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Primary Pulmonary Glomus Tumor, Diagnosed by Preoperative Needle Biopsy: Report of One Case and Literature Review

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Glomus tumors commonly occur in dermal and subcutaneous tissue in the subungal region of a finger. Some glomus tumors occur extracutaneously, including lung. Only 15 cases of primary pulmonary glomus tumor have been described in literature. In this report, we describe a case of primary pulmonary glomus tumor, which is the first case diagnosed before surgical resection. A 51-year-old man underwent a needle biopsy of a well defined coin-like mass in left lower lobe of the lung on chest radiography. Microscopic examination revealed a tumor composed of perivascularly arranged round to ovoid epithelioid cells with abundant eosinophilic cytoplasm. Tumor cells are immunoreactive for smooth muscle actin and vimentin, but negative for desmin, cytokeratin (AE1/AE3), chromogranin, or synaptophysin. A diagnosis of glomus tumor was then made. The lung mass was resected by wedge resection after being diagnosed by preoperative lung needle biopsy. Although primary pulmonary glomus tumor is rare, most cases follows a benign course. For proper treatment of the patient, glomus tumor should be considered as a differential diagnosis of solitary lung mass.

Key Words : Glomus tumor; Lung; Coin lesion; Pulmonary

Glomus tumors, also known as glomangiomas, are perivascular neoplasms that originate from a neuromyoarterial glomus or glomus body.[1] They are located at arteriovenous anastomosis sites, which functionally provide tissue with a blood supply and regulate temperature. The most commonly affected sites are the dermis and subcutaneous tissue, especially in the subungal region of the finger. However, some glomus tumors occur extracutaneously, e.g., in the gastrointestinal tract,[2] bone, cervix, ovary,[3] trachea,[4] or lung,[5-15] and in such cases the glomus body is devoid. Only 15 cases of primary pulmonary glomus tumor have been described in literature, and these include two malignant glomus tumors and one atypical glomus tumor.[5-15] Here, we report on a case of primary pulmonary glomus tumor, which is the first case diagnosed before surgical resection in the literature.

CASE REPORT

A 51-year-old man visited a local clinic with a cough, sputum, fever, and chill. Routine chest radiography revealed an abnormal nodule in the left lower lobe of the lung (Fig. 1A). He was then transferred to our hospital for further evaluation of the lung nodule. He had a long smoking history of one pack of cigarettes per day for thirty years. Chest computed tomography (CT) revealed a well defined small round nodule in the left lower lobe periphery (Fig. 1B). No other pulmonary lesion or lymphadenopathy was detected. Both lung bronchi were unremarkable by bronchoscopy. Ultrasonography guided percutaneous needle biopsy was performed. Microscopically, perivascular arranged round to ovoid epithelioid cells with abundant eosinophilic cytoplasm were observed (Fig. 2). Immunohistochemistry showed that tumor cells were positive for smooth muscle actin (SMA) and vimentin, but negative for desmin, cytokeratin (AE1/AE3), chromogranin, and synaptophysin (Fig. 3). The needle biopsy findings were interpreted as glomus tumor and then lung wedge resection was performed. Grossly, a 1.3×1.2 cm sized, pink, well demarcated round mass was identified (Fig. 4). Microscopically, the mass consisted of solid proliferative cells with round nuclei and eosinophilic cytoplasm. Tumor cells often showed an oval to spindle shaped nuclear appearance. These cells surrounded normal endothelial lined blood vessels. Immunohistochemical staining findings concurred with those of the needle biopsy.

