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Primary Mediastinal Myxoid Liposarcoma

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ABSTRACT

In this study, we report an extremely rare case of liposarcoma which arises primarily in mediastinum. The patient appeared to have progressive dyspnoea and prolonged cough for a duration of one year. Chest radiograph and Computed Tomography (CT) of the thorax revealed a large right mediastinal mass with fatty component. It was confirmed to be primary liposarcoma on histopathological examination.

Keywords: Mediastinal liposarcoma; liposarcoma

INTRODUCTION

Primary mediastinal liposarcomas are extremely rare tumours, which are composed largely of fat. They usually occur in adults, with an average presenting age of 56 years old[1]. These tumours grow to an enormous size, and their clinical symptoms are referable to compression and displacement of adjacent mediastinal structures[1].

Histopathologic examination is always necessary as much for diagnosis as prognosis, and thus, the optimum treatment is surgical removal[2]. Such an approach helps in establishing a tissue diagnosis, relieving patient's symptoms, and may result in a cure.

We described a primary mediastinal myxoid liposarcoma in a 42-year old lady who had complained of progressive dyspnoea and prolonged cough for one year.

CASE REPORT

Madam AA, a non-smoker 42-year old Malay lady, was referred to chest clinic complaining of progressive shortness of breath and chronic productive cough of two weeks' duration. These symptoms had recurred frequently for the past one year, and she had been treated with antibiotics and bronchodilators with transient improvement. Additionally, she reported a weight loss of 10 kilograms in a year and this was associated with her poor appetite. She denied history of fever, chest pain, haemoptysis or contact with pulmonary tuberculosis patients. There was no orthopnoea or paroxysmal nocturnal dyspnoea to suggest cardiac failure.

A physical examination revealed a thin middle aged lady with minimal respiratory distress. Percussion of the chest revealed dullness of the middle and lower zones of the right hemithorax. On auscultation, poor air entry was noted on the right side with end-inspiratory rhonci. The apex beat was displaced to the sixth left intercostal space and two centimetres lateral to the left midclavicular line.

An initial laboratory investigation showed a normal haemoglobin value of 13.7 g/ dl and a leukocyte count of 6600/ microlitre. Work-up for tuberculosis were negative. Urine analysis and blood chemistry results were within normal limits.

Chest radiograph (see Figure 1) revealed a homogenous opacity in the right middle and lower zones obliterating the right costophrenic angle. The mediastinum and trachea were displaced to the contralateral side.

Contrast-enhanced axial CT thorax (Figure 2) demonstrated a large heterogeneous soft tissue mass within the right hemithorax occupying the right lower lobe measuring around 23 × 24 × 15 cm. This mass has multiple septations with heterogeneous lesions of fat, fluid and soft tissue densities. Meanwhile, a focus of calcification was seen in the anterolateral aspect of the mass. The mediastinum was displaced to the contralateral side. Trachea was slightly deviated to the left side. The right upper lobe and the left lung were normal. No pleural effusion was demonstrated. Differential diagnoses of a teratoma, lipoma and liposarcoma were entertained.

A right thoracotomy was performed and a 27 × 15 cm lobulated soft to firm mass was found to be arising from the right mediastinum. The mass was yellowish to whitish in colour and moderately vascular. There was no abnormality noted on the surface of the collapsed right lung as well as the pleural surface. Similarly, no pleural effusion or hemothorax was also detected. The aorta and oesophagus were normal. The mass was completely resected.

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A gross examination of the cut sections showed that the tumour was fairly well-circumscribed and had a thin fibrous capsule. It consisted of mainly fat-like tissue, which was soft to firm and yellowish in colour. In some areas, there were nodules or tumour tissues which were slightly greyish in colour. These nodules were of various sizes and some of them contained hemorrhagic and necrotic tissue in the centre. Thin septae of fibrous tissue were also seen separating these nodules. Small cartilaginous nodules were also present within the tumour.

Histologically, the tumour tissue composed of loose and compact cellular areas. The loose areas were composed of cells with pleomorphic nuclei and florette-type of nuclei arrangement, with their surrounding areas showing plexiform vascular channels. There was no evidence of typical lipoblast. Occasional abnormal mitoses were discernible. In the other more cellular areas, the cells exhibited markedly pleomorhic nuclei with eosinophilic cytoplasm but there was no evidence of cross-striation present. Abnormal mitoses were observed. Areas of relatively mature cartilage and fibrous differentiation were noted, although there was no immature mesenchymal tissue, vascular or lymphatic permeation present. In conclusion, these features represent myxoid liposarcoma with chondroid metaplasia.

