Case Report

Malignant glomus tumor in pleural cavity

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Abstract: Glomus tumors, an uncommon hypervascular tumor, arise from modified smooth muscle cells of the glomus body that plays a significant role in the regulation of skin circulation. The tumors are usually located in the extremities, typically in the subungual region of the fingers. Primary glomus tumors of the chest are extremely rare, and to our knowledge, there are no cases have been described in thoracic cavity to date. We here report a case of intrathoracic glomus tumor in a 31-year-old man who presented with a persistent chest pain. Chest computed tomography scans demonstrated an irregularly shaped mass in the left thorax. Left thoracotomy was performed under the suspicious diagnosis of unexplained thorax tumor, and a tumor located in the left upper portion of thorax was founded. Complete resection of tumor along with the partial structure of chest wall was performed. Postoperative diagnosis was malignant glomus tumor.

Keywords: Glomus tumor; malignant; pleural cavity; surgery

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Introduction

Glomus tumor, first described in 1924 by Masson (1), arises from the neuro-arterial structure known as the glomus body. This tumor most frequently occurs in the deep dermis of the extremities and in the subungual region of the hands (2), but it is rarely seen in the pleural cavity. To our knowledge, there are no cases having been reported in pleural cavity to date. This study aims to present a primary malignant glomus tumor of thorax.

Case report

A 31-year-old man presented with intermittent chest pain was admitted to our hospital. He denied fever, cough, expectoration, and chest distress. Clinical history and family history were uneventful. The physical examination was normal. The chest X-ray displayed an abnormal lung shadow in the left upper field (*Figure 1*). Chest computed tomography (CT) scan demonstrated an 8 cm × 7 cm mass with uneven density in the upper left chest cavity (*Figure 2*). Fibrobronchoscopy revealed stenosis of the left upper lobe bronchus under external pressure. It was difficult to determine whether this large complex mass originated from the lung parenchyma or chest wall based on the CT scan appearance. Thus, he received transthoracic thin-needle aspiration biopsy before operation. However, only small amounts of degenerated atypical cells were obtained, which were insufficient for a preoperative histological diagnosis. Right posterolateral thoracotomy was performed in the fourth intercostal space. In surgery, an encapsulated tumor with rich blood vessels was found. This tumor looked to be derived from the parietal pleura, and slightly adhered with the upper left lung. After cutting off the adhesion, the tumor accompanying partial parietal pleura and intercostal muscles of the chest wall was completely resected.

Macroscopically, the circumscribed soft mass measured 8 cm × 7 cm, with white to pink cutting surface, focal hemorrhage, but no necrotic changes. Histologically, the tumor was encapsulated and composed of proliferations of small round to oval cells with minimal variation in size and shape; these cells showed some concentricity around the blood vessels (*Figure 3A*). The tumor cell had a centrally located nucleolus, clear cytoplasm, and distinct cell borders.

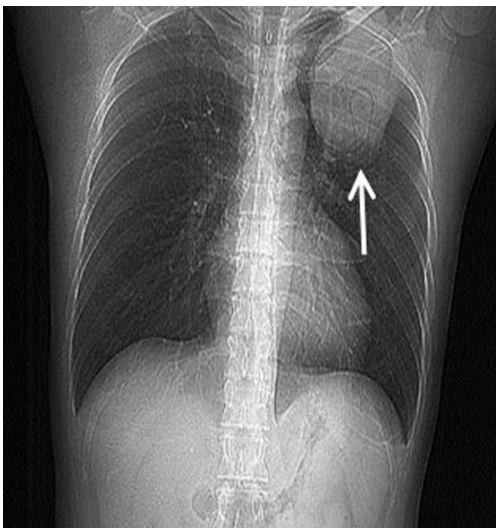

Figure 1 A chest X-ray displayed an abnormal lung shadow in the left upper field (arrow).

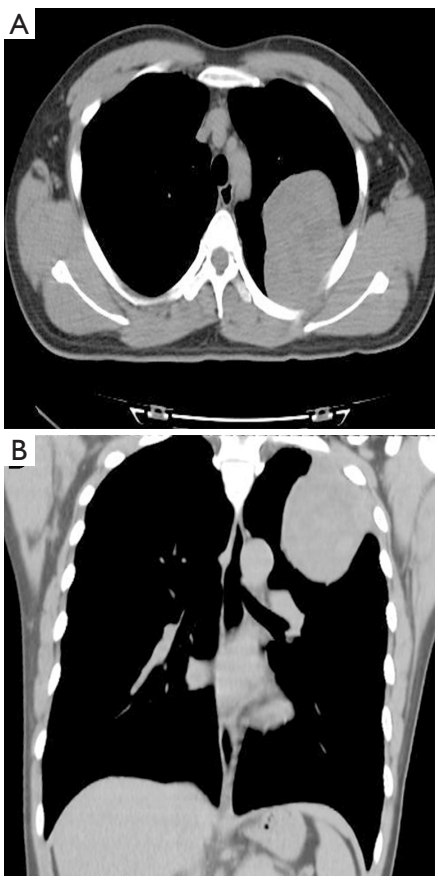

Figure 2 Chest CT scan demonstrated an 8 cm × 7 cm mass with uneven density in the upper of left chest cavity. CT, computed tomography.

