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Case Report

Myxofibrosarcoma of neck: A rare case

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ABSTRACT

Myxofibrosarcoma (MFS) is commonly seen among elderly patients, usually sixth to eighth decade of life. Most of these tumours arise in the extremities (lower limbs > upper limbs) and are less commonly seen on the Trunk, Head and Neck, Retroperitoneum and Pelvic areas. The World Health Organization (WHO) defines MFS as the malignant fibroblastic neoplasm characterized by cellular pleomorphism, variably prominent myxoid stroma, and prominent elongated, thin-walled stromal blood vessels. Head Neck MFS is rare site of occurrence with 19 cases reported worldwide till date, described subsites being Maxillary sinus, Infra Temporal fossa, Pterygopalatine fossa, or Parotid gland. The Surgical wide local excision is the main modality of treatment for Non metastatic stage followed by Adjuvant Radiotheraphy.

We are presenting a young adult male, presenting with Asymptomatic progressive neck mass, predominantly in posterior triangle distorting upper Aerodigestive tract anatomy. The Diagnosis of MFS done with MRI imaging of neck, Trucut biopsy and CECT scan thorax. With the anticipation of difficult intubation, Broncoscopic directed intubation is planned and done. The surgical wide resection amounted in excision of Sternomastoid, IJV, SAN and part of posterior triangle bed muscles for getting margin free status. The Histopathology showed spindle cells with Myxoid stroma, areas of necrosis with circumferential free margins, along with IHC showed positive for vimentin and CD34 with diagnosis as High Grade Myxofibrosarcoma. Post op patient was adviced Adjuvant Radiotheraphy.

1. Introduction

Myxofibrosarcoma (MFS) comprises a spectrum of malignant fibroblastic neoplasms with variably prominent myxoid stroma, cellular pleomorphism and distinctive curvilinear vascular pattern. This tumour commonly seen among elderly patients, usually sixth to eighth decade of life. It is commonly seen among males.[1] Most of these tumours arise in the extremities (lower limbs > upper limbs) and are less commonly seen on the trunk, Head and Neck, Retroperitoneum and Pelvic areas.[2] MFS is classified into low grade tumors, with low metastatic potential, and high grade tumours. High grade MFS has metastasis of 20-35% with Lung and Bone being the most common sites. Surgical wide local excision is the main modality of treatment in Resectable tumors in Non metastatic stage followed by Adjuvant Radiotheraphy for those with high risk features. The overall recurrence rate for MFS has been reported to be in the range of 50-60%.[3]

Here we present a case of Myxofibrosarcoma in the posterior triangle of neck extending to prevertebral space in a young adult.

2. Case Report

An 18year old young male patient presented with a painless progressive swelling in the right side of the neck from

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6 months. No history suggestive of tuberculosis/ Upper aerodigestive tract cancer. On examination was found to have a solitary, round, firm, lobulated, non tender swelling of 13x12cm located in the posterior triangle extending upto just short of base on the right side of neck.(Fig 1)The anterior border of the swelling was deep to Sternomastoid muscle, posterior border extends underneath anterior border of trapezius muscle. The swelling has restricted vertical mobility.The skin over swelling looked normal and pinchable. No other swellings/ Lymphnodes were palpable in the neck. There was no lower cranial nerve deficits noted on right side. The oral cavity, upper aerodigestive tract and thyroid gland were normal.

