Imaging of the craniocervical junction : a pictorial review

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Abstract

The craniocervical junction is a complex anatomical structure comprising the occiput, the atlas and the axis. The craniocervical junction plays an important role in maintaining stability, protection and supporting neurovascular structures. The craniocervical junction can be affected by a wide range of congenital variants, traumatic, degenerative, inflammatory and tumoral pathologies. The purpose of this pictorial review is twofold: (1) to review the normal anatomy of the craniocervical junction (2) to present the most common anatomic variants and pathologic conditions affecting the craniocervical junction.

Keywords

►Cervical spine
► Craniocervical junction
► Conventional Radiography
►Magnetic resonance imaging
► Computed tomography.

Anatomy of the cranicocervical junction

The craniocervical junction (CCJ) is a complex anatomical structure that includes the occiput, the atlas, the axis, a variety of ligaments and muscles. The CCJ is required for maintaining the stability and protection of vital neurovascular structures. On the other side a flexible mobility has to be preserved to allow neck mobility. The two major joints are the atlanto-occipital joint and the atlanto-axial joint. In the atlanto-occipital joint, the biomechanical features are mainly established by bony structures, whereas those of the atlanto-axial joint are primary determined by ligamentous structures[1].

The foramen magnum is located at the base of the occipital bone and compromises the clivus anteriorly, the squamosal portion posteriorly and the condyles in between[2]. The occipital condyles are angled inferomedially, limiting the mobility of the atlanto-occipital joint**.** The hypoglossal canal is located laterally of the condyles while the jugular foramen lays anterolateral of the condyles.

The first cervical vertebra, the atlas, is a ring-shaped structure that has no vertebral body or spinous process[2]. The atlas consists of two lateral masses that are connected via a short anterior arch and a long posterior arch. The superior surface of each lateral mass interacts with the occipital condyles while the inferior surface articulates with the superior articular facet of the axis. The groove for the vertebral artery is formed at the base of the posterior arch. The transverse foramen is located between the lateral mass of the atlas and the transverse process[2].

The second cervical vertebra is the axis and it contributes to the atlanto-axial joint (C1-C2). The ventral oval facet of the odontoid process projects cranially into the atlas and articulates

at the dorsal surface of anterior arch of the atlas. The spinous process of the axis is large and concave at its caudal border[2].

The ligamentous anatomy of the CCJ is complex but it is essential to provide motion and stability of the neck. It can be divided into external and internal ligaments (►**Fig. 1a-d**). The external ligamentous structures are further subdivided into anterior and posterior ligament and the ligamentum nuchae. The internal ligaments are subdivided into an anterior, middle and posterior group.

External ligaments

The anterior external ligaments of the CCJ are the anterior longitudinal ligament (ALL), the anterior atlanto-occipital ligament (AAOL) and the anterior atlanto-axial ligament (AAAL). The ALL runs anteriorly of the vertebral bodies. The AAOL connects the anterior aspect of the atlas to the anterior part of the foramen magnum, located posteriorly to the prevertebral muscles[3]. The AAAL attaches the caudal part of the anterior arch of the atlas to the anterior border of the axis body.
The posterior atlanto-occipital membrane (PAOM), the posterior atlanto-axial ligament (PAAL) and the extension of the ligamentum flava all make part of the posterior external ligaments. The PAOM is a wide ligament that attaches the posterior part of the foramen magnum with the posterior arch of the atlas, in continuity with the PAAL and the ligamentum flavum[3,4]. Anteriorly, this ligament is in proximity with the dura mater and posteriorly with the rectus capitis posterior minor muscle. The ligamentum nuchae is an extension of the supraspinous ligament connecting the spinous process of C7 to the external occipital protuberance[1].

Internal ligaments

The anterior part compromises a thin apical ligament that extends from the superior part of the odontoid process to the anterior base of the occiput. It is located between the AAOM and the cruciform ligament. The thicker alar ligaments have a V-shaped configuration that attaches the posterior surface of the dens with the base of the skull typically in a caudocranial direction[5].
The cruciform/cruciate ligament is one of the most important ligaments of the body, consisting of the transverse ligaments, accessory ligaments, superior and inferior extensions. The transverse ligament is 6-7 mm in height and connects the dorsal aspect of the odontoid process with a tubercle arising from the inner aspect of each lateral mass of the atlas[2,3,6]. It allows rotation at the atlanto-axial joint, serving as one of the primary stabilizers of the CCJ.
The tectorial membrane is the posterior group of the internal ligaments, situated dorsal to the cruciate ligament. It is a longitudinally oriented strong band of fibers that extends from the posterior longitudinal ligament attaching to the axis body and the body of the dens and finally attaching widely to the clivus of the occipital bone[3].

Variants and developmental abnormalities

There is a wide variety in developmental anomalies of the CCJ that may mimic fractures.

Variants and anomalies of the atlas

A condylus tertius is the result of a failure of integration of the proatlas with the condyle, leading to a well-delineated accessory ossicle located at the anteromedial margin of the occipital condyle[7].
Rachischisis can occur at the posterior and anterior arch of the atlas(►**Fig. 2**). Both variants are rare. Absence of the posterior arch can be associated with atlanto-axial subluxation and can result in spinal canal stenosis[1]. Split atlas refers to the combination of both posterior and anterior rachischisis[1]. *Developmental clefts* of the arch of the atlas are more common, frequently affecting the posterior arch. Most posterior arch clefts are situated at the midline, while lateral clefts account for 3%[8]. Clefts of the anterior arch can lead to dysraphic anomaly of the meninges or spinal cord and wide clefts may lead to atlas instability[8].
The *arcuate foramen* occurs due to calcification of the posterior atlanto-occipital membrane (►**Fig. S1 in online supplementary material**)[9].

