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Case Report

Giant Cervico-Mediastinal Myxolipoma in a 6 Year Old Child

Abstract

Lipomas are benign mesenchymal tumors that may present anywhere on the body. However, myxolipoma, the histological variant is rarely seen. We report a rare case of giant anterior cervico-mediastinal myxolipoma in a 6 year old child, with discussion over diagnostic and management strategies for same.

Introduction

Lipomas are benign mesenchymal tumors, seen all over body, usually occurring in 5th and 6th decades of life [1]. 25 % of lipomas are seen in Head & Neck area and mostly in subcutaneous region, posterior triangle neck [2]. Usually slow growing, they are generally asymptomatic, till reach great size to cause cosmetic defect or pressure symptoms on surrounding structures or impair function or mobility. Lipomas greater than 10cm in width or more than 1000g in weight are called giant tumors [3].

Myxolipoma is one of the extremely rare histological variant of lipoma, accounting for <1% of lipomas [4], characterized by mature adipose tissue and abundant mucoid substance.

Here we report successful surgical excision of giant anterior cervical myxolipoma in a child. The rarity of this variant and huge size of the tumor in a six year old makes this interesting.

Case Report

A six year male, child, resident of Saharanpur, Uttar Pradesh, presented to our out-patient department with complaint of gradual onset, painless, progressive single, swelling neck for three years. The child had no history of breathlessness but occasional difficulty in swallowing was present. There was no history of sudden increase in size, voice change or fever. There was no history of similar swelling in other three siblings. There was no history of previous medication or surgery for same.

On examination, the mass was 10cm X 5cmX 5cm, normal temperature, non-tender, lobulated, firm, fixed to underlying structure, but free overlying normal skin, extending from chin to clavicle involving left anterior triangle of the neck, displacing trachea to the right and left common carotid anteriorly (Figure 1). Finger could not be passed between the lower end of mass

and suprasternal notch, suggesting anterosuperior mediastinal extension. There was no other palpable mass or cervical lymph node. Oral cavity examination was within normal limits.

The Chest X- ray PA view confirmed tracheal deviation to right (Figure 2). Fine needle aspiration cytology revealed fragments of mature adipose tissue and fibrous tissue in background with no nuclear atypia. Ultrasonography Neck revealed lobulated soft tissue mass and Doppler ruled out hypervascularity as is usually seen in malignant mass.



Figure 1: Pre-operative clinical picture showing a huge neck mass extending from chin to clavicle, with common carotid overlying it.



Figure 2: Chest X-ray PA view showing left cervical mass pushing trachea to the right.

Contrast enhanced computed tomogram showed 10.2cmx5.5cmx5.2cm, large, unencapsulated diffusely infiltrative lipomatous mass showing multiple thin and thick septations with nodularity. Superomedially the mass was extending into the parapharyngeal space and deviating the left oropharynx airway towards right. Posterosuperiorly the mass was extending in the prevertebral region with extension into the cervical neural foramina and foramen transversarium at C4-5; C5-6; C6-7 levels, causing widening of neural foramina with thinning and scalloping of adjacent pedicles and lamina (Figure 3). Anterosuperiorly the mass is displacing the carotid sheath anteriorly and medially the esophagus, larynx, trachea and thyroid gland were displaced contralaterally with no luminal narrowing. Inferiorly, small extension was seen into the superior mediastinum with splaying at the origin of left common carotid artery and left subclavian artery (Figure 4).

All routine blood and urine investigations were within normal limits. The informed consent was taken from parents. The patient was taken up for excision of mass via cervical horizontal incision and raising superior, inferior subplatysmal flaps under general anaesthesia. The mass was seen pushing the left common carotid anteriorly and left internal jugular vein compressed posterolaterally. The mass was dissected from the surrounding structures carefully and removed completely, special care being taken to remove the extensions between the cervical vertebrae. The flaps were sutured over drain. The child received normal diet and intravenous antibiotics, analgesics. The drain was removed after 48 hours, suture removed after seven days and child was sent home.

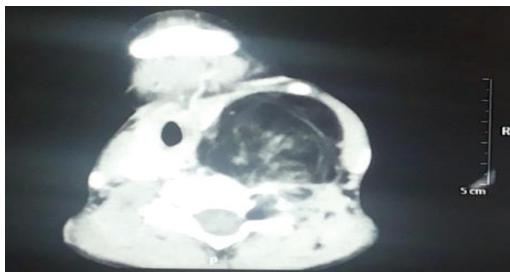


Figure 3: Axial section of Contrast enhanced computed tomogram showing diffusely infiltrative lipomatous mass with multiple thin and thick septations and extension into the cervical neural foramina and foramen transversarium.



