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Case Report

A huge high rising cervico-facial lymphangioma in an adolescent male: from pterygoid to hyoid

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ABSTRACT

Cervicofacial lymphangioma is a benign swelling of the neck and is commonly seen in paediatric population. Common sites of involvement are the submandibular space and the posterior triangle and rarely involves the face. We present a rare case of a huge cervico-facial lymphangioma in a 14 year old male where the swelling presented one year back and had progressively increased in size extending from the medial pterygoids and pterygomandibular space above to myelohyoid muscle below. Intraoperatively, it was closely abutting the submandibular gland mimicking salivary gland retention cyst and the facial artery was traversing through the mass. Complete surgical excision of the mass was performed along with removal of the ipsilateral submandibular gland. Salivary gland mucocele and cervicofacial lymphangioma were the two main differential diagnosis in our case. Intraoperative findings of macrocystic spaces with septations pointed towards a lymphangioma. Post operative histopathology confirmed it to be lymphangioma lined with endothelial cells. We concluded from this case that cervicofacial swellings in higher age groups should have lymphangiomas as differential diagnosis. As the swelling ascended up to the skull base, careful radiological assessment, meticulous surgical plan and cranio-caudal exposure from the zygomatic arch above to the hyoid below helped in complete surgical excision of the mass.

Keywords: Lymphangioma, Lymphatic malformations, Submandibular space, Masticator space, Submandibular gland

INTRODUCTION

Cervico-facial lymphangiomas are congenital benign tumours of head and neck region. Almost 90% of cervicofacial malformations have clinical manifestation before second year of life.[1] Cases which have reported adult onset lymphangiomas usually have trauma or infection as possible etiological factors.[2] In the head neck region, commonest site of cystic lymphangioma is the posterior triangle and submandibular space. Rarely it involves the lower part of face. We report a rare case of a large cervicofacial lymphangioma (macrocystic) in a 14 year old male child, which is a higher age group in the paediatric population for cystic hygroma. It is a first ever reported case of submandibular space lymphangioma extending superiorly upto masticator and pterygomandibular space in close relation to base of skull. Our work has been reported as per guidelines of SCARE criteria.[3,4]

CASE REPORT

A 14 year old male patient hailing from Mahasamund district of Chhattisgarh presented to our out patient department with complaint of gradual onset right side cheek and neck swelling since 1 year (Figure 1A). There

was no locoregional pain, restriction in mouth opening, dysphagia, dysphonia, hoarseness or respiratory distress. The only complaint of the patient was the progressive increase in size which caused cosmetic deformity.

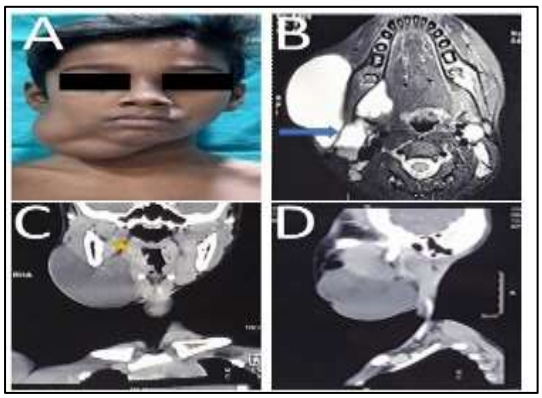

Figure 1: (A) 14 years old male with huge right sided cervico-facial swelling. (B) CEMRI of face and neck axial cuts showing cystic collection in multiple facial compartments submandibular, sublingual, masticator and pterygomandibular spaces. Facial artery (blue arrow). (C) CT scan coronal section showing non enhancing hypodense lesion extending from medial pterygoids above to myelohyoid below, pterygomandibular space (yellow asterisk). (D) CT scan sagittal section.

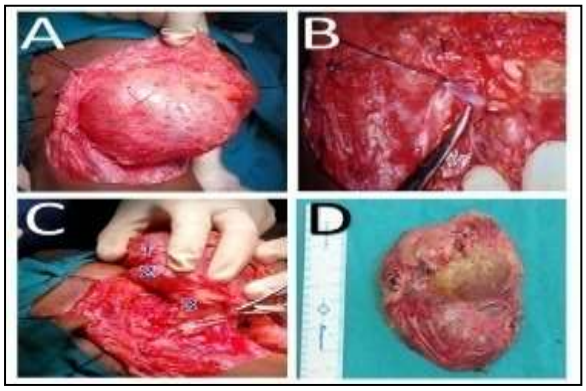

Figure 2: (A) Superior extent of the swelling upto zygomatic arch. (B) Facial artery traversing through the mass. (C) (1) and (2) parts of the swelling divided by a septation, (3) Sublingual part. (D) Entire specimen of lymphangioma excised in-toto.

On examination, there was a single globular swelling in the lower half of face and right submandibular region, size approximately 8×6 cm, skin over the swelling appeared to be normal. Superiorly extending up to zygoma, inferiorly up to the level of thyroid cartilage, posteriorly along the anterior border of the right sternocleidomastoid. Swelling was soft in consistency, fluctuation test positive and transillumination was positive. There was no bulge in the oropharyngeal wall or floor of mouth. The orifice of the right submandibular duct and the saliva flow was normal. All lower cranial nerves examination were within normal limits. Further the child was evaluated with USG neck which showed cystic lesion in right submandibular region with multiple septations with lesion measuring 4.75×3.38×4.9 cm and the lesion was seen rising from the superficial border of the right submandibular gland and facial artery and its few branches were traversing through the swelling. Cytological examination of the fluid aspirate from the swelling suggested benign lympho-epithelial cyst.