Post-operatively, the patient received radical radiotherapy of 64 Gy in 32 fractions. She had been well and symptom-free for four years after completing a radiotherapy course until recently when she came back with a recurrent myxoid liposarcoma in the right mediastinum. No evidence of distant metastasis was noted on imaging studies. The histopathological result confirmed the recurrent myxoid liposarcoma. As a result, she had another surgery and underwent a course of radiotherapy.

DISCUSSION

Despite liposarcomas being the most common soft tissue sarcoma in adults, primary mediastinal liposarcomas are rare, representing less than 1% of all mediastinal tumours[3]. These lesions usually remain asymptomatic until they reach a large size giving common presenting symptoms of chest pain, dyspnoea, cough and weight loss[1], as demonstrated by our patient as well as the finding of a large mediastinal mass on chest radiograph.

Meanwhile, the characteristic findings of a liposarcoma on Computed Tomography (CT) scan are that of heterogeneous soft tissue mass with fat density component; however, as they usually arise from primitive mesenchymal cell which has the property of lipogenesis[1], the appearance may vary, rendering it insufficient to establish the pre-operative diagnosis alone. A histopathological examination is required for final diagnosis[4]. CT scan is therefore valuable in determining the actual size, as well as the extent of the mass and involvement of the surrounding structures.

Liposarcomas are divided into five pathological subtypes, namely: well differentiated, myxoid, pleomorphic, mixed and round cell types; with myxoid liposarcoma accounts for 40 to 50% of the histological subtypes[1]. There are eleven recognizable histological patterns in myxoid liposarcoma with a variable of 1 to 7 patterns in one patient [5]. Chondroid metaplasia, in a traditional myxoid, contributed only 4% of all myxoid liposarcoma[5]. The most characteristic and defining features of the traditional myxoid pattern are its prominent delicate plexiform capillary network (so-called chicken-wire vasculature)[5], as demonstrated by our patient. Lipoblasts may or may not be visualized in this traditional myxoid pattern[5].

The survival rate seemed to depend on the pathological type, with only 40% of the patients with myxoid type were still alive after a mean interval of 13 months[1]. As for our case, the patient was fortunate as she is still alive after five years of diagnosis. These could be attributed by the early diagnosis and aggressive management, which include tumour resection and adjuvant radiotherapy.

Treatment of choice for mediastinal liposarcoma is surgical removal with total resection being the most desirable[4]. Radiotherapy and chemotherapy have been offered as adjuvant therapy for the liposarcoma to improve survival rates.

Recurrence rate is also very high in liposarcomas, which becomes apparent within the first 6 months after the resection in most cases. However, it may be delayed for 5 to 10 years after the initial excision[4]. As for our case subject, she had been disease-free for 4 years after resection and radiotherapy prior to the recurrence of the liposarcoma.

Almost all patterns of myxoid liposarcoma may develop metastases with round cell differentiation exhibiting an aggressive behaviour; however, the prognostic significance of the rest of the patterns has yet to be determined [5]. In conclusion, for primary mediastinal liposarcoma, being a rare tumour with large size at presentation, surgical resection is the treatment of choice to relieve the symptoms as well as to determine the histological subtypes. Thus, a careful follow-up is still needed due to high rate of recurrence.

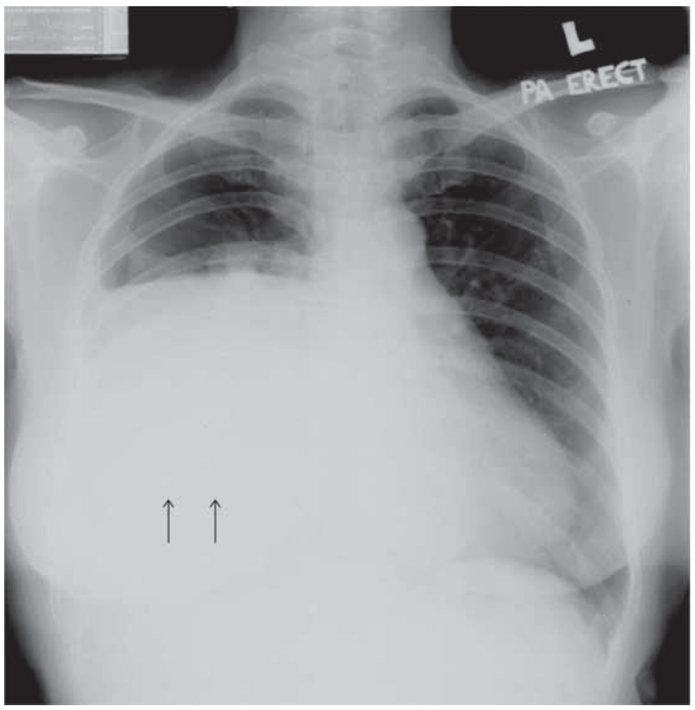

Figure 1. A frontal chest radiograph demonstrating homogenous opacity in the inferior half of the right hemithorax (black arrows), with trachea and mediastinum shift to the contralateral side.