Variants of the axis

A persistent *ossiculum terminale* is the failure of the fusion of the secondary ossification center of the dens, that normally fuses by the age of 12 year[1].
The more common *os odontoideum* is a well delineated separate ossicle situated cranially to a hypoplastic base of the dens at the location of the odontoid process (►**Fig. 3**). The anterior arch of the atlas may be hypertrophic[1]. Associated incompetence of the cruciate ligament can result in atlanto-axial instability[8]. This may further result in severe spinal canal stenosis due to a wider gap between the os odontoideum and the axis body extending above the level of the articular facets.

Pathology

Congenital disorders

Chiari malformation

Chiari malformation refers to a group of conditions affecting the posterior fossa that generally have the common feature of displacement of the cerebellar tonsils through the foramen magnum. It is subdivided into 5 subtypes (►**Table 1**).

Table 1	**Subtypes of Chiari malformation**
Subtype	*Characteristics*
Type 0	Significant syrinx in absence of tonsillar herniation
Type 1	Herniation of cerebellar tonsils in the foramen magnum
Type 1.5	Herniation of cerebellar tonsils and brainstem
Type 2	Type 1.5 + myelomenigocele
Type 3	Small posterior fossa + cervical/ occipital encephalocele +/- variable neural herniation
Type 4	Cerebellar hypoplasia

Type 5	Absence of cerebellum+ herniation of the occipital lobe in the foramen magnum

Patients with a type 1 Chiari malformation (►**Fig. 4**) will present with a wide variety of symptoms that can be categorized in three subgroups[10]. The first subgroup consists of headache due to cerebrospinal fluid obstruction, typically produced during Valsalva[11]. Hydrocephalus can be due to tonsillar herniation into the foramen magnum resulting in transient increase in intracranial pressure. The second subgroup exists of a wide variety of symptoms (absent gag reflex, hoarseness, nystagmus, …) caused by brainstem, cerebellar or cranial nerve dysfunction. Central sleep apnea is the result of brainstem or medullary compression, sometimes seen in children. The last subgroup manifests as spinal cord dysfunction resulting in upper and lower motor neuron signs due to syringomyelia.

MRI imaging is the preferred technique to evaluate Chiari malformation, particularly in the sagittal plane. The radiologic definition of Chiari malformation is inferior herniation of the cerebellar tonsils >5mm below the foramen magnum in adults (>3mm in children), measured on a midline sagittal image[12]. Although this finding may be found in the asymptomatic population, the presence of “peg-like” or “pointed” tonsils and effacement of CSF are radiologic features that are more likely associated with symptomatic patients[10,13].

Osteogenesis imperfecta

Osteogenesis imperfecta (OI) is a rare skeletal dysplasia, which is mostly inherited autosomal dominantly and rarely autosomal recessively. This genetic disorder affects type I collagen, that is mainly located in bone, ligaments, dentin and sclerae generally resulting in bone fragility and osteopenia[14]. It has an incidence of 1/10 000-20000 newborns and 4 subtypes are described[15,16]: *type I* is a non-bone-deforming OI with blue sclerae, *type II* is perinatal lethal associated with multiple intrauterine fractures, *type III* comprises severe progressive deformities, *type IV* results in variable bone deformities, a short stature and often normal sclerae. In the axial skeleton compression fractures, kyphoscoliosis and lumbar spondylosis are common[17].

In patients with OI the hallmark at the CCJ is basilar impression in association with basilar invagination[17]. Basilar impression is characterized by infolding of the occipital condyles and the base of the posterior fossa is getting elevated in the margins of the foramen magnum[17,18]. This leads to a horizontally oriented clivus, resulting in an abnormal rostrally located clivus-atlas-odontoid complex. These features give rise to the ”tam-o’-shanter” or “Darth Vader” skull (►**Fig. 5**). [14,18]. Basilar impression is sequentially accompanied by upward migration of the upper cervical column. Subsequently elevation and traction of the brainstem may produce brainstem dysfunction and impingement of the cranial nerves[17]. There is also predisposition to changes in cerebrospinal fluid circulation resulting in hydrocephalus. Basilar invagination can be demonstrated by conventional radiograph (CR) where the tip of the dens axis protrudes >5 mm above the Chamberlain line or >7 mm above the McGregor line. Visualisation of these landmarks may be challenging on CR due to the bone deformity and osteopenia.

Morquio syndrome

Morquio's syndrome, also named mucopolysaccharidosis type IV, is an autosomal recessive disease of glycosaminoglycan metabolism due to deficiency of the lysosomal enzyme galactosamine-5-sulphate sulphatase[19]. There are 2 types: type IV A and B. This syndrome leads to skeletal dysplasia with abnormalities of the cartilage and ligaments and is often diagnosed in the second year of life. Morquio's disease is a spondylo-epi-(meta)-physeal dysplasia[20]. Type IVA, which is the most severe type, is characterized by kyphoscoliosis, a small and abnormal thoracic cage, genu valgum, pectus carcinatum and a gait disturbance[19,20].

Cervical and thoracic platyspondyly together with beaked-shaped vertebra are common in MPS IVA[19,20]. At the CCJ Morquio's syndrome is typically characterized by a dysplasia of C2 and more specifically odontoid hypoplasia in the most severely affected patients (►**Fig. 6**). Another feature that may be present is a delayed ossification of the arches of C1, sometimes resulting in atlantoaxial instability. These two features may predispose and lead to atlantoaxial subluxation, basilar invagination, variable degree of spinal cord compression, myelopathy, secondary severe neurological deficit, quadriplegia, respiratory compromises and eventually death.