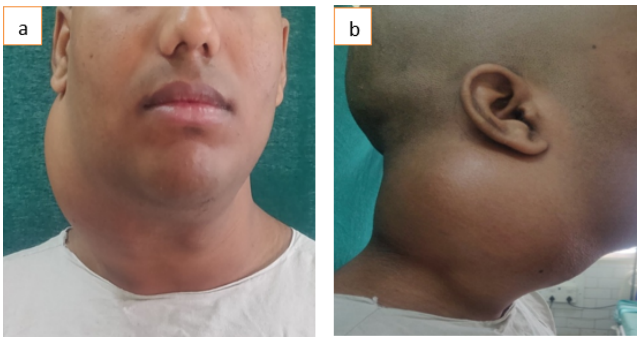

Fig. 1: a&b: Clinical picture showing Front and Side view of the Neck Swelling

The Ultrasonography of the neck showed a large well defined cystic lesion with solid areas taking colour, measuring 11.7 x 7.0 x 8.5cm in the right posterior triangle of neck. MRI of the neck was done for further evaluation which showed a large well defined multiloculated lesion (11.3 x 9.3 x 10.6cm) with its epicentre in right Posterior triangle extending to Paravertebral space, without intraspinal extension. The lesion is noted displacing the right carotid vessels, submandibular gland, parotid gland and sternocleidomastoid muscle anteriorly. Medially the lesion is noted abutting the posterior aspect of tongue and displacing the hypopharynx, esophagus, larynx and trachea to the left. A preoperative USG guided Trucut biopsy showed features of a MyxofibroSarcoma. The chest CECT scan done to rule out lung mets. Direct Laryngoscopy done, showed distorted anatomy of oro and laryngo pharynx with difficult visualization of cord movement, indicating difficult ET intubation.

The patient was worked up for Surgery and as planned awake Bronchoscopy guided Nasal intubation done with plexometallic tube. The patient is put in Supine position with neck extended turned to opposite side. The Curvilinear incision is taken extending from right mastoid to cricoid cartilage .The subplatysmal flaps are raised upper upto submandibular gland region and lower upto clavicle. Intraoperatively the mass of 12 x 8 cm was noted underneath the thinned out sternocleidomastoid muscle. The tumor

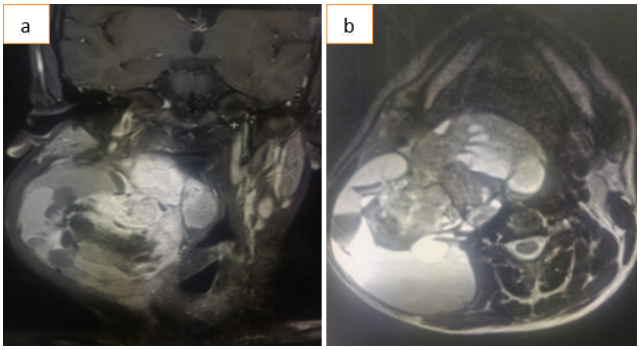

Fig. 2: a: MRI Axial section showing the tumor extent with compression of upper aerodigestive tract and pushing the Carotid-IJV anteriorly; **b:** MRI Sagittal section showing the tumor with Solid-cystic areas extending upto skull base and medially anatomical distortion of upper aerodigestivetract

found extending medialy upto pharyngeal musculature and Esoghagus, displacing carotid sheath contents anteriorly. The mass found deeply invoving prevertabral fascia as well Accessory Nerve, extending upto the base of the skull.

The Sternomastoid muscle transected at upper third and lower third, Carotid sheath dissected to find internal jugular vein involvement, hence it is transected by clamping, cut and doubly ligated. The carotid and vagus dissected out anteriorly. The tumor is mobilised inferiorly by ligating EJV posteriorly from Trapezius, SAN is sacrificed due to tumor infiltration. Then the tumor dissected along with prevertebral fascia upto level II area, identifying and preserving XII cranial nerve. The tumor bed is marked with titanium clips and hemostasis achieved wee.

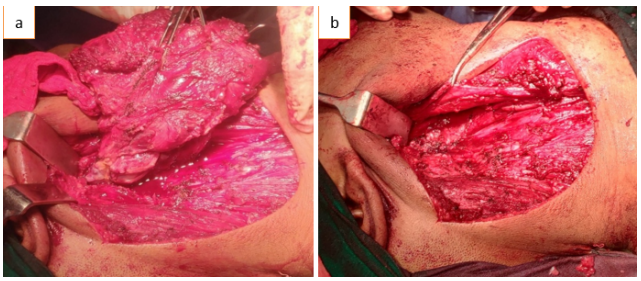

Fig. 3: a: The tumor being dissected from posterior triangle and separated from carotid-X CN, after ligating a segment of IJV; **b:**The tumor bed after resection, formed by trapezius, splenius, scalene and prevertebral muscles.

