Radiologic features of posterior mediastinal leiomyosarcoma presenting as a mimic of peripheral nerve sheath tumour

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Radiologic features of posterior mediastinal leiomyosarcoma presenting as a mimic of peripheral nerve sheath tumour

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ABSTRACT:

Leiomyosarcoma is one of the most common subtypes of malignant mesenchymal neoplasms, or in other words, it is around 20% of all new cases of soft tissue sarcomas. Though LMS occurs more frequently in the abdomen, retroperitoneum, large blood vessels, and uterus, it is exceptionally rare to meet it in the mediastinum, with less than 10% documented cases in the literature. A 62-year-old male patient having no history of significant illness has been brought with a six-month history of progressively worsening back pain. Magnetic resonance imaging done in view of clinical presentation revealed the imaging features of posterior mediastinal peripheral nerve sheath tumor. However, the biopsy of the tumor revealed positive immunohistochemical markers for leiomyosarcoma. Due to their wide variability in presentation and considerable risk for metastasis, leiomyosarcomas pose challenges in diagnosis and treatment. Advanced imaging, as well as a sophisticated appreciation of their pathological and genetic features, is therefore key to proper management and an evaluation of prognosis.

Keywords: posterior mediastinum, leiomyosarcoma, peripheral nerve sheath tumor, smooth muscle markers, paravertebral region

INTRODUCTION

Leiomyosarcoma is a rare malignant tumor originating from smooth muscle cells, typically occurring in the retroperitoneum, uterus, and large blood vessels [1]. Its posterior mediastinal location is extremely uncommon, thus posing difficulty in diagnosis and management because of its proximity to vital structures such as the descending aorta, esophagus, and nerve roots. Symptoms of mediastinal tumor's vary, ranging from being asymptomatic to causing significant compression of adjacent structures [2]. Due to their rarity and the confined nature of the posterior mediastinum, LMS can mimic more common masses such as neurogenic tumor's. Magnetic resonance imaging (MRI), play a crucial role in accurately assessing these tumors and their relationship with surrounding anatomy [3].

CASE PRESENTATION

This case involves a 62-year-old male with no remarkable past medical history, presenting with a six-month history of progressively worsening back pain radiating to the left axillary region. During the physical examination, significant tenderness was observed over the thoracic spinal and paraspinal areas. Laboratory tests showed elevated C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). Due to his worsening clinical symptoms, further diagnostic evaluation and imaging was done.

Imaging features

Radiograph dorsal spine shows focal soft tissue opacity in the mediastinum causing thickening of left paravertebral stripe in AP radiograph, as prevertebral opacity on lateral radiograph. The cardiac contour and hilar vessels can be seen through its inferior border, placing it in the posterior mediastinum ***Figure 1(a, b)***.

Magnetic resonance imaging of spine shows A well-defined, round to oval lesion is observed in the left paravertebral region, extending from the superior end plate of the D6 vertebra to the superior end plate of the D8 vertebra. The lesion measures 4.7 x 4.6 x 5 cm (AP x TR x CC) with smooth margins. The lesion exhibits the following imaging characteristics: on T1-weighted MRI, it appears hypointense with peripheral focal nodular hyperintensity; on T2-weighted MRI, it is heterogeneously iso- to hyperintense; and on the STIR sequence, it is hyperintense. The lesion abuts the abdominal aorta anteromedially without evidence of luminal narrowing. Medially and postero-medially, it contacts the D6 and D7 vertebrae without causing destructive or erosive changes and shows no extension into the neural foramina or spinal canal. Laterally, the lesion contacts the medial basal segment of the left lower lung lobe, with adjacent mild passive atelectatic changes. Inferiorly, it is connected to the left D7 exiting nerve roots. Additional findings include minimal heterogeneous areas of diffusion restriction, with post-contrast imaging revealing intense, heterogeneous, avid enhancement and focal nodular areas of non-enhancement. Based on these imaging findings, the differential diagnosis includes a neurogenic tumor, such as a schwannoma or neurofibroma, with a less likely consideration of a paravertebral paraganglioma. The mass does not appear to invade the bronchus, descending thoracic aorta, esophagus, vertebrae, or ribs ***Figure 2(a, b), Figure 3(a, b, c, d), Figure 4 (a, b, c, d).***

However, the Biopsy Results showed that the tumor cells arranged in intersecting fascicles. The cells are mildly to moderately pleomorphic, with elongated spindled nuclei and eosinophilic cytoplasm. Scattered highly pleomorphic cells are noted. Mitotic activity is low, at 1-2 per 10 high-power fields (HPF). No necrosis is observed. The tumor is positive for immunohistochemical markers SMA (clone IA4) and H-Caldesmon (clone EP19), with features suggestive of leiomyosarcoma.