CR of the cervical spine taken during flexion and extension can show atlantoaxial instability. Reactive thickening of the surrounding connective tissues, caused by the accumulation of glycosaminoglycan, results in a soft tissue mass at the CCJ, predisposing to spinal cord compression[19,20]. The soft tissue mass is of low to intermediate signal on T1-weighted images (WI) and low signal on T2-WI on MRI.

Table 2. Other congenital malformations affecting the craniocervical junction
Chondroosteo-dystrophy
Cleidocranial dysostosis
Klippel-feil syndrome
Down syndrome
Achondroplasia

Trauma

The CCJ is affected in 30% of cervical spine trauma[21]. The two major mechanism for CCJ injuries are a direct blow on the head and a deceleration of the body. Craniocervical instability may result in secondary neurological injury. CR has long been the first imaging technique to evaluate the cervical spine after trauma but has largely been taken over by CT. Since CCJ injuries have the highest association with blunt cerebrovascular injuries, CT angiography is the preferred vascular imaging modality to assess vascular damage. MRI is the best imaging modality to investigate ligamentous injury, spinal cord injury and soft tissue. MRI is required in case of suspected ligamentous injury, to further evaluate CCJ instability, patients with neurologic deficits with/without normal CT and patients with discrepancy between the symptoms and level of injury on CT[22].

Basi-occiput fractures and occipital condyle fractures

Basiocciput fractures often occur in high-energy trauma and is associated with a high mortality due to close proximity of the brainstem, cranial nerves and vascular structures. Retroclival epidural hematoma is a rare injury, associated with occipital fractures, more frequently seen in children.
Occipital fractures have a variable clinical presentation. These fractured can be classified into 3 types according to Anderson and Montesano classification (►**Fig. 7**)[23]. *Type 1* fracture is a comminuted impaction fracture without displacement of the bony fragments into the foramen magnum. *Type 2* fracture involves the basiocciput extending into the occipital bone, that preserves stability due to an intact tectorial membrane and alar ligaments. *Type 3* fracture is an avulsion fracture located at the occipital attachment of the alar ligament that may result in displacement of the fragment into the foramen magnum. It is considered unstable due to probable associated injuries of the tectorial membrane.

Craniocervical dislocations

Atlanto-occipital dislocation exists of a complete dislocation (AOD) or subluxation (AOS) of the joint, both resulting from a high-energetic trauma. AOS is more frequently seen in children due to an underdevelopment of the main ligamentous stabilizers and a less concave articulation of the atlas[1]. AOD is considered as a complete disruption of the ligaments of the atlas and the occiput. This is associated with a high mortality and morbidity due to a highly unstable severe injury [21]. AOD can be subdivided into 3 subtypes determined by the direction of luxation(►**Fig. S2 in online supplementary material**)[1]. A *type 1* is an anterior luxation of the occipital condyles towards the atlas, while a *type 2* injury is a vertical luxation and a *type 3* is a posterior displacement of the occipital condyles in relation to the atlas. Patients with congenital abnormalities (►**Table 2**) at the CCJ are more prone to dissociative injuries of the CCJ.

Atlanto-axial rotatory subluxation

Atlanto-axial rotatory subluxation (AARS) is an abnormal motion of the atlas in relation with the axis that is more common in de paediatric population. Atlanto-axial rotatory fixation refers to locking of the facets after trauma. The most frequently used classification is developed by Fielding and Hawkings (►**Fig. 8**). [24]. *Type 1* is a rotatory subluxation without anterior displacement of the atlas (<3mm). *Type 2* is an anterior subluxation of 1 lateral mass of the atlas of 3-5mm, while a *type 3* is an anterior bilateral displacement of the lateral mass with more than 5mm. *Type 4* is a posterior displacement of the atlas with respect to the axis. CT angiography may be indicated since there is an association with vertebral artery injury.

Atlas fractures (C1 vertebra)

Atlas fractures mostly occur due to an axial loading mechanism and is often accompanied with fractures of the axis and the subaxial spine. Atlas fractures can be classified into 5 subtypes according to Jefferson (►**Fig. 9**)[1,21]. *Type 1* fracture is an isolated posterior arch fracture, while a *type 2* only affects the anterior arch. *Type 3* fracture is a bilateral fracture of the anterior and posterior arch. A *type 4* fracture involves the lateral mass and a type 5 fracture is a transverse avulsion fracture of the anterior arch which is considered unstable

due to avulsion of the longus colli or the atlantoaxial ligament. Atlas fractures may be associated with injury of the transverse ligament that may result in instability of the CCJ junction. An atlanto-dental interval > 3mm in adults and > 5 mm in children on CR or CT is considered highly suggestive for involved ligamentous injury of the transverse ligament[1]. One should not misinterpret a 'pseudospread of the atlas' for a Jefferson fracture. This is an entity that occurs in children under 7 years of age consisting of a normal overhanging edges (up to 6 mm) of the lateral masses of C1 over the lateral edges of C2 due to a discrepancy in growth of the atlas and axis[25].

Axis fractures

Fracture of the C2 vertebra can be mainly subdivided into odontoid fractures and Hangman fractures.

Odontoid fractures are the most common fractures of the CCJ that is the result of a hyperflexion-hyperextension trauma. A classification system is developed by Anderson and D'Alonzo (►**Fig. 10**)[26]. A *type 1* fracture is an oblique avulsion fracture of the tip of the odontoid process that heals normally in case of an isolated fracture using a conservative treatment or halo immobilization. Fractures that are located at the base of dens and the vertebral body are categorized as an unstable *type 2* fracture. A type 2 fracture is more vulnerable for non-union A *type 3* fracture goes through the body of the axis extending in the lateral masses of the vertebral body. The last group has the best prognosis of healing.

