Case Report
Primary well differentiated liposarcomas in the mediastinum: two cases and a review of the literature

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Abstract: Well differentiated liposarcoma is a malignant mesenchymal tumor which occurs extremely rarely in the mediastinum, comprising less than 1% of all mediastinal tumor cases. The tumor is indistinguishable radiologically from other neoplasms occurring in the mediastinum including lipoma, teratoma and thymoma. Therefore, histological examination is the most important tool in the diagnostic process. Here, we report two cases of well differentiated liposarcoma in the mediastinum of Asian female. Both of the lesions were radiologically diagnosed as benign tumors, but the histological findings, including significant variation in cell size, focal nuclear atypia in adipocytes, and scattered lipoblasts, supported the diagnosis of well differentiated liposarcoma. Upon reviewing the literature, we have summarized the clinical presentation, prognosis, and treatment of liposarcoma in the mediastinum. We found that Computed tomography (CT) scans are the imaging modality of choice for the diagnosis of such lesions, but ultimately, histological examination is required for a definitive diagnosis.

Keywords: Liposarcoma, mediastinum, diagnosis, computed tomography, histological

Introduction

Atypical lipomatous tumor (ALT)/well differentiated (WD) liposarcoma is an intermediate (locally aggressive) malignant mesenchymal neoplasm, typically occurring in the lower extremities and retro peritoneum. And accounting for 10-16% of all sarcomas, it is the most common type of soft-tissue sarcoma [1]. When this tumor occurs in the mediastinum, obtaining a wide margin surgically is nearly impossible, resulting in a high propensity for recurrence, so the diagnosis of “well differentiated liposarcoma” is recommended. Primary liposarcoma in the mediastinum is extremely rare, comprising less than 1% of all mediastinal tumor cases [2]. It shares radiographic features with other mediastinal tumors and can be misdiagnosed as the more commonly occurring lipoma, teratoma or thymoma. The two cases we report were initially diagnosed as benign tumors by the radiologist due to the density of the adipose tissue on the computed tomography (CT) scan and because of the patient’s lack of worrisome clinical features, such as weight loss. Histologically, the tumor cells displayed significant variation in cell size with focal adipocyte nuclear atypia and scattered lipoblasts, suggestive of the diagnosis of well differentiated liposarcoma.

Case presentation

Case one is a 33-year-old female patient who presented to the hospital with dyspnea, post-exercise orthopnea, as well as an occasional cough producing white phlegm. Case two is a 55-year-old Asian female who presented to the hospital with a three year history of dyspnea and three month history of a cough productive of white phlegm. Their visit to the hospital was prompted by worsening of symptoms. No other neurological deficits were identified in both two patients. Both of the two patients had no history of trauma or cranial nerve abnormalities. Both of the two patients declared for no family history of malignant tumor.
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Materials and methods

The patients underwent computed tomography (CT) and magnetic resonance imaging (MRI) examinations. Pathology examination was performed, the resected tissues were embedded in paraffin blocks and sectioned, H&E staining was done according to the standard protocol. This study was prospectively performed and approved by the institutional Ethics Committees of China Medical University and conducted in accordance with the ethical guidelines of the Declaration of Helsinki. Written informed consent was obtained from the patient for the publication and accompanying images.

Result

Case one

The contrast-enhanced CT scan of her thorax revealed a large mediastinal mass near the right lung characterized by a heterogeneous density with local fat shadow but no distinct enhancement following contrast administration (Figure 1A, 1B). Radiologically, the differential diagnosis for the lesion included lipoma, teratoma or thymoma. The tumor was surgically resected and submitted for pathologic evaluation. On gross examination, the lesion measured 10 x 8 x 7 cm, was well-circumscribed, lobulated in shape with no macroscopic evidence of invasion into surrounding tissue. The cut surface of the tumor was white-yellow, firm, and there was no grossly apparent necrosis or hemorrhage. Microscopically, adipocyte-like tumor cells showed significant variation in size and nuclear atypia. Scattered lipoblasts could also be seen among the mesenchymal cells. The histological appearance was most consistent with WD liposarcoma (Figure 2A and 2B). Clinical follow-up based on review of the patient’s electronic medical record showed no recurrence by CT examination at 3 and 6 months after surgical treatment.