DISCUSSION

Glomus tumors are infrequently encountered, and accounted for only 1.6% of 500 consecutive soft tissue tumors in one series.[16] Usually glomus tumors occur in the dermis and epidermis of the subungal region, but they can occur at other extracutaneous areas. To our knowledge, primary pulmonary glomus tumor has been reported on 16 occasions (including the present case), as summarized in Table 1 (recently a case was reported in Portuguese,[15] so which was reviewed by abstract, only). The mean age of primary pulmonary glomus tumor patients is 47.8 years (range between 20 and 69 years) and the male-to-female ratio is 2.2:1 (11:5). Approximately a half of these patients experienced no specific symptoms. Most masses were incidentally visualized by chestradiography, as well circumscribed (coin-like)

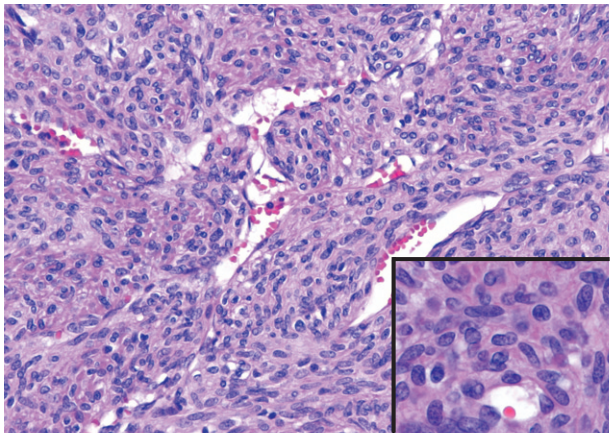

Fig. 2. Perivascularly arranged ovoid epithelioid cells with abundant eosinophilic cytoplasm were observed (× 200, H&E stain).

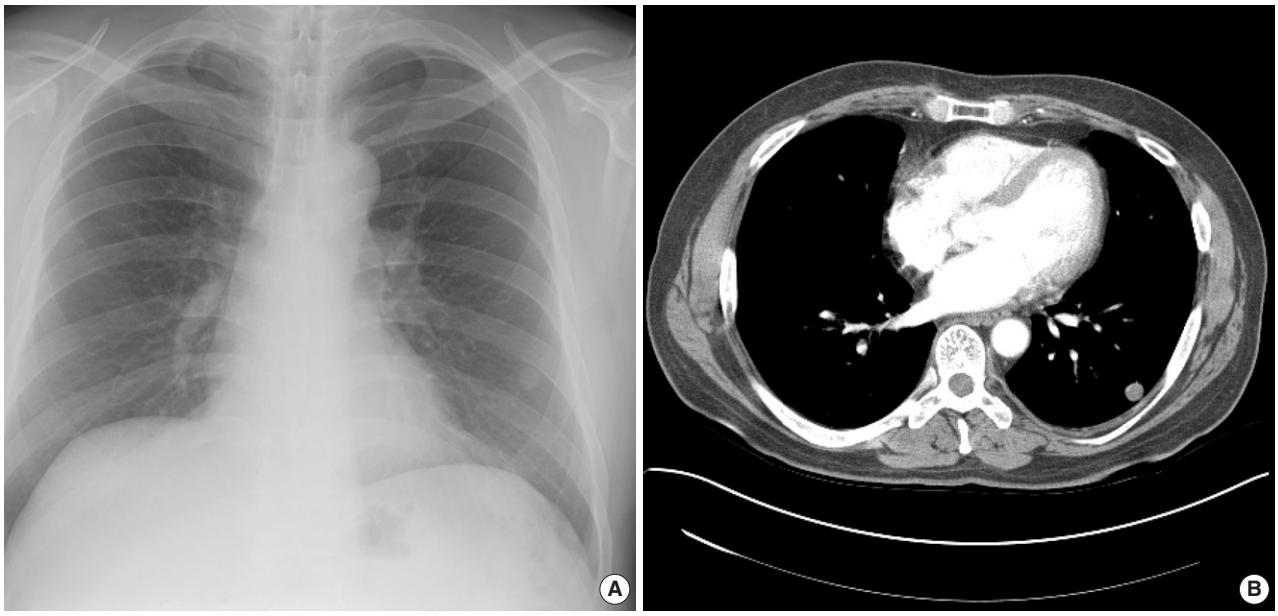

Fig. 1. Coin-like nodule with a sharp outline was revealed in the left lower lobe by chest radiography (A), and chest computed tomography (CT) visualized a well defined small round nodule in left lower periphery (B).