CECT neck showed a large non enhancing hypodense lesion (7.8×7.5×5.2 cm) in the right side of neck which has epicentre in the right submandibular space, extending supero-laterally up to masticator space and abutting the masseter muscle, infero-medially seen abutting and displacing the myelohyoid muscle, superiorly seen abutting the medial pterygoid muscle, inferiorly extending to the floor of the mouth and occupying the sublingual space. A claw of submandibular tissue is noted in the posteromedial aspect (Figure 1C, 1D). MRI which showed a cystic lesion extending from the medial pterygoids above to myelohyoid muscle below (Figure 1B). The treatment was planned for surgical excision of the mass under general anesthesia based on radiological and cytopathological studies. The surgical approach was made with a horizontal neck incision along the neck crease. Skin flap was elevated superiorly up to zygomatic arch and a huge cyst was noted (Figure 2A). The mass was meticulously dissected from the masseter muscle and a well demarcated plane of separation was present. The marginal mandibular nerve was identified and retracted and facial artery was seen traversing through the mass (Figure 2B) which was ligated and divided. The cystic mass had an extremely thinned out wall with multiple septations and was closely abutting the submandibular gland. As submandibular salivary gland mucocele was a probable differential diagnosis, the cyst along with the submandibular gland was excised in toto to prevent any future recurrence (Figure C, D). The specimen was sent for histopathological examination, which revealed fibro-collagenous cyst wall lined by cuboidal epithelium and endothelial cells. Immunohistochemistry stain was positive for CD 34 and CD 240 and negative for EMA, which favoured the diagnosis towards lymphangioma. There was no recurrence of swelling post six months of follow up.

DISCUSSION

Cervicofacial lymphatic malformations account for 5.6% of all benign lesions of infancy and childhood.[5] They arise from obstruction of lymphatic drainage into the jugular lymph sacs of the venous system. The cystic spaces are filled with lymphatic collection. Three different histological varieties have been described, namely, lymphangioma simplex, cavernous lymph-angioma and cystic lymphangioma (cystic hygroma /cervicofacial lymphangioma.[6,7] Ultrasonographic findings have classified lymphangioma as septate and non-septate lesions.[8]

Lymphangiomas can be diagnosed based on clinical symptoms, FNAC and radiographic appearance on MRI. Clinically, the swellings are soft, cystic in consistency and are found to occur in head, neck, axilla, groin, oral cavity and facial soft tissues.[9] They tend to grow extensively along tissue planes and have multiple outpouching sacs. Histologically the fluid contains cholesterol crystals, lymphoid cells and endothelial cells.[10] The clinical presentation is mainly based on mass effect and compression to surrounding neurovascular structures. Lymphangiomas of head neck primarily present with dysphagia, dysphonia, or airway compromise apart from a visible neck swelling.[11] Rarely, macrocystic ones may extend to involve multiple fascial compartments of head and neck which can be both disfiguring and potentially life-threatening if it extends to skull base.

MRI is an important diagnostic tool which also helps in surgical planning and determining the anatomical spread of disease. Surgical excision remains the treatment of choice for these lesions. Other options are laser surgery, cryosurgery, electrocautery, steroid administration, sclerotherapy injection, embolization and radiation therapy. Partial removal can lead to symptomatic recurrences. In selective cases, tumour reduction with CO2 or Nd-YAG laser might reduce symptoms and sclerotherapy using Picibanil (OK-432) remains an effective treatment.[12-14]

Present case was a huge swelling which involved submandibular, sublingual, masseter and pterygomandibular space and had extended superiorly up to pterygoid plates. The huge swelling caused cosmetic deformity. There was no complaint of dysphagia or respiratory distress. No history of trauma or upper respiratory tract obstruction was seen. Cranial nerve evaluation revealed no abnormality. Owing to its location in submandibular space, cystic consistency, late age of presentation, the probable differential diagnosis was salivary gland mucocele and lymphangioma. Intraoperative findings revealed that the mass was macrocystic in nature with internal septations and was closely abutting the submandibular gland, extending upto the sublingual space. Superiorly, the mass was well delineated from the masseter and medial pterygoids which revealed that the mass was extending but not involving the base of skull. The facial artery was traversing through the mass and was duly ligated. As preoperative FNAC was a lymphoepithelial cyst, the submandibular gland was excised along with the mass to prevent future recurrence. Post-operative histopathology confirmed it to be a lymphangioma and no salivary glandular elements were found. The difficulties in surgical management is due to the proximity of the swelling to vital structures like branches of facial nerve (marginal mandibular nerve), facial artery, masseter and pterygoid muscle, submandibular gland and makes complete surgical excision a challenge. The patient is on regular follow up for six months and there is no recurrence of swelling till date.

Lymphangiomas of neck and face although common below 2 years of age, can present in higher age groups and should be considered as differential diagnosis in these age groups. As the preoperative differential diagnosis was a salivary gland mucocele and a lymphoepithelial cyst, adjoining salivary gland excision was vital to prevent recurrence in future. Radiologically, the mass extended to multiple facial compartments ascending up to skull base. A wide cranio-caudal exposure from the zygomatic arch to the hyoid was necessary to access the superior extent of the facial component and prevent spillage of the contents. As first surgical intervention is the best opportunity for complete removal, meticulous surgical plan and tailor-made intraoperative steps were necessary to avoid injury to adjacent neuro-vascular structures and ensure complete excision.

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