

Case Report

Myxoid liposarcoma presenting like a cystic neck swelling

**Harmeet Sahni*, Raj Gautam, Vikash Lal, Tanveer Parvez Shaikh,
Karishma Palshetkar, Aditya Oak**

Department of Surgery, DY Patil College of Medicine, Nerul, Navi Mumbai, Maharashtra, India

Received: 13 March 2015

Revised: 20 March 2015

Accepted: 03 April 2015

***Correspondence:**

Dr. Harmeet Sahni,
E-mail: h.sahni@ymail.com

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ABSTRACT

Soft tissue sarcomas are rare and unusual neoplasm's, accounting for approximately 1% of adult human cancers and 15% of pediatric malignancies. Most liposarcomas occur in deep soft tissues of the extremities and retroperitoneum; only a very small percentage (as low as 2%) occurs in the head and neck. Here we present a case of a 48yr old male with a painless mass in the neck clinically presenting as a cystic swelling. The CT neck was suggestive of cystic lesion in the neck representing benign cystic lesion. The swelling was excised and sent for histopathology which was suggestive of myxoid liposarcoma. The patient was planned for wide excision of the edges and scar and adjuvant chemo and radio therapy.

Keywords: Myxoid, Liposarcoma, Cystic swelling, Sarcoma, Neck swelling

INTRODUCTION

Sarcoma presenting in soft tissue are rare and usually with unusual type of neoplasm and they account for 1% of adult population and 15% of pediatric cancers.¹⁻³ Sarcoma classification is based on site of tumor. Soft tissue sarcoma is malignancy arising from muscles, fat, vessels, peripheral nervous system and fibrous tissue. It is divided in 4 types based on histology and they are a) well differentiated liposarcoma, b) myxoid round cell liposarcoma, c) pleomorphic liposarcoma, d) non differentiated liposarcoma. The final clinical course of the disease and prognosis depends on the histology type.

CASE REPORT

A 48 year old man came to the outpatient department with history of swelling over the right side of neck since 3 years (Figure 1). The swelling was gradually increasing in size and was painless. The patient had no history of loss of weight, appetite. Patient had no history of difficulty in swallowing, breathlessness, change of voice.

The patient had no history of addictions to tobacco or alcohol except chewing beetle nut leaves. The patient was a known diabetic on oral hypoglycaemic agents with no history of hypertension, Koch's, Koch's contact, bronchial asthma, ischaemic heart disease. The patient had no previous medical or surgical history and no history of previous similar illnesses or drug allergies.



Figure 1: Swelling in the neck.



Figure 2: X-ray neck of the patient.

On examination the patient was moderately built and moderately nourished. Local examination revealed a soft cystic 10x8 cm sized swelling in the right anterior triangle of the neck with no skin changes, scars, sinuses or dilated veins. The swelling was well circumscribed in the anterior triangle of the neck and did not move with deglutition or protrusion of tongue. The swelling was brilliantly transilluminant and was freely mobile in all the planes. Provisionally the diagnosis was considered as cystic hygroma or branchial cyst due to the brilliantly transilluminant nature and the cystic consistency. X-ray of neck was done (Figure 2).

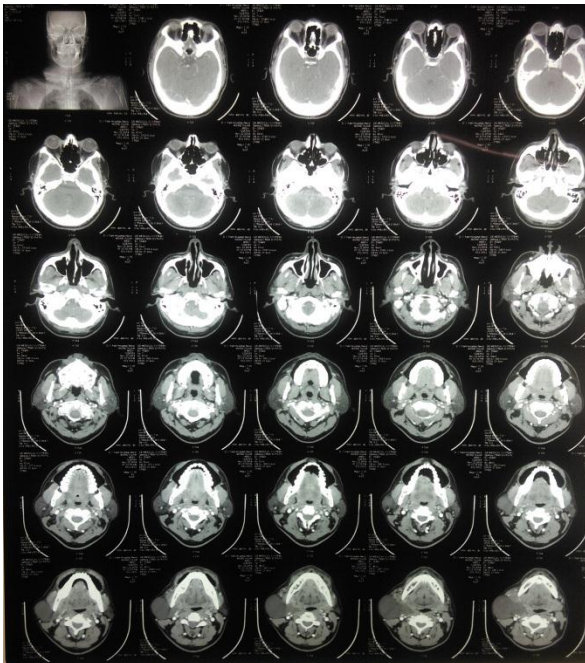


Figure 3: CT scan image.