DISCUSSION

Leiomyosarcoma of the posterior mediastinum is an exceptionally rare tumor, accounting for only a small percentage of primary mediastinal tumors. Diagnosis and treatment are

challenging due to their diverse presentation and the complex anatomy of the mediastinum. The posterior mediastinum, an anatomical space containing vital structures such as nerves, vessels, and the spine, is a common site for various tumor's, especially those of neurogenic origin [4,5]. As neurogenic tumors represent the majority of posterior mediastinal masses, there is a significant potential for the misdiagnosis of leiomyosarcoma as a peripheral nerve sheath tumor. The imaging characteristics of posterior mediastinal leiomyosarcomas are variable, often showing heterogeneous signals on MRI with evidence of tissue invasion [6,7]. In this case, MRI revealed a well-defined lesion with hyperintensity on T2-weighted images and the STIR sequence, as well as peripheral nodular hyperintensity on T1-weighted images, indicative of a complex mass [8].

Differentiating between leiomyosarcoma and peripheral nerve sheath tumors based solely on imaging is difficult, as both can present as well-defined masses with varying degrees of heterogeneity. However, the lack of involvement of adjacent structures in this case, despite the tumor's size, helped narrow down the differential diagnosis in this case [9,10]. Leiomyosarcomas are known for their potential to grow significantly before symptoms occur, largely due to the loose connective tissue of the retroperitoneum and mediastinum.

Histopathological examination remains the gold standard for diagnosis, as imaging alone cannot reliably distinguish leiomyosarcomas from other spindle cell neoplasms, such as malignant peripheral nerve sheath tumor. Immunohistochemical staining plays a crucial role in confirming smooth muscle differentiation, typically showing positivity for smooth muscle markers, including muscle-specific action, desmin, and h-caldesmon. Some subtypes, such as epithelioid leiomyosarcoma, may require additional stains for accurate identification, as they can mimic carcinoma by staining positive for epithelial markers. In such cases, stains like histone deacetylase-8 and myocardin can aid in confirming the diagnosis. Furthermore, immunopositivity for p16 and p53, along with a high Ki-67 proliferation index, has proven effective in distinguishing leiomyosarcomas from leiomyomas [11,12].

In this case, the diagnosis was confirmed by a core needle biopsy, allowing for timely optimization of the patient before surgical intervention. The primary treatment for mediastinal leiomyosarcomas, particularly large posterior mediastinal tumor's, remains surgical resection. The preferred operative approach is a posterolateral thoracotomy, which provides optimal access and offers the best chance for a positive outcome. Smaller lesions may be managed using video-assisted thoracoscopic surgery, though this method offers limited exposure for larger tumor's [13,14].

Preoperative planning is crucial, given the mediastinum's complex anatomy and the tumor's proximity to vital structures such as the aorta, esophagus, airways, and spinal canal. While most large tumor's tend to compress rather than invade adjacent structures, en bloc resection might be necessary if the tumor extends into the lung, chest wall, or spinal area. If diaphragm resection is required, reconstruction can be performed using a polytetrafluoroethylene graft. In rare cases involving extensive spread, a thoracoabdominal approach may be employed. The risk of local recurrence and distant metastasis, even after aggressive surgical management, emphasizes the need for close, long-term follow-up. The role of adjuvant therapies, including chemotherapy and radiotherapy, remains a topic of debate and depends

on the tumor's histopathology. These adjuvant treatments aim to reduce recurrence and metastasis, especially in malignant cases, which require regular monitoring for early detection of recurrence. A multidisciplinary approach, involving radiologists, surgeons, oncologists, and pathologists, is essential for optimizing patient outcomes [15].

CONCLUSION

Posterior mediastinal leiomyosarcoma is an extremely rare entity, which can closely mimic neurogenic tumor's, including peripheral nerve sheath tumor's, on imaging. Overlap between these entities creates a diagnostic dilemma that warrants complete evaluation with advanced imaging techniques along with histopathological confirmation. The successful surgical resection of the leiomyosarcoma without involvement of any adjacent tissue underlines the potential for favorable outcomes when this tumor is identified and treated appropriately. This case brings out the importance of considering leiomyosarcoma in the differential diagnosis of posterior mediastinal masses, guiding clinicians toward the optimal management approach. The main factors in improving the prognosis of this rare tumor are earlier and more exact diagnosis, along with a multidisciplinary therapeutic approach.

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