Hangman fractures are bilateral fractures of the pars interarticularis of the axis that occurs due to a hyperextension trauma. It can be classified using the Levine and Edwards (►**Fig. S3 in online supplementary material**) classification or the AO spine classification.

Inflammation and infection

Rheumatoid arthritis

Rheumatoid arthritis (RA) is a systemic chronic inflammatory disease, often symmetrical, affecting the peripheral skeleton. It has a female predilection[27]. When affecting the axial skeleton, the cervical spine is one of the most common sites involving the CCJ. There is a wide variability in clinical presentation of patients with RA when the CCJ is affected. Most patients are asymptomatic but symptomatic patients can present with neck pain, decreased range of motion, compression neuropathy, occipital headache and in severe cases spinal cord compression may lead to stroke and sudden death[28,29]. Progressive RA disease will result in the formation of pannus and chronic inflammatory infiltration, secondary leading to bone erosion and laxity of the ligamentous insertion. Finally, this results in instability of the CCJ[27,29].

CR is the first-line imaging technique to investigate cervical involvement of RA. Atlanto-axial subluxation (AAS), vertical subluxation (VS), lateral AAS (LAAS) and sub-axial subluxation (SAS) must be evaluated initially. *AAS* is the abnormal movement between C1 and C2 in different directions, where anterior displacement is the most common one. Lateral displacement of the atlantoaxial joint is rare, best evaluated on open mouth view. VS is the superior displacement of dens resulting in basilar invagination[28]. SAS means there is instability below

the axis with> 3.5 mm horizontal displacement on lateral radiographs of two vertebral bodies relative to each other [30]. It can involve multiple levels then leading to staircase deformity[31].

CT is a superior imaging modality compared with CR for more precise measurement for cervical instability. It also better demonstrates bony destruction, ankylosis and pseudoarthrosis. CT is indicated for patients in case of pre-operative planning and with CT angiography the vertebral artery can be evaluated[28,30]. Contrast-enhanced CT is useful for patients to investigate the inflammatory pannus in case of contra-indications for MRI[31].

MRI should be performed in case of abnormal CR/CT or neurological deficits. It is most sensitive for detection of inflammatory degenerative pseudotumor (►**Fig. 11**). Contrast-enhanced MRI differentiates between the various forms of pannus inflammation[28,32]. Hypervascular pannus has a high signal intensity on T2-WI and will enhance while hypovascular pannus is of intermediate signal on T2-WI and shows moderate enhancement. Fibrous pannus is hypo-intense on T2-WI and does not enhance. MRI can also be used for assessment of the cervicomedullary angle (normal >135°) to evaluate VS[31]. Dynamic MRI is used to evaluate the subarachnoid space in case of normal static MRI[28,29].

Tuberculous spondylitis

Mycobacterium tuberculosis infection has a predilection for the respiratory system, although spinal involvement affects approximately 1% of infected patients[33]. The disease is often located at the thoracolumbar region and the cervical spine is rarely affected[34,35]. Bacterial tuberculous spondylodiscitis, also called Pott's disease, is a destructive form of tuberculosis, resulting from hematogenous spread[33].

CR remains the initial screening imaging technique. In later stages of the disease, erosion of the vertebral endplates, loss of disk height, vertebral erosion, bone sequestration, sclerosis and para-/prevertebral abscess is seen[34,36].
CT can detect the features earlier than CR and evaluates bony destruction optimally. Calcification in the paravertebral masses is characteristic for tuberculous spondylodiscitis and is best visualized on CT[34,37]. Multiloculated granulomatous abscess may be seen on contrast-enhanced image.
MRI can detect the disease in the earliest stage. There are 4 different MRI patterns of the disease described[33,34,36,38]. First, para-discal lesion is the most common pattern that begins in vertebral metaphysis leading to direct disk involvement. The earliest findings are characterized by low T1 signal intensity and high signal intensity on T2/ STIR resembling a type 1 Modic changes. An obvious enhancement of the affected disc is seen following gadolinium contrast administration. Abscess formation is caused by spreading of the infection into the paravertebral soft tissue. Secondary, anterior lesions are located subperiostally in the corner of the vertebral body. It evolves in striping the periosteum and the anterior longitudinal ligament from the anterior vertebral body. Disease progression of anterior lesions leads to subligamentous abscess with sparing of the disc. Third, central lesions typically occur intravertebral and the intervertebral disc remains healthy. Finally lesions of the posterior elements posterior lesions are rarely seen and is often the result of complete vertebral disease.

Crowned dens syndrome

The crowned dens syndrome is an inflammatory disease characterized by the crystal deposits in the ligaments surrounding the odontoid peg. They present with acute pain at the base of the skull or the high cervical region together with fever, general signs of inflammation and elevated inflammatory markers[39]. The crystal deposits can be the result of both calcium pyrophosphate dihydrate or calciumhydroxyapatite disease[40]. It mostly affects the elderly. CT is the best imaging modality and demonstrates periodontal calcification mostly located in the cruciform ligament(►**Fig. 12**)[41].

Tumor

Plasmacytoma

Plasmacytoma is the most common primary bone neoplasm in adults consisting of discrete masses due to focal proliferation of malignant monoclonal plasma cells. It can be subdivided in solitary plasmacytoma (70%) and extramedullary plasmacytoma (30%)[42,43]. The mean age of patients with plasmacytoma is around 55 year. The thoracic spine is most frequently affected. Location at the CCJ is rare. Solitary plasmacytoma may evolve to multiple myeloma, which are the most common primary bone malignancy in adults[44,45].