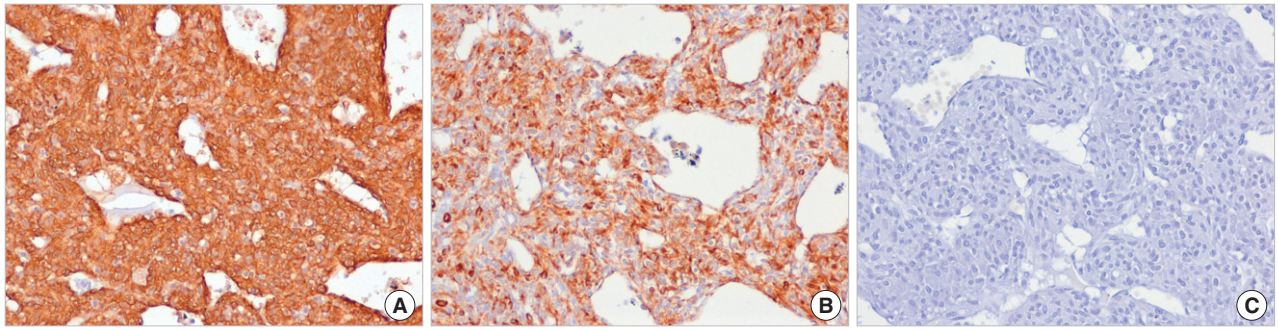

Fig. 3. Tumor cells were strongly positive for smooth muscle actin (A) and vimentin (B), but negative for chromogranin (C) (× 200, immunohistochemical stain).

Table 1. Reported cases of primary pulmonary glomus tumors

Case no. (reference)	Age/sex	Symptom	Size (cm)	Location	Preoperative study	Preoperative diagnosis	Treatment	Final diagnosis	SMA	vimentin	desmin	CK	CG
1 (5)	67/F	RUQ, epigastric pain	6.5×5	LLL			Lobectomy	GT					
2 (6)	34/M	ASX, CXR abn	2.0	RUL	Needle biopsy	Nondiagnostic	Shelled out	GT					
3 (7)	50/M	ASX, CXR abn	1.1	RL			Wedage resection	GT	+	+	+	−	−
4 (7)	41/M	ASX, CXR abn	1.5	RLL			Wedage resection	GT	+	+	−		
5 (8)	20/M	Pneumothorax	1.4×1.3	LMB	BS biopsy	Carcinoid tumor	Sleeve resection	GT	+	+	+	−	−
6 (8)	65/F	ASX, CXR abn	3.0	RL, infrahilar			Wedage resection	GT	+		−	−	−
7 (8)	40/M	ASX, CXR abn	1.1 and 0.5	RLL	Needle biopsy	Suspicious for hemangiopericytoma	Lobectomy	GT	+	+	−	−	−
8 (8)	69/M	Hemoptysis	9.5×9.0	RUL, invl azygos vein			Lobectomy	Gloman-giosarcoma	+	+	−	−	−
9 (9)	29/F	Cough, dyspnea, chest pain	1.5×1.0	LMB	BS biopsy	Carcinoid tumor	Bronchostomy and mass extripation	GT		+		−	−
10 (10)	41/F	ASX, CXR abn	1.0	RLL	Needle biopsy	Negative for tumor	Wedage resection	GT		+		−	−
11 (11)	29/M	Right chest discomfort	2.5×2.0	RH, RMB	BS biopsy	Bronchial adenoma	Sleeve lobectomy	GT with atypical feature	+	+	+, f	−	−
12 (12)	53/M	Dry cough	2.5×2.0	RLL	BS biopsy	Not enough for diagnosis	Lobectomy	Gloman-giosarcoma	+, f	+		−	−
13 (13)	50/M	ASX, CXR abn	4.0	RUL	BS cytology exam	Not be made	Lobectomy	GT	+	+	−		
14 (14)	64/M	ASX, CXR abn	3.5	LLL			NA	GT	+			−	
15 (15)	62/F	Chest pain, dyspnea	1.9	LLL			NA	GT					
Present case	51/M	Cough, sputum	1.3×1.2	LLL	Needle biopsy	GT	Wedage resection	GT	+	+	−	−	−

