CASE REPORT

MYXOID LIPOSARCOMA ORIGINATING IN THE ANTERIOR MEDIASTINUM

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Myxoid liposarcomas of the head and neck are mostly metastatic in nature, with the primary lesion usually in lower extremities or retroperitoneum. Primary mediastinal liposarcomas are even rarer. Although there have been previous cases reporting their incidence, there is no evidence of any case comprising of both the neck and the mediastinal region yet. We here present a case of a 43 year old male with a primary liposarcoma of the myxoid subtype originating in the anterior mediastinum. This is the first time such a case has presented in literature. The patient presented with a right sided neck swelling associated with pain and shortness of breath upon lying down. CT neck revealed an enhancing mass extending from right cervical region to axilla and encasing the subclavian artery and involving a portion of the right lung. The patient underwent wide local excision of the mass along with chemoradiotherapy to prevent recurrence.

Keywords: Myxoid; liposarcoma; soft tissue sarcomas; head and neck

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INTRODUCTION

Myxoid liposarcoma is a subtype of liposarcoma which is categorized under soft tissue sarcomas. Soft tissue sarcomas (STSs) are malignant neoplasms originating from non-epithelial sources. They are mesenchymal tumours which are very common in adults. Their occurrence in the head and neck region however, is rare making up only 1% of all head and neck cancers. With an annual incidence of 2.5 cases per million population , liposarcomas are the second most common type of soft tissue sarcomas (14%) with malignant fibrous histiocytoma (24%) being the most common soft tissue sarcoma.[1,2] Majority of the liposarcomas in adults are detected more commonly in the lower extremities (43%), in particular the buttocks and the thighs, and at other instances, the retroperitoneum.[3] Histopathologically speaking, liposarcomas are further divided into five subtypes: well-differentiated, dedifferentiated, round cell, myxoid and pleomorphic types.[4]

Little work has been done on addressing the head and neck liposarcomas. The few case reports that have been published have only discussed a small number of patients since this tumour makes up only approx 4% of liposarcoma cases.[5] Also, majority of the cases involving the neck and the mediastinum liposarcomas have reported it to be secondary in relation to metastasis from another primary commonly located in the extremities. In this case report, we present a very rare case of primary myxoidliposarcoma discovered in the neck region with mediastinal invasion and encompassment of the subclavian artery in a 43 year old gentleman. There has not been any report or evidence of a primary myxoidliposarcomas comprising both the neck and the mediastinal region previously. We have extensively reviewed the literature on pub med and Google scholar and no cases of primary myxoid liposarcomas involving the neck and mediastinum could be found. This case report also reviews the relevant literature related to this presentation.

CASE REPORT

A 43 years old gentleman, known case of hypertension for last 2 years, reported with complaint of a painful right sided neck mass since 1 ½ years. The mass was associated with a 3 month history of progressive pain which was pressing in character and this was the reason for his visit to the clinic. Pain had become constant in nature since 7–10 days. There was also history of shortness of breath on lying down since the past 2 months. No history of fever, dyspepsia or change in voice. His sleep, energy, appetite, and mood were unaffected since the onset of the disease.

Local examination of the neck revealed right sided neck swelling in Level III-V extending below clavicle inferiorly and laterally, measuring around 4×10 cm, rounded, firm, non-tender, not attached to overlying skin with no other palpable neck nodes. ENT examination was within normal limits.

Ultrasound of the neck region done previously showed a neck mass on the right side with Supraclavicular part measuring 4.7×2.2 cm and Infraclavicular part measuring 4.3×2.0 cm

respectively. FNAC also carried out previously revealed spindle cells with a myxoid background indicative of myxoid liposarcoma.

Patient was advised MRI but he got CT scan done which showed an enhancing mass measuring 4.8×12.6 cm. The mass exhibited slight heterogeneity, which is characteristic of the myxoid subtype and extended from Right cervical to below supraclavicular region and axilla (Figure-1). A small projection was found in Right lung apex but there was no rib erosion. The mass was partly encasing (abutting) the right subclavian artery and fully encasing subclavian vein (Figure-2).

A tumour board meeting was called to discuss the case. Tumour board review is a treatment planning approach in which a number of doctors who are experts in different medical specialties review and discuss the medical condition and treatment options of a patient. Tumour board planned for wide local excision of the right neck and thoracic mass, along with thoracotomy for surgical resection of the involved potion of the lung to be performed by the Cardiothoracic Surgeon. An ENT surgery team was also involved as the tumour was enclosing the subclavian artery.

In the current case, the patient underwent an extensive surgery which involved the ENT and the cardiothoracic surgery teams working together since the tumour had a very invasive spread and was enclosing the subclavian artery. The tumour was completely excised and chest drains were placed. The affected portion of the right lung was resected. Intraoperative findings included a lobulated mass arising from the supraclavicular fossa extending into the Infra-clavicular space, abutting the subclavian vessels and right chest wall but not invading it or entering into right pleural cavity. Histopathology report showed features of myxoidliposarcoma with some component of round cell 5% Grade 2. The patient was recommended to undergo radiotherapy (at radiation dose 60Gy) upon discharge with the aim of strengthening the effect of the treatment and reducing the risk of recurrence.

Postoperatively, diet was started, patient mobilized and analgesic control provided. Chest drains were kept on suction and later removed. Chest physiotherapy was carried out with incentive spirometry.

