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Case report - Thoracic general

Intrapulmonary glomus tumor in a young woman

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Abstract

A 21-year-old female patient presented with pneumonia and on chest roentgenogram a solitary pulmonary nodule was incidentally found. After an observation period she underwent left upper lobectomy because of documented tumor growth. Pathology showed an intrapulmonary glomus tumor of the proper type, which is a very rare occurrence. Literature review revealed only 11 published cases of this subtype. Radiological investigation is helpful for localization and characterization of the tumor. However, pathological examination is required for definitive diagnosis. Complete surgical excision is the treatment of choice. Although uncommon, glomus and carcinoid tumors should be considered in the differential diagnosis of solitary pulmonary nodules in young patients.

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Keywords: Lung, benign lesions; Glomus tumor; Immunohistochemistry; Thoracotomy; Lobectomy

1. Introduction

Glomus tumors are benign neoplasms originating from glomus bodies in the dermis or subcutis of the extremities [1,2]. Extracutaneous presentations occur but are rare, especially in visceral organs where glomus bodies are sparse or even absent. A case of an intrapulmonary glomus tumor is presented.

2. Case report

A 21-year-old non-smoking female patient was referred with a persisting intrapulmonary nodule in the left upper lobe. She was treated for a lobar pneumonia 18 months before and the solitary nodule was incidentally found on chest X-ray. This coin lesion grew slowly but steadily over time. On admission, the patient was asymptomatic. Apart from allergic asthma, her history was negative. Physical examination and routine laboratory tests were normal. Chest computed tomography (CT) showed a nodule without inlying calcifications in the parahilar region of the left upper lobe, with a size of 2.5×2.3 cm (Fig. 1a). Positron emission tomographic (PET) scanning showed a low to moderate isotope uptake. No other lesions were detected. Bronchoscopy showed no endobronchial involvement. Due to its central localization, the nodule was judged to be inaccessible for transthoracic puncture or thoracoscopic biopsy.

The patient underwent a muscle-sparing left anterolateral thoracotomy. After freeing the major fissure of the left lung, the nodule was palpated between the branches of

the bronchus and the pulmonary artery. Wedge excision of the nodule was not feasible because of the central position of the nodule inside the lobe and an upper lobectomy was performed. Macroscopically, a well-circumscribed soft mass of 2.5×2.2×2.4 cm was found. The non-encapsulated nodular lesion consisted of highly vascularized beige-to-pink tissue. Microscopically, the biopsy specimen showed a uniform distribution of epithelioid cells with a wide cytoplasm and homogeneous round to oval nuclei, occasionally with a round nucleolus (Fig. 1b and c). In between the epithelioid cells, thin-walled vascular structures and some spindle-shaped cells were present. No bronchi or pleura were involved in the tumoral process and all separately removed lymph nodes were negative. There were no signs suggestive of malignancy. Immunohistochemical study revealed tumor cells to be positive for CD34 and S-100; and negative for chromogranin, synaptophysin, h-caldesmon, α -smooth muscle actin, desmin, pancytokeratin, CD1- α , CD10 and CD31. The final pathological diagnosis was intrapulmonary glomus tumor of the proper type. The patient made an uneventful recovery and was discharged soon after surgery. Radiological examination after four months showed a normal status after left upper lobectomy.

3. Discussion

Glomus tumors are uncommon neoplasms composed out of uniform round cells, which are arranged in sheets that form collars around capillary-sized vessels [2]. They find their origin in the neuromyoarterial glomus bodies, which are arteriovenous connections that contribute to the regulation of blood flow and temperature on the surface of the skin. Benign glomus tumors can be divided into three subtypes [3]. In the glomus tumor proper type round glomus

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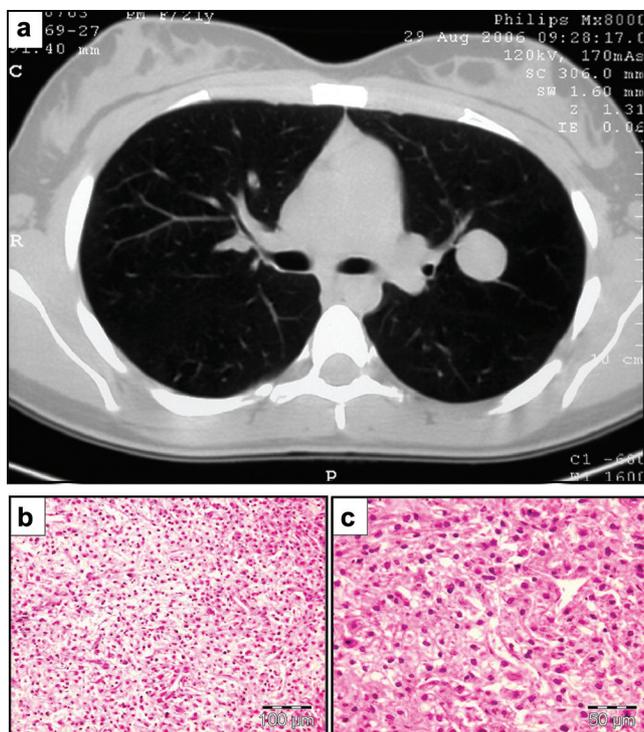


Fig. 1. (a) Axial computed tomography (CT) image showing a solitary nodule in the left parahilar region. (b and c) Biopsy specimen showing an abundance of epithelioid cells. Hematoxylin-eosin stain (b: $\times 200$; c: $\times 400$).

cells are the largest component as in our case, whereas in the glomangioma and glomangiomyoma types blood vessels and spindle cells are predominant. Despite that intrapulmonary glomus tumors are generally benign neoplasms,

four malignant cases have been described [4]. In the lower respiratory tract (excluding the trachea) 22 cases of glomus tumors have been reported in the literature, including only 11 cases of the proper subtype (Table 1). The average age of detection of the tumor is 48 years; with men affected twice as much as women. Tumor size is variable and ranges from 0.5 to 9.5 cm with most nodules in the 1–5 cm interval. As a result, these intrapulmonary lesions are significantly larger than their cutaneous counterparts; but in contrast, they usually remain asymptomatic and are accidentally found on routine chest roentgenogram [1].

