Case Report

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Radiology Section

Efficacy of Intra-lesional Bleomycin and Sodium Tetradecyl Sulphate Combination in Management of Vascular Malformation in Head and Neck Region in a Child-A Case Report

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ABSTRACT

Vascular malformations of the head and neck region can cause cosmetic and functional disturbances. These may lead to enormous psychosocial problems for patients and their families affecting activity of daily living. We present an unusual case of large low flow vascular malformation of left cheek and upper part of neck, involving left pterygopalatine fossa and left para-pharyngeal space. Our case demonstrates the effective management by combined intra-lesional sclerotherapy of injection Bleomycin and injection Sodium tetradecyl sulphate. Multi Detector Computed Tomography (MDCT) played an important role in determining the extent of lesion, feeding vessels, nature of lesion and also in tailoring the management of vascular malformations.

Keywords: Low flow vascular malformation, Multi detector computed tomography, Sclerotherapy

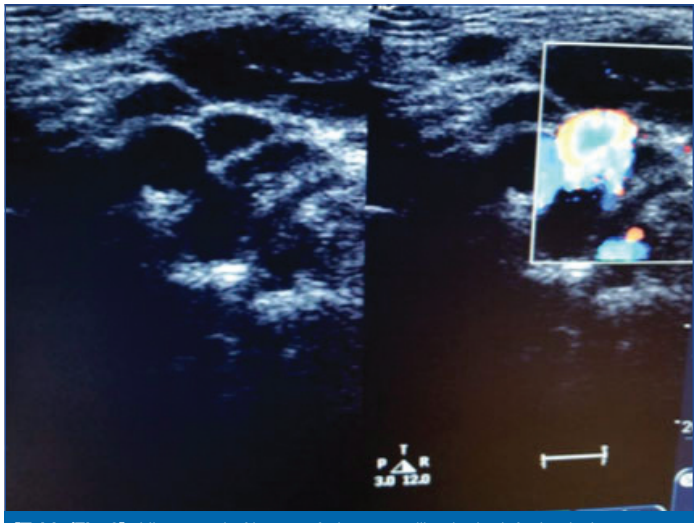
CASE REPORT

A 12-year-old female patient presented to Department of Ear, Nose and Throat (ENT) on 11th July 2018 with complaints of swelling in the left side of face and neck since 10 years, with progressive increase in size. There was no history of trauma or any previous surgeries or similar complaints in the family. The patient had received 3 doses of injection sodium tetradecyl sulphate with Injection triamcinolone acetonide under local anaesthesia intra-lesionally in October, December 2013 and March 2014, each dose two month apart in a tertiary care hospital in Bangalore, Karnataka, India, following which the lesion size had minimally reduced. The lesion started increasing in size after six months of initial therapy. On inspection there was a diffuse swelling around 7×5×2 cm, on left cheek and left side of upper part of neck and it was extending up to the: a) superiorly, at the level of lower border of tragus; b) inferiorly, up to the level of body of hyoid bone, left submandibular region; c) medially, the margin of swelling was around 2 cm away from angle of mouth; d) laterally, it was involving the concha and symba concha of pinna, external auditory canal, post auricular region with obliteration of post auricular grove and mastoid area with its tip. Intraorally, there was a medial displacement of left tonsil. On local examination, there was no local rise in temperature, tenderness was present over the angle of the mandible, and surface was smooth with few nodularities and it was immobile and non-pulsatile with no associated vascular bruit. The swelling was spongy in consistency with few nodular sub centric swellings in between [Table/Fig-1]. Systemic examination and laboratory investigations were within normal limits. Clinical diagnosis of a slow growing vascular malformation was made.

[Table/Fig-1]: Clinical profile showing extensive swelling of the left cheek. Intra orally there was a medial displacement of left tonsil.

The patient was referred to Department of Radio-diagnosis for ultrasound of the soft tissue swelling in the left cheek and Contrast Enhanced Computed Tomography (CECT) of neck for further evaluation.

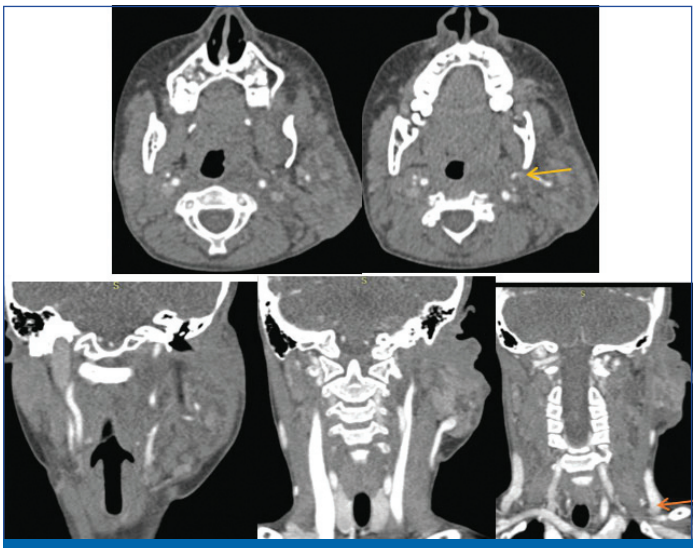
Ultrasound of the soft tissue swelling in the left cheek demonstrated a large heterogeneous echotexture lesion with multiple anechoic areas which are compressible and internal vascularity noted within these anechoic areas suggestive of vascular malformation [Table/Fig-2].