The patient was discharged oral pain killers and antibiotics. Chemotherapy 25 cycles with Lipozoid has been planned Post-operatively, the patient has been healthy for 10 months. Follow-up surveillance will continue.

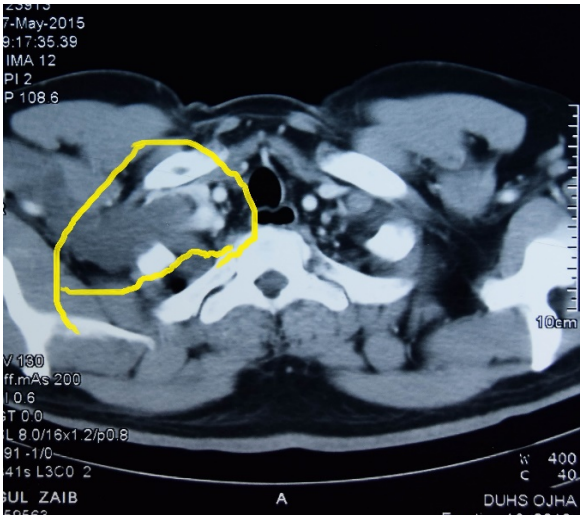

Figure-1: Computed Tomography scan of the chest with contrast showing the lesion (axial section)

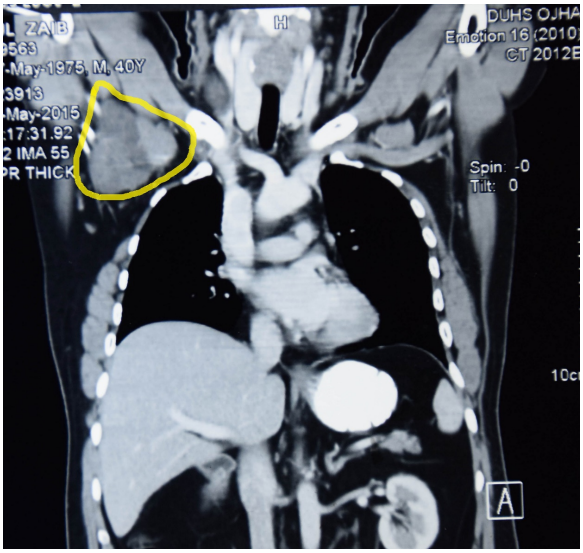

Figure-2: Computed Tomography scan of the chest with contrast showing the lesion (coronal section)

DISCUSSION

Liposarcomas arise from the deep situated adipose tissues of the body and have the characteristics of malignant mesenchymal neoplasms. Russell *et al*[6] has found in a review of soft tissue sarcomas that only 221 out of the 1215 sarcoma cases were reported to be liposarcomas. More importantly, only 10 (4%) out of those 221 were found to be located in the head and neck region. Primary liposarcomas in the mediastinum are very rare. In a study conducted by Hahn and Fletcher, it is seen that there are only 24 cases of mediastinal liposarcoma over the course of 13 years (1992–2005). Amongst those 24, only 9 were found to be originating in the anterior mediastinum and of those, only 2 were of the myxoid subtype.[7]

As already mentioned, liposarcomas are classified in further five subtypes. These subtypes basically help in correlating the clinical picture of the disease with its prognosis and also in deciding the treatment options. The most common subtype amongst these is the myxoid, found to be present in almost 45–55% of the cases. Myxoid tumours are generally of low or intermediate grade, and are seen to have a decent prognosis. Its high grade counterpart is said to be the round cell subtype which represents about 15% of the total number of liposarcoma cases. The rest comprise of well-differentiated subtype with the lowest grade and the best prognosis, and the pleomorphic subtype with the highest grade and the worst prognosis. Also, the pleomorphic and the round cell types have been seen to have a higher rate of local recurrence and also, a higher risk of distant metastasis. According to the histopathology reports, our patient had myxiod subtype of liposarcoma with some component of round cell.

These neoplasms are believed to be a result of a number of chromosomal abnormalities. The pathogenesis behind the myxoid subtype is translocation, with the most common being the t (12;16) (q13;p11.2) translocation. This involves the fusion of CHOP gene present on the chromosome and is normally responsible for regulating the process of adipocyte differentiation with the TLS gene present on chromosome 16.[8,9]

Liposarcomas in the neck are usually misdiagnosed since they mostly have an asymptomatic course and appear very similar to a number of more commonly encountered benign lesions such as lipomas on gross inspection.[1,9,10] Hence, multiple imaging is required with MRI being the best option. In our scan, an MRI was advised but the patient went for a CT scan which exhibited slight heterogeneity, which is characteristic of the myxoid subtype. The gold standard for the diagnosis is however the biopsy. In our patient, a fine needle aspiration biopsy was performed which confirmed the diagnosis of the myxoidliposarcoma of a low grade with multiple number of lipoblasts present. Low grade myxoid tumors usually have an abundance of spindle-shaped cells and are considered to be of the least cellular form.[11–13]

Once identified, the recommended treatment options include surgery with wide local excision performed and a consideration to undergo radiotherapy following the operation. Postoperative radiotherapy is considered to be the standard approach for nearly all intermediate or high-grade soft tissue sarcomas and the recommended postoperative radiation dose is 60–66 Gy in 1.8–2 Gy fractions. Local recurrence can develop in approximately 13% of the patients. According to a study done on the outcome of conservative surgery and radiation therapy in patients with liposarcoma, the 5 year, 10year, and 15-year survival rates were 79%, 69%, and 61%, respectively.[14]

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