The differential diagnosis consists of a wide variety of neoplasms, most notably: carcinoid tumor, hemangiopericytoma, paraganglioma, smooth muscle neoplasms and metastatic tumors. Carcinoid tumors are most commonly confused with glomus tumors, since they possess a similar cytological appearance [5]. In spite of this, they were excluded because of the absence of the somewhat typical coarsely granular to salt-and-pepper chromatin – in contrast to the finer chromatin pattern of glomus tumors – and the negative staining for neuroendocrine markers. Hemangiopericytoma is another rare tumor that should be considered. Nevertheless, a glomus tumor differs because of its round epithelioid cells and regular oval to round nuclei, whereas hemangiopericytoma consists of more polygonal to spindle-shaped cells with elongated nuclei [6]. Although spindle cells were found in the present case as well, their low quantity and focal distribution were not very suggestive for hemangiopericytoma. Moreover, the ramifying to staghorn vasculature pattern, which is archetypical for hemangiopericytoma, was absent. Paraganglioma, on the other hand, could be excluded because of the

Table 1
Studies reporting glomus tumors in the lower respiratory tract (excluding the trachea)

Author	Year of publication	Age at diagnosis (years)	Gender	Largest diameter nodule (cm)	Histological differentiation	Localization
Tang et al.	1978	67	Female	6.5	Glomangioma	Pulmonary
Mackay et al.	1981	19	Male	2.5	Malignant glomus tumor	Pulmonary/bronchogenic
Alt et al. [6]	1983	34	Male	2.0	Glomus tumor [†]	Pulmonary/bronchogenic
Koss et al.	1998	40	Male	1.1	Glomus tumor proper type	Pulmonary
		51	Male	1.5	Glomus tumor proper type	Pulmonary
Gaertner et al. [1]	2000	46	Female	7	Glomus tumor, locally infiltrative	Mediastinal [‡]
		20	Male	1.4	Glomus tumor proper type	Endobronchial
		65	Female	3	Glomus tumor proper type	Pulmonary
		40	Male	1.1 and 0.5	Glomus tumor proper type	Pulmonary
		69	Male	9.5	Glomangiosarcoma de novo	Pulmonary (+metastases)
Oizumi et al.	2001	48	Male	0.7	Glomus tumor [†]	Endobronchial
Yilmaz et al. [5]	2002	29	Female	1.5	Glomus tumor proper type	Pulmonary/bronchogenic
Hishida et al. [4]	2003	53	Male	2.5	Glomangiosarcoma arising within a glomus tumor	Pulmonary/bronchogenic
Hamaguchi et al.	2003	70	Female	1.0	Glomus tumor [†]	Pulmonary
Zhang et al.	2003	29	Male	2.5	Glomus tumor [†]	Pulmonary/bronchogenic
Ueno et al.	2004	50	Male	4	Glomus tumor [†]	Pulmonary
Vailati et al. [9]	2004	40	Male	5	Glomus tumor [†]	Pulmonary/bronchogenic
De Weerd et al.	2004	37	Male	n/a	Glomus tumor [†]	Endobronchial
Hariri et al.	2005	49	Male	n/a (large >2)	Malignant glomus tumor	Endobronchial
Sousa et al.	2006	62	Female	1.9	Glomus tumor [†]	Pulmonary
Rössle et al.	2006	64	Male	3.5	Glomangioma	Pulmonary
Katabami et al. [3]	2006	56	Female	5.5	Glomangiomyoma	Pulmonary/bronchogenic
Takahashi et al. [8]	2006	67	Male	1.5	Glomus tumor proper type	Endobronchial
Present case	2008	21	Female	2.5	Glomus tumor proper type	Pulmonary

[†]No further specifications available, most likely glomus tumor proper type. [‡]No pulmonary glomus tumor. n/a, data not available.

absence of sustentacular cells and the typical ‘Zellballen’ pattern, combined with the negative staining for neuroendocrine markers [7]. Other neoplasms, such as smooth muscle tumors and secondary metastatic lesions have distinctive histological and immunohistochemical features and were effortlessly differentiated from glomus tumors.

Since glomus tumors are essentially benign lesions, surgical resection is the treatment of choice [1,2,5,8]. However, in a case where the tumor is strictly confined to the bronchial wall and lumen, non-surgical intervention by means of bronchoscopic techniques such as electrocoagulation, forceps extraction and cryotherapy may be indicated [9]. According to the size and localization of the tumor, different surgical techniques can be employed. Wherever possible, a sublobar resection by means of a wedge, anatomical segmentectomy or sleeve resection is the preferred therapy [8,10]. However, in case of central localization where the intersegmental plane cannot be developed and in the absence of significant contraindications, lobectomy may be necessary as in our case to obtain free margins around the lesion [10]. In all cases, prognosis is excellent when a complete resection has been performed [5,8,10].

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