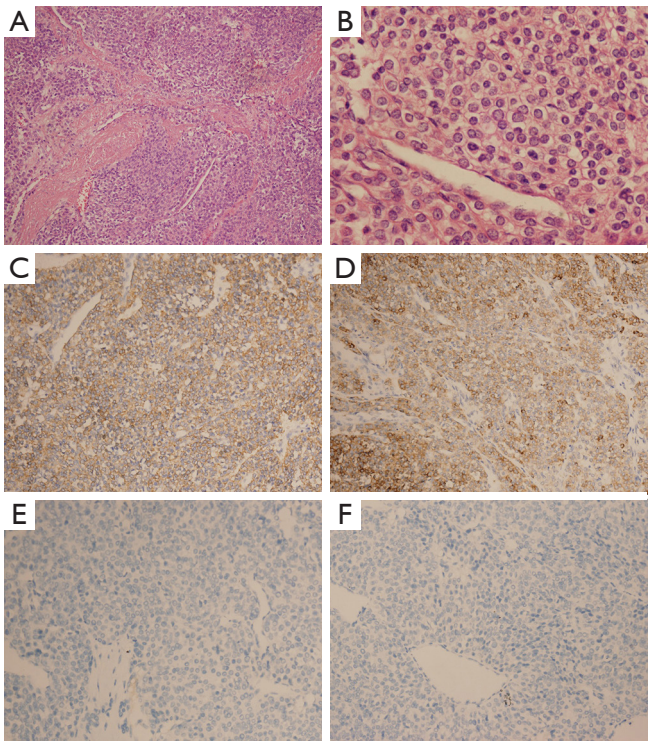

Figure 3 (A) The tumor was composed of uniform small round to oval cells that showed some concentricity around blood vessels; (B) the tumor cell have a centrally located nucleolus, clear cytoplasm and cell borders. (H&E staining, A: 100×; C: 400×); (C,D) immunohistochemical stains showed positivity for SMA and vimentin (200×); (E,F) immunohistochemical stains showed negative for S-100, CD31 (200×).

No nuclear atypical and mitotic figures were observed (*Figure 3B*). Immunostaining demonstrated that the tumor cells were positive for alpha smooth muscle actin (SMA) and vimentin (*Figure 3C,D*), while negative for S-100, CD31, and cytokeratin (*Figure 3E,F*). Based on these findings, we made a final diagnosis of malignant glomus tumor.

Discussion

Glomus tumors are rare neoplasms arising from neuromyoarterial glomus bodies, which are arteriovenous connections that contribute to the management of blood flow and temperature on the surface of the skin (3). Glomus tumors account for 1.6% of all soft tissue tumors (4), which are usually located in the upper extremities, mostly in the subungual region of the fingers. These tumors occasionally occur in internal organs, such as stomach, liver, kidney, and

Table 1 Reported cases of glomus tumor in chest

N	Author	Age (year)	Sex	Size (cm)	Location	Symptom	Treatment	Follow-up
1	Tang, 1978 (12)	67	F	6.5×5.0×5.0	Pulmonary	Bellyache	Lobectomy	No recurrence
2	Fabich,1980 (13)	63	M	2.5×2.0×1.0	Tracheal	Cough	Sleeve resection of trachea	No stated
3	Shin, 1990 (14)	47	F	1.5×1.0×1.0	Tracheal	Hemoptysis	Sleeve resection of trachea	No recurrence
4	Hirose, 1996 (15)	26	F	1.4×2.5×3.0	Mediastinum	Back pain	Complete resection	No recurrence
5	Koskinen, 1998 (16)	66	M	2.0×3.0	Tracheal	Hypertension	Radiotherapy	No recurrence
6	Gowan, 2001 (17)	73	M	1.6×1.3×0.6	Tracheal	Cough, Dyspnea	Segmental resection of trachea	No recurrence
7	Hishida, 2003 (18)	53	M	2.5×2.0×1.8	Pulmonary	Cough	Lobectomy	No recurrence
9	Kleontas, 2010 (19)	74	M	4.0×2.6	Pulmonary	Cough	Lobectomy	No recurrence
10	Santambrogio, 2011 (20)	39	M	1.3×1.1	Pulmonary	Nothing	Wedge resection of lung	No recurrence
11	Hohenforst-Schmidt, 2012 (21)	35	F	No stated	Pulmonary	Chest pain	Pneumonectomy	No stated
12	Huang, 2013 (22)	28	M	8.0×6.0×6.0	Pulmonary	Hyperpyrexia	Lobectomy	No recurrence
13	Zhu, 2013 (23)	30	F	4.0×0.5×0.5	Endobronchial	Polypnea	Complete resection	No recurrence
14	Fan, 2013 (24)	15	M	2.3×2.0	Tracheal	Hemoptysis	Sleeve resection of trachea	No recurrence
15	Rychlik, 2014 (25)	59	M	2.0×2.0×2.0	Mediastinum	Chest pain	Complete resection	No recurrence
16	Elkrinawi, 2012 (26)	61	F	4.0×5.0	Pericardium	Dyspnea	Complete resection	Recurrence
17	Bali, 2013 (27)	49	F	7.6×4.9×4.2	Esophagus	Dysphagia	Esophagectomy	No recurrence
18	Ghigna, 2013 (28)	70	M	2.0×2.0	Tracheal	Hemoptysis	Segmental resection of trachea	No recurrence
		40	M	1.4×1.0	Tracheal	Hemoptysis	Segmental resection of trachea	No recurrence