Figure 4: Sagittal section CECT Neck showing the mass displacing the carotid sheath anteriorly and small extension into the superior mediastinum.

Pathological examination of the excised specimen revealed grey white encapsulated soft tissue mass measuring 12x7x5cms, grossly nodular, with cut section revealing grey white to grey-yellow firm areas with no evidence of haemorrhage or necrosis (Figure 5). The microscopic examination showed benign well circumscribed encapsulated mesenchymal tumor comprising of mature adipose tissue separated by fibrous septae, with areas of myxoid change consisting of stellate shaped cells (Figure 6). Myxoid areas stained strongly with alcian blue (pH 2.5). No cellular atypia, mitotic figures, hemorrhage or areas of necrosis were seen.

The patient is under follow-up for last one year, with no function loss and good cosmetic scar, without any recurrence.

Discussion

Lipoma is rarely observed in the anterior neck [5]. The fibrous capsule around, differentiates it from normal adipose tissue seen in obese people. They have slow growth and are usually asymptomatic. Numerous lipoma are mainly seen in men, while single lesion is more common in females. They are mostly seen in adult, yet some congenital lipoma has been seen [6].

There are varieties of histological variants of lipoma, like angiolipoma, fibrolipoma, spindle cell lipoma, osteolipoma, myelolipoma, chondrolipoma, myolipoma and myxolipoma. Myxolipoma is a lipoma admixed with abundant mucoid substance [7]. This mucoid substance stains positively with alcian blue and gets digested by hyaluronidase. This



Figure 5: Excised lipomatous mass of 12x7x5cms and grossly nodular.

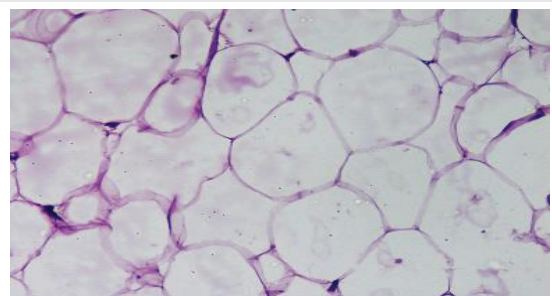


Figure 6: The H&E stain, 40x magnified histopathology slide microscopic picture showing mature adipose tissue separated by fibrous septae, with areas of myxoid change consisting of stellate shaped cells.

characteristic differentiates this from mucoid substance seen in Chondrolipoma. The presence of atypical lipoblasts (absence of vacuoles, irregular shaped nuclei and increased cell size) and rich capillary network seen in myxoid liposarcoma, differentiates it from myxolipoma on histopathology.

The exact etiology of lipoma is difficult to identify, but genetic, endocrine and blunt trauma are considered the most common. Rupture of the fibrous tissue and anchorage connections between the skin and deep fascia after blunt trauma may result in proliferation of the adipose tissue [8]. It is also postulated that trauma induced cytokine release triggers pre-adipocyte differentiation and maturation [9]. Lipoma are also known to be associated with various syndromes such as Gardner's (intestinal polyposis, osteomas), Madelung's (lipomatosis of head, neck, shoulder and proximal upper extremities) and Dercum's disease (multiple painful subcutaneous lipomas) [1,6]. Our patient had no identifiable etiological factor or any syndrome.

The Computed Tomogram (CT) scan for lipoma show homogeneous mass with encapsulation, no contrast enhancement and scanty septation [10]. On Magnetic Resonance Imaging (MRI) lipoma have high signal intensity on T1-weighted images [10].

There are diverse methods of treatment available for lipoma. The treatment may vary from conservative like steroid injections, to cosmetic surgery like liposuction, to complete surgical excision [6]. Despite the range of treatment options available, the main treatment followed for giant cervical lipoma is complete excision, which was done in our case. Steroid injections, cause fat atrophy and are used for smaller lipomas, but they require several sitting and may even depigment the overlying skin [6]. Liposuction is also used in small lipomas, however removal of entire tumor is difficult and it leaves fibrous capsule [11].

Possible complications after surgery are hematoma, seroma, ecchymosis, adjacent vessel or nerve injury, fat emboli, infection and excessive scarring [5,6]. Recurrence is rarely seen, but residual is common, often due to large size, surrounding adhesions or infiltration into adjacent muscle [6]. None of the above complications was seen in our case.

Conclusion

Myxolipoma is a rare variant of lipoma, which contains mature adipose tissue and mucoid substance. They are asymptomatic, progressive and reach huge size before patient seeks treatment. Main treatment is complete surgical excision as it avoids recurrence and other complications associated with conservative measures.

Informed consent

Written informed consent was taken from the patient parents.

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