CR shows a well-delineated osteolytic lesion [44]. CT better assesses the lytic and expansile bony component of plasmocytomas. The soft tissue component is often hyperdense and enhance homogeneously on CT. On MRI the lesion is of hypointense signal on T1-WI and intermediate to high signal on T2[44]. There are thickened cortical struts probably due to stress reaction of the lytic process of the lesion that results in a characteristic feature of a “minibrain” on axial MR/CT images[46]. There is a heterogeneous contrast enhancement on MRI.

Metastasis

The spine is the most frequent site for bone metastasis occurring in 10 % of cancer patients, although metastasis that occur at the CCJ only account for 0,5% of all the spine metastasis[47]. Metastasis causing deformity of the CCJ may lead to instability, relevant pain, cranial nerve palsies and sometimes death. It frequently affects patients older then 50 years old: in women the most common cause is breast cancer, while in men prostate cancer is the most common[44]. Three types of bone metastasis have been described: osteolytic (71%), osteoblastic (8%) and mixed osteolytic-osteoblastic (21%)[44].

On CT osteoblastic bone metastasis appears as an hyperdense sclerotic lesion. Osteolytic metastasis is a radiolucent lesion with a soft tissue density on CT, demonstrating bone destruction. Osteoblastic metastasis is hypo-intense on both T1-WI and T2-WI on MRI. The halo sign, which is a slight hyperintense rim surrounding the lesion representing bone marrow edema, is seen on T2-WI and STIR images[48]. An osteolytic metastasis appears as a hypo-intense lesion on T1-WI and hyperintense lesion on T2-WI. Both types of metastasis will generally enhance. When a bone metastasis extends epidural, this results in a draped curtain sign due to a posterior displacement of the lateral parts of the posterior longitudinal ligament and a strong medial fixation, giving it a bilobular appearance, best seen on axial MR images.

Meningioma

Meningioma is the most common benign slow-growing tumor and it is the second most common spinal cord tumor, accounting for 12% of all meningiomas[50]. A meningioma located at the CCJ is rather a rare entity originating from the meninges of the clivus and the upper part of the axis[51]. It has a female predominance usually present after the age of 50. Multiple meningiomas are associated with neurofibromatosis type 2[44]. There is a wide variety of symptoms depending on the extent of the tumor, the growth rate and the tumor localization.

On CT meningioma appears as a well-circumscribed hyperdense lesion. In a minority of the cases, calcifications can be seen. Meningiomas have a wide dural attachment and are hyper/iso-intense on T2-WI and intermediate/hypointense on T1-WI. The tumor shows a moderate homogenous enhancement on MRI (►**Fig. 13)** and CT. The dural tail sign is a non-specific feature associated with a meningioma[52].

Schwannoma

Schwannoma is a benign peripheral nerve sheath tumor, composed of Schwann cells. It is mostly located intracranial. Spinal schwannoma located at the CCJ is uncommon. It is often found intradural and extramedullary and have a peak incidence in the fourth and fifth decade[44]. It is associated with neurofibromatosis type 2. Generally, schwannoma is asymptomatic until compression of the spinal cord resulting in radiating pain, sensory or motor deficit.

CT shows pedicle erosion, remodeling with secondary widening of the neuroforamen, posterior vertebral scalloping and a paravertebral hypo- or slightly hyperdense soft tissue mass[44,53]. Schwannoma is hypo-intense on T1-WI and hyper-intense on T2-WI and STIR on MRI. Homogenous intense contrast enhancement is seen. A target sign may be seen in schwannoma, which consist of a hyperintense rim with a central area of low signal[54]. The main differential diagnosis is a neurofibroma. Occasionally schwannoma is associated with hemorrhage, cyst formation and fatty degeneration which are rare features in neurofibroma[44].

Chordoma

Chordoma is a rare malignant, slow-growing, tumor arising from notochordal rests. It often presents in middle-aged patients in the fifth to seventh decades with a male predilection[44,55,56]. Chordoma is typically located at the midline and mostly affects the sacrum (50%) followed by the skull base (35%), where it is often seen in the clival area, and 15 % along the spine. It is an aggressive tumoral lesions with a high risk of local recurrence and metastases to the lung and bone[57]. Generally, chordoma is both sporadic and hereditary lesions. Patients presents with symptoms due to mass effect on the surrounding structures.

On CR, most lesions are osteolytic or mixed. Vertebral sclerosis resulting in an ‘ivory vertebral body’, is less common. Multilevel involvement is frequent. CT is essential for accurate evaluation of the degree of bony destruction and vertebral sclerosis. An associated lobular

soft tissue component may cause pressure erosion of the outer cortex and adjacent spiculated periosteal reaction[59]. Another feature consists of irregular intratumoral calcifications, but this is more common when located in the sacrum[55,56]. Enhancement is mild, either diffuse or with a peripheral pattern.

MRI shows a low to intermediate signal on T1-WI and T2-WI hyperintense signal with intralesional hypo-intense septa on T2-WI (►**Fig. 14**), similar to signal of the nucleus pulposus of the disk. Intratumoral hemorrhage and/or proteinaceous mucinous material is responsible for multiple small foci of T1 hyperintensity[60]. There is thick inhomogeneous peripheral and septal enhancement.

Ecchordosis physaliphora

Ecchordosis physaliphora (EP) is a rare congenital hamartomatous lesion, originating from an ectopic notochordal rest. It is often found incidentally and is asymptomatic. Large lesion may cause compression of the brainstem or the cranial nerves. EP is typically located in the prepontine region at the retroclival midline where it is attached to the posterior wall of the clivus with a small pedicle, in close proximity of the basilar artery[61,62].

