Leiomyosarcoma of Esophagus Presenting a Huge Posterior Mediastinal Mass: A Case Report

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Smooth muscle tumors presenting as mediastinal soft tissue masses are rare and are often mistaken for other neoplasms. In this context, we presented a case of leiomyosarcoma of the esophagus presenting as a huge posterior mediastinal mass. The computed tomographic findings were discussed.

*Key words: Esophagus; Leiomyosarcoma; Posterior mediastinal tumor*

Leiomyosarcoma is a high-graded, smooth muscle tumor of mesenchymal origin that can arise in the gastrointestinal tract. In esophagus, it is rare, only accounting for less than 1% of malignant tumors. Diagnosis is based on pathologic and immunohistochemical findings. Because of its location in the submucosal layer, it usually grows to a huge mass by the time symptoms appears [1]. The computed tomographic (CT) appearance is not specific and can be mistaken for other tumors. It is usually large in size, showing low attenuation areas of central necrosis. Calcification is rather uncommon [2].

We report a case of an esophageal leiomyosarcoma presenting as a huge posterior mediastinal mass.

**CASE REPORT**

A 36-year-old Chinese woman suffered from chest pain for 10 days. She denied having fever, cough or loss of weight. She had a chest radiograph in a clinic (Fig. 1) which showed a huge soft tissue mass in right lower lung field with blunting of right cardiophrenic angle. Patient was then referred to our institution. Ultrasound-guided thoracentesis was performed but the result was unremarkable. Bronchoscopy and bronchial washing in the lateralobasal and posterobasal segments of the right lower lobe did not reveal malignant cells. Computed tomography (CT) (Fig. 2a) showed a huge mediastinal mass (111mm) at right cardiophrenic space adjacent to the diaphragm and esophagus (Fig. 2c). No calcification was seen. On post IV contrast scanning (Fig. 2b), there was heterogenous enhancement of the solid component in the medial aspect and no enhancement at the lateral portion suggestive of necrosis. The patient was advised to undergo excision of the mediastinal mass.

Surgical findings revealed a huge well encapsulated, hypervascular mass about 12 × 10 cm. The lesion was adherent to right lower lung with pedicle...
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arising from distal third of the esophagus. No pleural seeding was noted. About 200 ml of clear yellowish fluid was evacuated. Pathologic and immunohistochemical analysis revealed a 12 × 10 cm encapsulated, soft-tissue mass, light-yellow to gray in color with hemorrhage weighing 1500 grams (Fig. 3). Microscopic analysis revealed a well-defined tumor composed of fascicles of spindle cells with elongated nuclei and pink cytoplasm. Focal hemorrhage and several foci of coagulation necrosis accompanied by acute inflammation were also noted. Mitotic number count was 1/50 HPF (high power field) but immunohistochemical study for ki-67 showed 20-25% mitotic index (Fig. 4). On immunohistologic analysis, tumor cells were positive for desmin (muscle marker) and negative for CD117, CD34 and S-100. Considering the tumor size and overall features, it was diagnosed to be a leiomyosarcoma.

DISCUSSION

Leyomyosarcoma is a rare tumor of mesenchymal origin. It is a high-graded soft tissue tumor that can arise in any tissue containing smooth muscle fibers [1]. Forty percent of leiomyosarcoma are found in the alimentary tract mostly in stomach and small bowel; few have been found in colon and esophagus [2, 3], 24% in genitourinary tract, 19% in retroperitoneum and 14% in other soft tissues [2]. It apparently arises between muscularis propria and muscularis mucosa layers of the bowel wall but the exact etiology remains in question [3]. As a result, the tumor tends to grow submucosally and extraluminally, leading to late obstruction. Tumors are usu-

Figure 1. Chest x-ray suggested a huge soft tissue mass in right lower lung field with blunting of right cardiophrenic angle (arrow).

Figure 2. a. Pre-contrast CT scan axial view showed a huge (111 mm) mediastinal mass (arrow) at right cardiophrenic space. b. Contrast enhanced CT scan axial view showed the mediastinal mass at right cardiophrenic space with heterogeneous enhancement of the solid component in the medial aspect (arrow) and unenhanced lateral portion suggestive of necrosis (arrowhead). c. Contrast enhanced CT scan coronal view showed the huge mediastinal mass (arrow) abutting the esophagus (curve black arrow) and diaphragm (straight black arrow).
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ally rather large when it is initially presented [3, 4].

Esophageal leiomyosarcoma accounts for 0.5% of all esophageal malignancy and 5% of gastrointestinal sarcomas [5]. It is usually located in the middle or distal third of the esophagus [6] and is usually found in middle-aged or elderly patients with slight male predominance [1, 7]. The most common symptom is dysphagia [7, 8], some with nausea, vomiting, weight loss or bleeding [3]. Growth pattern are classified as infiltrating or polypoid [1, 8].

Radiographic appearance of leiomyosarcoma especially on CT scan is non-specific. It usually presents as a large mass with low attenuation areas of central necrosis or cystic change and myxoid degeneration [2, 9], some with extraluminal gas or contrast material within the tumor [9]. There is moderate peripheral rim enhancement of the large primary tumor. Calcification in the tumor is rather uncommon. [2]. Our patient presents with the same characteristics of large posterior mediastinal mass with necrosis on its lateral portion and an enhanced solid part on its medial aspect. No calcification was seen. Similar findings have been reported with leiomyosarcoma of stomach and small bowels, indicating that these tumors have identical gross and pathologic features regardless of their location [9, 10, 11].

Leiomyosarcoma cannot be differentiated from leiomyoma and other large tumor merely by radiographic features [6]. Levine et al. have mentioned that large intramural lesion with ulceration and tracking should suggest the diagnosis but accurate diagnosis still depends on histopathologic evaluation which includes mitoses, cellularity, nuclear pleomorphism and tumor necrosis [12]. The lack of relationship in a nerve cell with strong immunoreactivity for smooth muscle actin and desmin and lack of staining with S-100 protein argued in favor of smooth muscle over peripheral nerve sheath differentiation [13].

CONCLUSION

By anatomic divisions, posterior mediastinum includes descending aorta, ayzygous vein, superior intercostals vein, thoracic duct, esophagus and vagus nerve. Several tumors can grow without symptoms and varies from benign to malignant. Most common posterior mediastinal tumors are neurogenic neoplasm either benign or malignant, lymphoma and mediastinal cysts but sarcoma group like leiomyosarcoma should be included in the differential diagnosis, especially in patient who presents with huge mediastinal mass that has very minimal or even without symptoms.

The CT features of leiomyosarcoma is non-specific. It usually appears as a large mass with frequent areas of cystic and myxoid degeneration; while, calcification is rather uncommon. These features can be easily confused with neural or other neoplasm. Therefore, imaging modalities are not enough for definitive diagnosis and usually requires definite histopathologic analysis.

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Figure 3. Gross appearance of the excised huge mediastinal mass.

Figure 4. Ki-67 stain showed mitotic index of 20-25% (arrowhead).
食道平滑肌肉瘤以巨大後縱隔腔腫瘤表現：
病例報告及文獻回顧

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平滑肌肉瘤發生在胸腔是非常少見，文獻報導常發生於縱隔腔，而以巨大後縱隔腔腫瘤表現則是非常罕見，我們報告一個後縱隔腔巨大食道平滑肌肉瘤的電腦斷層影像表現。本文中的病例我們將敘述在電腦斷層影像中的發現。

關鍵詞：食道；平滑肌肉瘤；後縱隔腔