adrenal glands (5-8). Review of the published literature revealed rare involvement of the pleural cavity. Our case is the first report of a large intrathoracic glomus tumor.

According to the relative prominence of glomus cells, vascular structures, and smooth muscle, glomus tumors are subdivided into solid glomus tumor, glomangioma, and glomangiomyoma (9,10). Although glomus tumors are usually benign, malignant transformation has been reported in the literature (10,11). Folpe *et al.* (11) presented the following diagnostic criteria for malignant glomus tumors of soft tissue: (I) deep location and size of more than 2 cm; (II) presence of atypical mitotic figure; or (III) combination of moderate to high nuclear grade and mitotic activity (5 mitoses/50 high-power fields). Based on the aforementioned criteria, our patient was diagnosed as malignant glomus tumor.

Pathologically, solid glomus tumors consist of small uniform rounded glomus cells that have small uniform nuclei, clear cytoplasm, and distinct cell borders, without nuclear atypia or mitotic figures (11). The microscopic characteristics were similar to some neuroendocrine tumor, such as carcinoid. Therefore, immunohistochemistry is essential in the differential diagnosis of glomus tumors with some neuroendocrine tumor. In general, immunohistochemical staining of glomus tumors for SMA and vimentin were positive. However, they were negative for neuron specific enolase (NSE), synaptophysin (Syn), S-100, and chromogranin. In contrast, immunohistochemical stainings of neuroendocrine tumor for those markers of nervous tissue were exclusively positive.

Glomus tumor occurring in the chest is extremely rare. The literature review (*Table 1*) revealed that the most common sites in the chest are lungs, trachea, and mediastinum (2,12-25). Rare cases occurring in the esophagus and pericardium have also been reported (26,27). To date, there was no report about this condition occurring in the pleural cavity. The clinical presentation of intrathoracic glomus tumor is nonspecific and varies depending on the site and size of the tumor, which is similar to other common thoracic tumors. Patients may present chest pain, cough, and breathlessness. In this patient, the main symptom was chest pain because of the involvement of the chest wall. Radiographic findings are also nonspecific. X-ray or CT scan usually reveals a circumscribed soft tissue mass without calcification. The

rarity of intrathoracic glomus tumors causes confusion with other similar tumors, such as mesotheliom, hemangiopericytomas, and hemangioendotheliomas. Transthoracic thin-needle aspiration biopsy may occasionally contribute to preoperative diagnosis. However, limited tissue samples reduce the success rate of diagnosis.

Surgical resection is the first choice of diagnosis and treatment for thoracic glomus tumor. In light of the tendency toward malignancy, radical resection including the surrounding normal tissue should be performed. Edges of the tumor must be determined to ensure negative margin resection during operation. In our case, given that the tumor was closely associated with the chest wall, a complete resection of the tumor along with the partial parietal pleura and intercostal muscles of the chest wall was performed. Although the morbidity and recurrence rates appear to be low in most reported cases, close follow-up is required.

Conclusions

Primary glomus tumors in chest are extremely rare, which usually occur in the lung or trachea. There are no cases in the pleural cavity having been described in literature up to now. Preoperative diagnosis of glomus tumor in pleural cavity is difficult, since their clinic symptom and radiology features overlap with more common thoracic tumors. Here we reported a very rare case of a primary malignant glomus tumor occurring in the pleural cavity. Due to the lack of the evidence in the current literature, the therapeutic effect cannot be assessed exactly and close follow-up is necessary.

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