CT reveals a well-delineated radiolucent clival defect. Presence of an osseous stalk is pathognomonic[61,62]. On MRI, EP is a well-circumscribed, round, extra-axial lesion that is hypo-intense on T1-WI and hyperintense on T2-WI (►**Fig. 15**). The lesion has a signal similar to cerebrospinal fluid. The lesion does not enhance which may be a helpful feature in differentiating it from a chordoma[61,63]. Surgery is only necessary in symptomatic cases.

Chondrosarcoma

Chondrosarcoma (CS) is a rare malignant nonlymphoproliferative neoplasm, originating form cartilage tissue. Its main feature is the formation of chondroid matrix[55]. It mostly affects the thoracic spine and 2% is located at the skull base[64]. CS of the skull base most frequently originate from the cartilage of the petroclival region and spheno-ethmoidal sutures[57]. There is a male predilection and generally presents as a low-grade tumor. Therefore, CS is often found paracentral which is a main difference in comparison with chordomas that are typically located at the midline[64,65].

CT and MRI are complementary in characterizing a chondrosarcoma. On CT, the ring-and-arc calcification is characteristic for neoplasms of chondroid origin[66]. CS is hypo-intense on T1-WI and hyperintense on T2-WI due to the nonmineralized areas of hyaline cartilage (►**Fig. 16**). A peripheral and septal contrast enhancement, similar to chordoma, is generally seen in chondrosarcoma. On diffusion WI, CS has a higher ADC value in comparison with a chordoma, which may be helpful in differentiating a chordoma from a chondrosarcoma[65].

Hemangioma

Hemangioma is common benign tumor of vascular origin and is often incidental findings on imaging. A primary intraosseous cavernous hemangioma (PICH) is frequently seen in the vertebral body followed by the skull, where it mostly affects the calvarium and is rarely found

at the clivus[67]. PICH is asymptomatic in most cases, but it can result in a pathologic fracture or causing mass effect on surrounding structures[55].

CR presents PICH as a well-delineated radiolucent lesion. The honeycomb appearance, corduroy sign and white polka dot sign are features often seen in thoracal vertebrae. At the calvarium PICH presents as a lytic area with thickened trabeculae, which has a spoke-wheel appearance (►**Fig. S4 in online supplementary material**). The appearance on MRI depends on the volume of fat intratumorally and vascular components. Generally, on T1-WI PICH manifests as a hyperintense lesion in case of lipid-rich PICH and intermediate to low signal intensity on T1-WI when lipid-poor[67,68]. Due to the vascular nature, PICH is hyperintense on T2-WI and shows vivid or heterogeneous contrast enhancement[69].

References

1. Offiah CE, Day E. The craniocervical junction: embryology, anatomy, biomechanics and imaging in blunt trauma. *Insights Imaging*. Feb 2017;8(1):29-47.
2. Menezes AH, Traynelis VC. Anatomy and biomechanics of normal craniovertebral junction (a) and biomechanics of stabilization (b). *Childs Nerv Syst*. Oct 2008;24(10):1091-100.
3. Tubbs RS, Hallock JD, Radcliff V, et al. Ligaments of the craniocervical junction. *J Neurosurg Spine*. Jun 2011;14(6):697-709.
4. Tubbs RS, Wellons JC, Blount JP, Oakes WJ. Posterior atlantooccipital membrane for duraplasty. Technical note. *J Neurosurg*. Sep 2002;97(2 Suppl):266-8.
5. Krakenes J, Kaale BR, Rorvik J, Gilhus NE. MRI assessment of normal ligamentous structures in the craniovertebral junction. *Neuroradiology*. Dec 2001;43(12):1089-97.
6. Fiester P, Rao D, Soule E, Orallo P, Rahmathulla G. Anatomic, functional, and radiographic review of the ligaments of the craniocervical junction. *J Craniovertebr Junction Spine*. 2021;12(1):4-9.
7. Jain N, Verma R, Garga UC, Baruah BP, Jain SK, Bhaskar SN. CT and MR imaging of odontoid abnormalities: A pictorial review. *Indian J Radiol Imaging*. 2016;26(1):108-19.
8. Smoker WR, Khanna G. Imaging the craniocervical junction. *Childs Nerv Syst*. Oct 2008;24(10):1123-45.
9. Ahn J, Duran M, Syldort S, et al. Arcuate Foramen: Anatomy, Embryology, Nomenclature, Pathology, and Surgical Considerations. *World Neurosurg*. Oct 2018;118:197-202.
10. McClugage SG, Oakes WJ. The Chiari I malformation. *J Neurosurg Pediatr*. Sep 01 2019;24(3):217-226.
11. Tubbs RS, Beckman J, Naftel RP, et al. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. *J Neurosurg Pediatr*. Mar 2011;7(3):248-56.
12. Pindrik J, McAllister AS, Jones JY. Imaging in Chiari I Malformation. *Neurosurg Clin N Am*. Jan 2023;34(1):67-79.
13. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM. Significance of cerebellar tonsillar position on MR. *AJNR Am J Neuroradiol*. 1986;7(5):795-9.
14. Wallace MJ, Kruse RW, Shah SA. The Spine in Patients With Osteogenesis Imperfecta. *J Am Acad Orthop Surg*. Feb 2017;25(2):100-109.
15. Van Dijk FS, Sillence DO. Osteogenesis imperfecta: clinical diagnosis, nomenclature and severity assessment. *Am J Med Genet A*. Jun 2014;164A(6):1470-81.
16. Marom R, Rabenhorst BM, Morello R. Osteogenesis imperfecta: an update on clinical features and therapies. *Eur J Endocrinol*. Oct 2020;183(4):R95-R106.
17. Menezes AH. Specific entities affecting the craniocervical region: osteogenesis imperfecta and related osteochondrodysplasias: medical and surgical management of basilar impression. *Childs Nerv Syst*. Oct 2008;24(10):1169-72.
18. Janus GJ, Engelbert RH, Beek E, Gooskens RH, Pruijs JE. Osteogenesis imperfecta in childhood: MR imaging of basilar impression. *Eur J Radiol*. Jul 2003;47(1):19-24.
19. Hughes DG, Chadderton RD, Cowie RA, Wraith JE, Jenkins JP. MRI of the brain and craniocervical junction in Morquio's disease. *Neuroradiology*. May 1997;39(5):381-5.
20. Padash S, Obaid H, Henderson RDE, et al. A pictorial review of the radiographic skeletal findings in Morquio syndrome (mucopolysaccharidosis type IV). *Pediatr Radiol*. Jan 11 2023?
21. Izzo R, Popolizio T, Balzano RF, et al. Imaging of cranio-cervical junction traumas. *Eur J Radiol*. Jun 2020;127:108960.