Macroscopically, the mass was heterogenous in appearance with solid -cystic spaces filled with myxoid material, necrotic and haemorrhagic areas. Microscopy showed spindle shaped cells having oval to elongated pleomorphic nucleus with scant cytoplasm and indistinct cell borders. > 10 mitoses/10 hpf were noted. Circumferential resected margins were free of tumour.IHC showed positive for vimentin and CD34 with diagnosis as

High Grade Myxofibrosarcoma.

The postop recovery Uneventful and the Patient advised for adjuvant radiotheraphy.

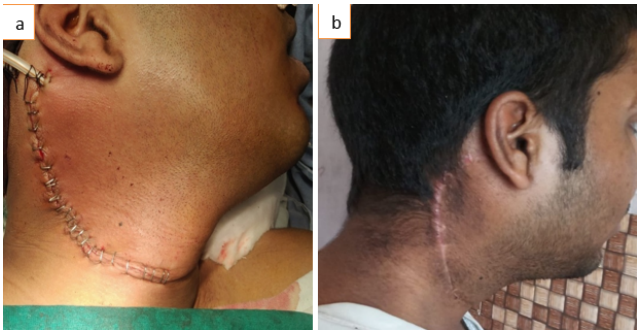

Fig. 4: a: Neck incision closure with suction drain; **b:** Patient at 4 weeks post surgery.

3. Discussion

Myxofibrosarcoma (MFS) is a fibroblast-derived sarcoma, which accounts for approximately 5–10% of all soft tissue malignant tumors.[1] The World Health Organization (WHO) defines MFS as the malignant fibroblastic neoplasm characterized by cellular pleomorphism, variably prominent myxoid stroma, and prominent elongated, thin-walled stromal blood vessels.[2] The mean age in patients with MFS is between the fifth and seventh decades.[3] Although some studies show a slight male predominance,[4] the current evidence suggests no significant gender predilection.[5] About 77% of MFS cases occur in the extremities with a predilection for the upper extremities. Other areas of the body including the trunk (12%), retroperitoneum or mediastinum (8%),[4,6] abdominal wall heart have also been reported.

There are very few reports of cases (19 cases) in the head and neck regions are available in the literature with varied location like maxillary sinus, infra temporal fossa, pterygopalatine fossa, or parotid gland. The most common site of metastatic disease in cases of extremity myxofibrosarcoma is the lungs, while no such data is available with regards to head and neck myxofibrosarcoma.[3]

In our case, patient being young (18 years old), presenting painless progressive swelling in posterior neck, made us to suspect as lymphnodal mass or soft tissue mass. Being located in posterior triangle neurogenic tumor being rare, with the help of Ultrasound we could get detailed information of the tumor with possibility of Sarcoma in view of hypoechoic necrotic areas and hyperechoic myxomatous material. We did MRI imaging and tissue diagnosis with Trucut biopsy as Fibrosarcoma with myxoid component, metastases to lung is ruled out with CECT thorax. As per imaging the tumor looked operable with sacrifice of Spinal accessory Nerve, Sympathetic chain and Internal jugular vein. As the upper aerodigestive tract anatomy was distorted imaging wise due to tumor compression and non-visualization of both vocal cord in Direct laryngoscopy, difficult intubation is anticipated and Bronchoscopy guided intubation is planned. It is unusal for a majority posterior triangle tumor to have difficult airway, as we experienced in our case

Macroscopically MFS can be categorised into two groups: Dermal (subcutaneous/superficial) and Intramuscular (Subfascial / deep). The dermal group of tumors infiltrate and spread extensively in a longitudinal manner in the subcutaneous plane while the tumors arising in the Intramuscular plane were noted to form a single discrete mass.[5] In our case, as the tumor was both in dermal and inter muscular plane, on table we experienced ill defined plane with the neck muscles, at the deeper surface. The theoretical margin of 2-3 cms is not possible in the large neck mass due to closeness to important anatomical structures. Adjuvant Radiotheraphy has to be considered marking the tumor bed with titanium clips.

4. Abbreviations

MFS-Myxofibrosarcoma, MRI- Magnetic Resonance Imaging, IJV-Internal Jugular Vein, CECT- Contrast Enhanced Computerised Tomography, SAN-Spinal Accesory Nerve.

5. Source of Funding

None

6. Conflicts of Interest

None

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