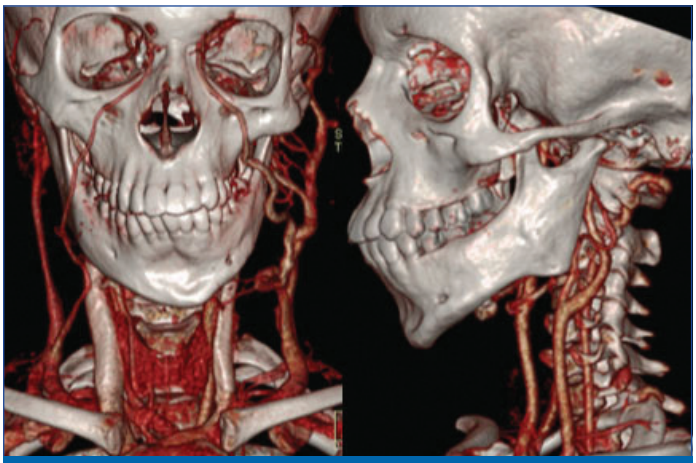
[Table/Fig-2]: Ultrasound of large soft tissue swelling in the left cheek demonstrates a large heterogeneous echotexture lesion with multiple anechoic areas which are compressible and internal vascularity noted within these anechoic areas suggestive of vascular malformation.

CECT demonstrated a large heterogeneous enhancing lesion with epicenter of the lesion in the left parotid space with extension into the masticator space involving the left temporalis and masseter muscles, pre auricular, auricular and post auricular regions, left pterygopalatine fossa. Left para pharyngeal space was obliterated and displaced medially. Anteriorly, the lesion was extending till the left submandibular gland with displacement of the tonsil medially. Scalloping and cortical thinning of the left ramus and condylar process of the left mandible was noted. Feeding vessels were noted

from the external carotid artery branches with drainage into the left subclavian vein. On delayed images, there was minimal washout. No midline extension or calcifications/phleboliths were noted [Tables/Fig-3,4].



[Table/Fig-3]: Axial and coronal contrast enhanced computed tomography demonstrates large heterogeneous enhancing lesion with epicenter of the lesion in the left parotid space with extension into the masticator space involving the left temporalis and masseter muscles. Feeding vessels were noted from the external carotid artery branches (yellow arrow) with drainage into the left subclavian vein (orange arrow).

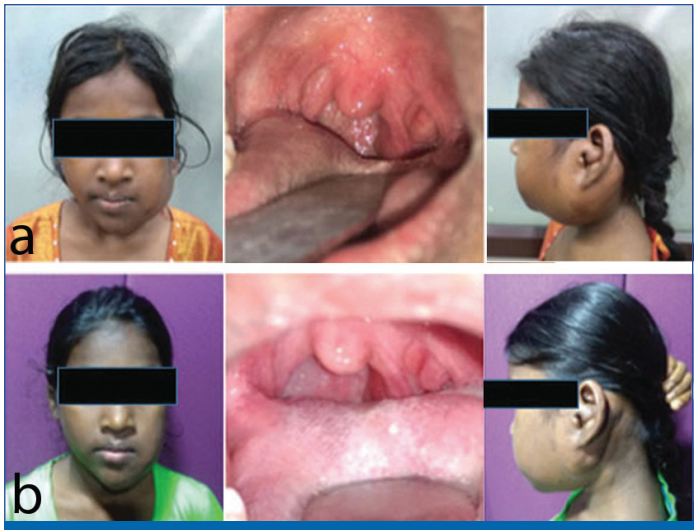


[Table/Fig-4]: Volume rendered computed tomography demonstrates the feeding vessels from the external carotid artery branches with drainage into the left subclavian vein.

Three doses of sclerotherapy under local anaesthesia with injection bleomycin 12.5 International Units (IU) and injection sodium tetradecyl sulphate 1cc, diluted with 5cc of 5% normal saline was administered, each one month apart. Initially after first dose, post procedure there was a minimal increase in the size of swelling, later there was a gradual reduction in the size. Patient was followed up regularly initially every month for three months, followed by once in three months. After one year of follow-up from the date of first dose, there was a significant reduction in the size of the swelling and there was no recurrence or increase in the size after last dose of sclerotherapy. Patient is doing fine till date without any complications or side effects post procedure [Table/Fig-5a,b].