22. Santos-Nunez G, Lo HS, Kotecha H, Jose J, Abayazeed A. Imaging of Spine Fractures With Emphasis on the Craniocervical Junction. *Semin Ultrasound CT MR*. Aug 2018;39(4):324-335.
23. Anderson PA, Montesano PX. Morphology and treatment of occipital condyle fractures. *Spine (Phila Pa 1976)*. Jul 1988;13(7):731-6.
24. Fielding JW, Hawkins RJ. Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). *J Bone Joint Surg Am*. Jan 1977;59(1):37-44.
25. Lustrin ES, Karakas SP, Ortiz AO, et al. Pediatric cervical spine: normal anatomy, variants, and trauma. *Radiographics*. 2003;23(3):539-60.
26. Anderson LD, D'Alonzo RT. Fractures of the odontoid process of the axis. *J Bone Joint Surg Am*. Dec 1974;56(8):1663-74.
27. Chu EC, Wong AY, Lee LY. Craniocervical instability associated with rheumatoid arthritis: a case report and brief review. *AME Case Rep*. 2021;5:12.
28. Ellatif M, Sharif B, Baxter D, Saifuddin A. Update on imaging of the cervical spine in rheumatoid arthritis. *Skeletal Radiol*. Aug 2022;51(8):1535-1551.
29. Shlobin NA, Dahdaleh NS. Cervical spine manifestations of rheumatoid arthritis: a review. *Neurosurg Rev*. Aug 2021;44(4):1957-1965.
30. Joaquim AF, Ghizoni E, Tedeschi H, Appenzeller S, Riew KD. Radiological evaluation of cervical spine involvement in rheumatoid arthritis. *Neurosurg Focus*. Apr 2015;38(4):E4.
31. Drosos AA, Pelechas E, Voulgari PV. Radiological Findings of the Cervical Spine in Rheumatoid Arthritis: What a Rheumatologist Should Know. *Curr Rheumatol Rep*. May 13 2020;22(6):19.
32. Stiskal MA, Neuhold A, Szolar DH, et al. Rheumatoid arthritis of the craniocervical region by MR imaging: detection and characterization. *AJR Am J Roentgenol*. Sep 1995;165(3):585-92.
33. Kumar Y, Gupta N, Chhabra A, Fukuda T, Soni N, Hayashi D. Magnetic resonance imaging of bacterial and tuberculous spondylodiscitis with associated complications and non-infectious spinal pathology mimicking infections: a pictorial review. *BMC Musculoskelet Disord*. Jun 05 2017;18(1):244.
34. Rivas-Garcia A, Sarria-Estrada S, Torrents-Odin C, Casas-Gomila L, Franquet E. Imaging findings of Pott's disease. *Eur Spine J*. Jun 2013;22 Suppl 4(Suppl 4):567-78.
35. Teka M, Ghozlen HB, Zaier AY, Hnia MB, Naouar N, Abid F. Cervical spine tuberculosis. *Pan Afr Med J*. 2020;37:7.
36. Garg RK, Somvanshi DS. Spinal tuberculosis: a review. *J Spinal Cord Med*. 2011;34(5):440-54.
37. Tali ET. Spinal infections. *Eur J Radiol*. May 2004;50(2):120-33.
38. Currie S, Galea-Soler S, Barron D, Chandramohan M, Groves C. MRI characteristics of tuberculous spondylitis. *Clin Radiol*. Aug 2011;66(8):778-87.
39. Scutellari PN, Galeotti R, Leprotti S, Ridolfi M, Franciosi R, Antinolfi G. The crowned dens syndrome. Evaluation with CT imaging. *Radiol Med*. Mar 2007;112(2):195-207.
40. Feydy A, Lioté F, Carlier R, Chevrot A, Drapé JL. Cervical spine and crystal-associated diseases: imaging findings. *Eur Radiol*. Feb 2006;16(2):459-68.
41. Lee GS, Kim RS, Park HK, Chang JC. Crowned dens syndrome: a case report and review of the literature. *Korean J Spine*. Mar 2014;11(1):15-7.
42. Nahi H, Genell A, Wålinder G, et al. Incidence, characteristics, and outcome of solitary plasmacytoma and plasma cell leukemia. Population-based data from the Swedish Myeloma Register. *Eur J Haematol*. Sep 2017;99(3):216-222.