CT neck (Figure 3, 4) was suggestive of large well defined cystic lesion in the anterior triangle of neck on right side measuring 8x6.7x6 cm. The lesion showed few thin enhancing septae within representing benign cystic lesion most likely lymphangioma. Superiorly the lesion reached the submandibular region displacing the submandibular gland anteriorly. Posteriorly the lesion extended along the right sternocleidomastoid muscle and

abutted the CCA and ICA. Medially there was focal extension of the lesion through the thyrohyoid membrane in the right para-laryngeal space and compressed the laryngeal ventricle. Anteromedially the lesion abutted the strap muscles of the neck. The naso-oropharynx, thyroid, parotid and submandibular glands were normal.



Figure 4: CT scan image.

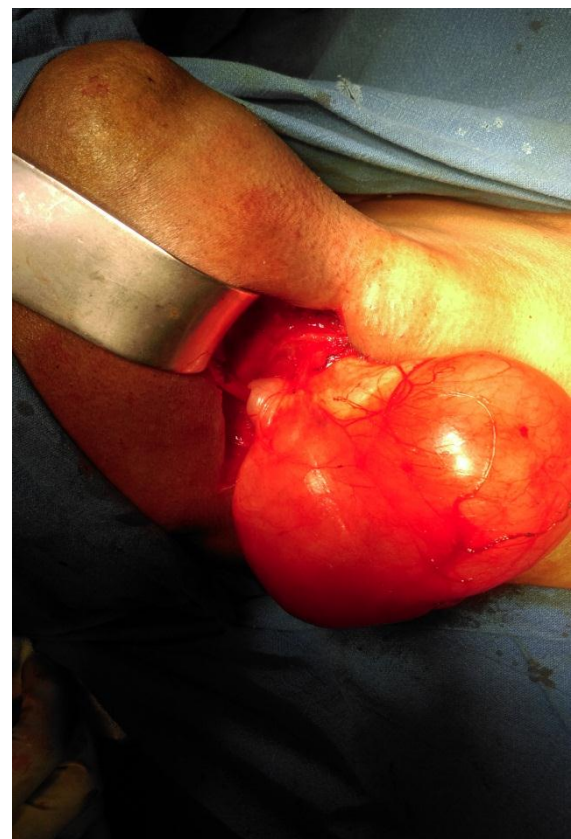


Figure 5: Intra op image.



Figure 6: Histopathology image.



Figure 7: Histopathology image after cutting the tumor.

Oesophagoduodenal scopy was done and was suggestive of the swelling indenting just above right vocal cord due to which the right cord was not visualized. Intra operatively (Figure 5, 8) the swelling was present in the subcutaneous plane and was dissected out from all sides and excised. The swelling was a single soft globular swelling within a capsule. The swelling was cut open and white jelly like in consistency. This was sent for histopathology examination to Hinduja Hospital. Histopathology specimen was 12x10x7 cm size weighing 300 gm (Figure 6, 7). It was a multinodular mass which was slimy internally as well as externally. Cut surface was multinodular grey white myxoid. Microscopy showed myxoid liposarcoma with characteristic hypocellular nodules and myxoid stroma. Tumor showed characteristic plexiform capillary network with cyst like areas. Scattered lipoblasts were also seen. It was diagnosed as moderate grade myxoid liposarcoma. Post operatively (Figure 9), an Oncology opinion was taken and patient was advised adjuvant chemo and radiotherapy and revision of the surgery with wide excision of edges and scar excision.



Figure 8: Intra op image of tumor excision.