Case two

A 55-year-old Asian female presented to the hospital with a three year history of dyspnea and three month history of a cough productive of white phlegm. A contrast enhanced CT scan of the thorax demonstrated a large heterogeneously dense, non-enhancing mass, with a clear boundary, located in the midline of the posterior mediastinum (Figure 1C, 1D). The lesion was surgically resected and gross examination revealed a 15 x 10 x 7 cm well-circumscribed, encapsulated, white-yellow, and firm mass. Microscopically, lipoma-like tumor cells with atypical features including enlarged, irregular nuclear and scalloped/signet ring-like lipoblasts were easily identified. Ultimately, the mass was pathologically classified as liposarcoma.
patients older than 40, with a mean age range of 43-58 years. There is no apparent difference in the incidence rate between genders [6, 7]. Both of our patients were women. One of our patients presented at the age of 33, which is younger than the mean age published in the literature; and the other presented at 55, coinciding with the typical mean age. Presenting symptoms vary depending on the location and size of the lesion. Mediastinal liposarcomas commonly grow to a large size and by exerting a mass effect on surrounding organs, they result in clinical symptoms such as shortness of breath, chest pain, and tachypnea [8], as was seen in our cases. Some patients are asymptomatic and their lesions are detected incidentally on imaging, making the diagnosis more difficult [9]. Radiologic investigation is the primary diagnostic approach, specifically CT scan of the thorax. The detected of a heterogeneous low-density shadow similar to that of fat with clear boundaries and no obvious parenchymal enhancement should raise the possibility of a liposarcoma. The radiographic diagnosis of liposarcoma can be challenging due to its similarities with other lesions, including lipoma, teratoma and thymomas by CT.

Histopathological examination is required for a definitive diagnosis. Grossly, liposarcomas are typically large, well-circumscribed, and lobulated masses. Rarely, an infiltrative growth pattern may be seen. Color varies from yellow to white depending on the proportion of adipocytic fibrous/myxoid components present, respectively. Necrosis is commonly apparent in larger lesions. The tumors in our cases were encapsulated and well-circumscribed with a white-yellow cut surface, no obvious necrosis.

ALT/WD liposarcoma can be subdivided morphologically into four main subtypes: adipocytic (lipoma-like), sclerosing, inflammatory, and patients older than 40, with a mean age range of 43-58 years. There is no apparent difference in the incidence rate between genders [6, 7]. Both of our patients were women. One of our patients presented at the age of 33, which is younger than the mean age published in the literature; and the other presented at 55, coinciding with the typical mean age. Presenting symptoms vary depending on the location and size of the lesion. Mediastinal liposarcomas commonly grow to a large size and by exerting a mass effect on surrounding organs, they result in clinical symptoms such as shortness of breath, chest pain, and tachypnea [8], as was seen in our cases. Some patients are asymptomatic and their lesions are detected incidentally on imaging, making the diagnosis more difficult [9]. Radiologic investigation is the primary diagnostic approach, specifically CT scan of the thorax. The detected of a heterogeneous low-density shadow similar to that of fat with clear boundaries and no obvious parenchymal enhancement should raise the possibility of a liposarcoma. The radiographic diagnosis of liposarcoma can be challenging due to its similarities with other lesions, including lipoma, teratoma and thymomas by CT.

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ALT/WD liposarcoma can be subdivided morphologically into four main subtypes: adipocytic (lipoma-like), sclerosing, inflammatory, and
spindle cell [10-13]. The presence of more than one morphological pattern in the same lesion is common, particularly in retroperitoneal tumors. The two cases we report were of the adipocytic type, characterized by a proliferation of relatively mature adipocytic call, focal nuclear atypia, and scattered lipoblasts. Immunohistochemistry and molecular testing play a very minor role in the diagnosis of ALT/WD liposarcoma because of their classic morphological features. MDM2 amplification studies were not performed on our cases because the characteristic histological appearance did not warrant further testing.

In conclusion, liposarcomas are rare tumors in mediastinum. CT scan is the imaging modality of choice, but distinguishing these lesions from radiographically similar lipomas, teratomas, and thymomas is challenging. Therefore, histological examination is the most reliable approach for obtaining a definitive diagnosis.

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Disclosure of conflict of interest

None.

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References