DISCUSSION

Extracranial vascular anomalies are abnormal communication between feeding arteries and draining veins without an intervening capillary bed. These are relatively common in infancy and childhood and cover an array of pathologies. The most common site being head and neck region accounting for about 60% of cases [1,2]. Vascular malformations are usually congenital, with a propensity to grow without regression and can be devastating both functionally and aesthetically [3,4]. Mathur NN et al., for 'slow flow' lesions coined



[Table/Fig-5]: Photographs and intra oral examination of the patient a) pre and b) post sclerotherapy (day 30) demonstrating significant resolution of the soft tissue swelling in the left cheek.

the term 'haemodynamically less active' [1,5]. The pathogenesis of venous malformation is not clearly understood. It is hypothesised to be caused by developmental defects of the venous system [6]. Mulliken JB et al., classified these extracranial vascular anomalies into hemangioma and vascular malformation [1,7]. Haemangiomas appear during the first month of life and vascular malformations are always present at birth. Haemangiomas proliferate rapidly during first two years of life and regress slowly. Vascular malformations do not regress and continue to grow as the child grows. Waner M and Suen JY have classified vascular malformation depending on their predominant vasculature into slow or low flow and fast or high flow vascular lesions [8] in which lymphatic, capillary and venous malformations are grouped under slow flow vascular lesions and arterial, arteriovenous fistula and arteriovenous malformation comes under fast flow vascular lesions [1,8].

Depending upon the radiographic imaging of the draining veins, venous malformation is divided into four types [9,10]: Type I-isolated malformation without venous drainage; Type II-malformation with drainage into normal veins; Type III-malformation with drainage into dilated veins and Type IV-dysplastic venous ectasia. This classification is useful for assessing candidates to sclerotherapy. For first two types, mild sclerosants are considered like pingyangmycin/bleomycin and for last two types lesions, strong and aggressive sclerosants such as ethanol was suitable as they have a drainage into a dilated vein [1,9,10].

Surgical excision is the treatment of choice for lymphangiomas and vascular malformations but it carries lots of complications due to radical resection leading to disfigurement or dysfunction or incomplete resection [1,4]. Embolisation materials like onyx, N-butyl cyanoacrylate and polyvinyl alcohol can be used to treat extracranial vascular malformations but has a high recurrence rate [11-15]. Ethanol has been used in type III and IV lesions as it has low recurrence rates, but not used routinely as it is a toxic embolic agent causing serious complications like pulmonary hypertension, heart failure and nerve damage [15].

Intra-lesional bleomycin injection has been recently shown to be an effective treatment for vascular lesions and used extensively as a nonsurgical treatment because of its low cost, easy availability, low toxicity as chemotherapeutic agent and high sclerosing effect on vascular endothelium and an apoptotic effect on rapidly growing immature cells inducing DNA degradation [1,3].

Bajpai H et al., had reported on comparison of intra-lesional sclerotherapy with sodium tetradecyl sulfate and bleomycin in the management of low flow craniofacial soft tissue vascular malformation and found that bleomycin was more effective than sodium tetradecyl sulphate. A total of 16 patients with craniofacial

region venous malformation were divided into two groups of eight members each. Intra-lesional bleomycin was administered to Group 1 and sodium tetradecyl sodium sulphate was administered to group 2 patients. All the 16 cases were followed up for 2 to 3 years, most of the vascular lesions of group 1 resolved after first dose with no recurrence with 87.5% cure rate. In Group 2 patients, 4-6 doses with a mean of five repeated dosage of intra-lesional sodium tetradecyl sulphate was required before the lesions started to resolve and three patients showed recurrence within 2 years, with an effective response rate of 62.5% [2].

In an another study by Sachin K et al., in which retrospective analysis of 358 patients with vascular malformations showed combination of sodium tetradecyl sulphate and bleomycin injection have a cure rate of 82% to 94% depending on the size of lesion with no recurrence, with minimal to no complications [4].

Jin Y et al., conducted a pilot study on treatment of extracranial arteriovenous malformations in early stages with intra-lesional interstitial bleomycin injection. In this study out of 34 patients, 27 patients were responsive to bleomycin with complete response in nine patients. There was no response in four patients. Anaphylactic shock was a major complication in one patient. Other reported minor complications including hyperpigmentation, pruritus and bullae [15].

In our study, we used combination of injection bleomycin and injection sodium tetradecyl sulphate as it was a case of recurrence which was previously treated with injection triamcinolone acetonide and sodium tetradecyl sulphate intra-lesionally. A total of three doses were given intra-lesionally, each one month apart. At the end of 3rd dose, patient showed significant reduction in the size of the swelling with very minimal or almost nil cystic spaces within the lesion on ultrasonography. There was no local or systemic complications post treatment. Patient was followed up for one year from last dose and there is no recurrence till date.

CONCLUSION

Management of Low Flow Vascular Malformations (LFVMs) can be very much challenging, especially in growing children and must involve a multimodality approach. Combination sclerotherapy with intra-lesional bleomycin and sodium tetradecyl sulphate injections are a promising treatment option or an adjunct in children with LFVMs. This may preclude the need or morbidity associated with surgical excision, particularly in the head and neck region. MDCT is a valuable tool to know the extent and involvement of various spaces. It also aids in the vascular mapping of the malformation prior to the treatment.

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