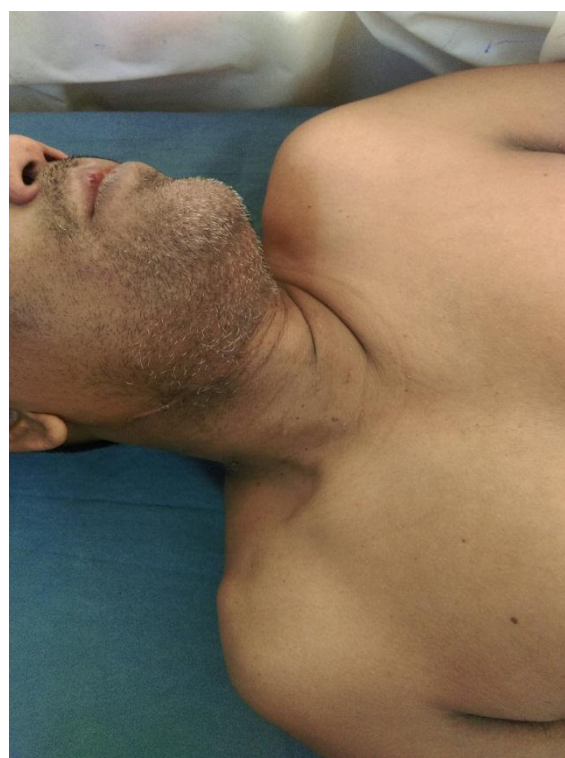


Figure 9: Post op scar.

DISCUSSION

Liposarcomas account for 8 to 17% of soft tissue sarcomas.⁴ The common sites of occurrence of liposarcoma are extremities and retroperitoneum. Head and neck liposarcomas are rarer with as low as 2 to 3% incidence.⁴ myxoid and round cell liposarcomas have a peak incidence among those in their 30s.⁵ Liposarcoma is classified into three biologic groups encompassing five histologic subtypes, based on strict morphologic features and cytogenetic aberrations - well-differentiated, dedifferentiated, myxoid, round cell, and pleomorphic. The well-differentiated and dedifferentiated subtypes account for 42% and 21% of liposarcomas, respectively, and are more commonly found in the retroperitoneal location; the myxoid, round cell, and pleomorphic subtypes account for 25% and 8% of liposarcomas, respectively, and are usually located in the extremity.⁵

Well differentiated and myxoid liposarcomas are usually associated with good prognosis, whereas young age at presentation and female patients tend to suffer from high grade tumors and a poor prognosis.⁴

Myxoid liposarcomas account to 45 to 55% of all liposarcomas.⁴ Pure myxoid liposarcomas rarely metastasize to the lymph nodes. In contrast to other liposarcomas they can metastasize to the lungs, bone, pleura, and pericardium. Myxoid Liposarcomas are best treated with surgical excision with a wide margin. They have a survival rate of 80% at 5 years and 50% at 10 years,⁶ which is usually determined by the negative margins achieved at the time of surgery. Myxoid liposarcomas are known to have a high risk of local recurrence. The evidence of positive excision margins increases the risk of tumor recurrence (around 60%) in the absence of re-excision.⁷ This resumption must be scheduled from the results of a MRI Scan which looks for potential tumor anatomical residue and should be performed once proper healing of the initial surgery has been achieved. The re-excision procedure takes place at the initial site of resection and is performed with a wider margin of at least two centimeters of healthy tissue. All tissue potentially exposed to viable tumor cells at the initial procedure should be removed at that time, including the surgical scar and the orifices of drainage. A complementary radiation therapy should be implemented on the operating site, with a minimal safety margin of five centimeters, combined with adjuvant chemotherapy.^{8,9}

CONCLUSION

Myxoid liposarcoma rarely presents in the head and neck. It can mimic as a cystic lesion as well. Re excision of the initial site of resection including at least two centimeters of healthy tissue along with the surgical scar and orifices of drainage along with adjuvant radio and chemotherapy have to be implemented for better survival rate.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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DOI: 10.5455/2320-6012.ijrms20150547

Cite this article as: Sahni H, Gautam R, Lal V, Shaikh TP, Palshetkar K, Oak A. Myxoid liposarcoma presenting like a cystic neck swelling. Int J Res Med Sci 2015;3:1281-4.