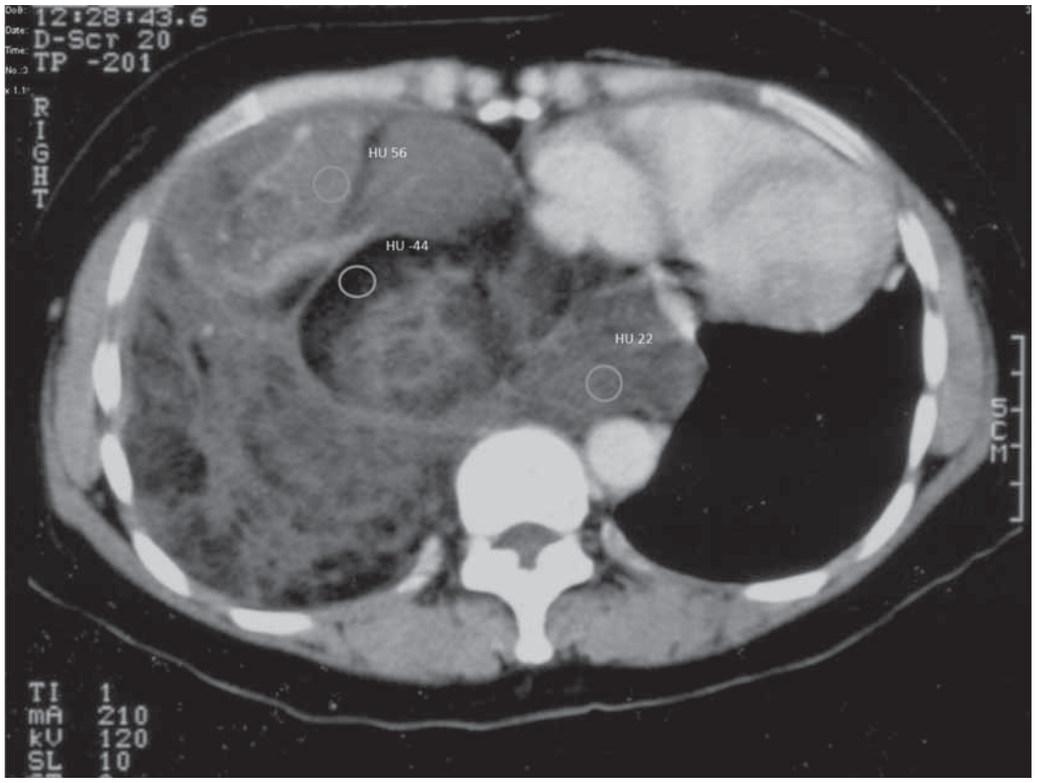

Figure 2. Contrast-enhanced axial CT thorax showing large multiseptated heterogeneous soft tissue mass with fat (HU-44), fluid (HU 22) and soft tissue (HU 56) densities. The mass is causing mediastinal displacement to the left side.

REFERENCES

[1] Hirai S, Hamanaka Y, Mitsui N, Uegami S, Matsuura Y. Surgical Resection of Primary Liposarcoma of the anterior mediastinum. Ann. Thorac Cardiovasc. Surg. 2008; 14: 38-41.

[2] Punpale A, Pramesh CS, Jambhekar N, Rajesh C Mistry. Giant Mediastinal Liposarcoma: A case report. Ann Thorac Cardiovasc Surg. 2006; 12: 425-7.

[3] Macchiarini P, Ostertag H. Uncommon primary mediastinal tumours. Lancet Oncol. 2004; 5: 107-18.

[4] Thomaz FB, Marchiori E, Guimarães AN, De Magalhães IF, Magalhães FV, Gonçalves LP *et al*. Primary mediastinal liposarcoma- computed tomography and pathological findings: A case report. Cases J. 2009; 2: 8703.

[5] Fritchie KJ, Goldblum JR, Tubbs RR, Sun Y, Carver P, Billings SD *et al*. The expanded histologic spectrum of Myxoid Liposarcoma with an emphasis on newly described patterns: Implications for diagnosis on small biopsy specimens. AJCP. 2012; 137: 229-39.