43. Ozsahin M, Tsang RW, Poortmans P, et al. Outcomes and patterns of failure in solitary plasmacytoma: a multicenter Rare Cancer Network study of 258 patients. *Int J Radiat Oncol Biol Phys*. Jan 01 2006;64(1):210-7.
44. Pinter NK, Pfiffner TJ, Mechtler LL. Neuroimaging of spine tumors. *Handb Clin Neurol*. 2016;136:689-706.
45. Saad A, Azzopardi C, Haleem S, Czyz M, James SL, Botchu R. Tumours of the odontoid peg revisited. *Indian J Radiol Imaging*. 2020;30(4):420-426.
46. Major NM, Helms CA, Richardson WJ. The "mini brain": plasmacytoma in a vertebral body on MR imaging. *AJR Am J Roentgenol*. Jul 2000;175(1):261-3.
47. O'Sullivan MD, Lyons F, Morris S, Synnott K, Munigangaiah S, Devitt A. Metastasis Affecting Craniocervical Junction: Current Concepts and an Update on Surgical Management. *Global Spine J*. Dec 2018;8(8):866-871.
48. Schweitzer ME, Levine C, Mitchell DG, Gannon FH, Gomella LG. Bull's-eyes and halos: useful MR discriminators of osseous metastases. *Radiology*. Jul 1993;188(1):249-52.
49. Shah LM, Salzman KL. Imaging of spinal metastatic disease. *Int J Surg Oncol*. 2011;2011:769753.
50. Setzer M, Vatter H, Marquardt G, Seifert V, Vrionis FD. Management of spinal meningiomas: surgical results and a review of the literature. *Neurosurg Focus*. 2007;23(4):E14.
51. Dührsen L, Emami P, Matschke J, Abboud T, Westphal M, Regelsberger J. Meninigiomas of the Craniocervical Junction--A Distinctive Subgroup of Meningiomas. *PLoS One*. 2016;11(4):e0153405.
52. Sotoudeh H, Yazdi HR. A review on dural tail sign. *World J Radiol*. May 28 2010;2(5):188-92.
53. Parmar HA, Ibrahim M, Castillo M, Mukherji SK. Pictorial essay: diverse imaging features of spinal schwannomas. *J Comput Assist Tomogr*. 2007;31(3):329-34.
54. Bhargava R, Parham DM, Lasater OE, Chari RS, Chen G, Fletcher BD. MR imaging differentiation of benign and malignant peripheral nerve sheath tumors: use of the target sign. *Pediatr Radiol*. Feb 1997;27(2):124-9.
55. Orguc S, Arkun R. Primary tumors of the spine. *Semin Musculoskelet Radiol*. Jul 2014;18(3):280-99.
56. Murphey MD, Minn MJ, Contreras AL, et al. Imaging of spinal chordoma and benign notochordal cell tumor (BNCT) with radiologic pathologic correlation. *Skeletal Radiol*. Mar 2023;52(3):349-363.
57. Yamazawa E, Takahashi S, Shin M, et al. MRI-Based Radiomics Differentiates Skull Base Chordoma and Chondrosarcoma: A Preliminary Study. *Cancers (Basel)*. Jul 03 2022;14(13).
58. Nibu Y, José-Edwards DS, Di Gregorio A. From notochord formation to hereditary chordoma: the many roles of Brachyury. *Biomed Res Int*. 2013;2013:826435.
59. Cui JF, Hao DP, Chen HS, Liu JH, Hou F, Xu WJ. Computed tomography and magnetic resonance imaging features of cervical chordoma. *Oncol Lett*. Jul 2018;16(1):861-865.
60. Smolders D, Wang X, Drevelengas A, Vanhoenacker F, De Schepper AM. Value of MRI in the diagnosis of non-clival, non-sacral chordoma. *Skeletal Radiol*. Jun 2003;32(6):343-50.
61. Lakhani DA, Martin D. Ecchordosis physaliphora: Case report and brief review of the literature. *Radiol Case Rep*. Dec 2021;16(12):3937-3939.
62. Sarkar N, Chakravarthy S, Chakravarty R, Mukhopadhyay S. Radiological Diagnosis of a Rare Prepontine Lesion: Ecchordosis Physaliphora. *Cureus*. Apr 2022;14(4):e24335.

63. Chihara C, Korogi Y, Kakeda S, et al. Ecchordosis physaliphora and its variants: proposed new classification based on high-resolution fast MR imaging employing steady-state acquisition. *Eur Radiol*. Oct 2013;23(10):2854-60.
64. Awad M, Gogos AJ, Kaye AH. Skull base chondrosarcoma. *J Clin Neurosci*. Feb 2016;24:1-5.
65. Hasegawa H, Shin M, Niwa R, et al. Revisitation of imaging features of skull base chondrosarcoma in comparison to chordoma. *J Neurooncol*. Sep 2022;159(3):581-590.
66. Flemming DJ, Murphey MD, Carmichael BB, Bernard SA. Primary tumors of the spine. *Semin Musculoskelet Radiol*. 2000;4(3):299-320.
67. Liu JK, Burger PC, Harnsberger HR, Couldwell WT. Primary Intraosseous Skull Base Cavernous Hemangioma: Case Report. *Skull Base*. Nov 2003;13(4):219-228.
68. Hoyle JM, Layfield LJ, Crim J. The lipid-poor hemangioma: an investigation into the behavior of the "atypical" hemangioma. *Skeletal Radiol*. Jan 2020;49(1):93-100.
69. Alexiou GA, Lampros M, Gavra MM, Vlachos N, Ydreos J, Boviatsis EJ. Primary Intraosseous Cavernous Hemangioma of the Cranium: A Systematic Review of the Literature. *World Neurosurg*. Aug 2022;164:323-329. doi:10.1016/j.wneu.2022.05.107