SMA, smooth muscle actin; CK, cytokeratin; CG, chromogranin; RUQ, right upper quadrant of the abdomen; ASX, asymptomatic CXR; abn, chest radiography (X-ray) abnormality; LLL, left lower lung; RUL, right upper lung; RL, right lung; RLL, right lower lung; LMB, left main bronchus; invol, involving; RH, right hilum; RMB, right main bronchus; BS, bronchoscopic; NA, not available; GT, glomus tumor; f, foccal.

round masses. An average tumor size is 2.6 cm (range between 0.5 and 9.5 cm), and in terms of immuohistochemical profiles, glomus tumors show positivity for smooth muscle actin (SMA), and vimentin, and negativity for cytokeratin, and chromograin. Importantly, the results for these markers have been 100% positive or negative in all performed cases. Desmin positivity has been reported in 3 of 9 cases.

In most cases, tumors have been benign, though rarely they demonstrate an aggressive and malignant clinical course and histological characteristics such as nuclear atypia, necrosis and mitotic activity.[12,17] Ultrastructurally, glomocytes have smooth muscle features, and histologically, are subdivided into three general categories based on the prominence of glomocytes, vessels, and smooth muscle: 1) the common glomus tumor type (glomus tumor proper), 2) a glomangioma type, and 3) a glomangiomyoma type.[5,17] The major feature of the differential diagnosis of glomus tumor concerns carcinoid tumors.[1,3,7] However, in glomus tumors, cells have a fine chromatin pattern, which in contrasts with the coarse granular or salt and pepper chromatin seen in carcinoid tumors.[11] Moreover, the typical organoid arrangement of the tumor cells observed in carcinoid tumors is not prominent in glomus tumors,[8] and glomus tumors are positive for smooth muscle actin, vimentin, and variably positive for desmin, but negative for cytokeratin and neuroendocrine markers, which contrasts with the positivity shown by carcinoid tumors. Hemangiopericytoma shares histologic features with glomus tumors,[18]

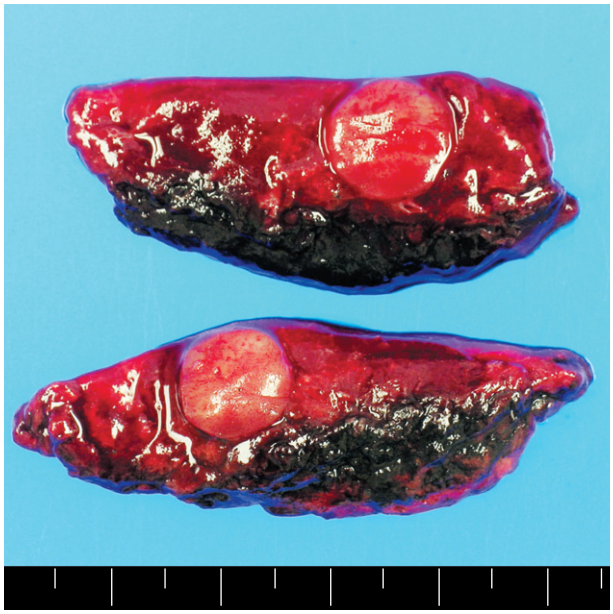

Fig. 4. Grossly, the tumor was a well demarcated, pinkish round mass, measuring 1.3 × 1.2 cm in size.

and glomus tumors may have a focal hemangioperipericytoma like vascular pattern.[8] However hemangiopericytomas are invariably composed of spindle cells with elongated nuclei, whereas glomus tumors are composed of epithelioid cells with round central nuclei.[19,20] In addition, hemangiopericytomas are negative for smooth muscle actin. Other aspect of differential diagnoses are extraskeletal Ewing's sarcoma/primitive neuroectodermal tumor (PNET), and smooth muscle neoplasms (including epithelioid leiomyosarcoma, paraganglioma and metastases).[2,4,6,8]

Primary glomus tumor of the lung is rare entity and most cases follow a benign course and show typical histologic findings and immunohistochemical results. Therefore, the diagnosis of glomus tumor, especially by preoperative diagnostic needle biopsy, is importantly required to avoid unnecessary treatment.

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