CYSTIC HYGROMA WITH ASSOCIATED ANEURYSM OF THE COMMON FACIAL VEIN: A CASE REPORT

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ABSTRACT

Cystic hygromas (CH) are congenital malformations of the lymphatic system and consists of internal cystic areas of variable sizes. CH is commonly seen in the cervicofacial region, axilla, mediastinum, groin and abdominal viscera. CH can be asymptomatic if small however can cause variable symptoms from respiratory distress to infection. It has been associated with number of syndromes and pathologies however its association with venous anomalies and aneurysm has been rarely reported in the literature. We report a case of cystic hygroma in left submandibular region with venous aneurysm of the left common facial vein and ectatic lingual vein.

Key words: Cystic hygroma, venous aneurysm, submandibular

Introduction

Cystic hygromas (CH) are congenital malformations of lymphatic system. Cystic hygroma occurs more frequently as compared to other types of lymphangioma, and consists of single or multiple macrocystic lesions having poor communication with normal lymphatic channels.[1] Lymphangiomas are usually classified as capillary, cavernous or cystic or may be classified as microcystic (<2cm cysts), macrocystic (>2cm cysts) and mixed lymphangiomas which consists of cysts of variable sizes.[2]

CH most commonly occur in the cervicofacial regions, particularly at the posterior cervical triangle, other common sites are axilla, mediastinum, groin and below tongue. Sometimes these may occur in liver, spleen, kidney and intestine. Omental cyst and mesenteric cyst represents parallel lesions at these locations.[3]

Cystic hygroma can be associated with aneuploidic anomalies and non aneuploidic anomalies.[4] Venous anomalies are rarely seen with cystic hygromas.[5] We report a case of cystic hygroma of the subman-dibular region with venous aneurysm of the common facial vein.

Case Discussion

A 3 years old baby resident of Afghanistan presented with left submandibular soft, non tender swelling since birth. The mass was compressible and fluctuant. According to her mother it had increased in size with growth of the baby. There was no history of cyanosis, choking, fever and chills or erythema. She was advised a CE CT neck by plastic surgeon who suspected it as a case of cystic hygroma however wanted to see the relation and the extent of the lesion. CE CT done on 128 slice Toshiba prime aquilion showed a 5.2 x 4.9 x 3.5cm hypodense cystic lesion with minimal enhancement of the internal septa suggestive of cystic hygroma in the left submandibular region extending

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superiorly along the ramus of the mandible abutting left masseter muscle. No internal solid component/ calcification/ hemorrhage were seen. Medially it was extending upto the floor of mouth abutting the left submandibular gland. Posteromedially extending to the left carotid sheath. Posteriorly it was abutting the left sternocleidomastoid muscle. A dilated aneurysmal common facial vein, (Fig.1) (tributary of internal jugular vein) measuring 1.5 x 1.4 x 1.2cm, was seen within the posteroinferior aspect of cystic hygroma. Ectaticlingual vein waspassing medially to cystic hygroma. The facial vein was further seen traversing through the lesion. Further it was noted that facial and lingual arteries (branches of external carotid artery) were also traversing though the lesion. Final diagnosis of cystic hygroma with intralesional venous aneurysm of common facial vein along with facial vein, facial artery and lingual artery traversing through the lesion was given. Her baseline investigations were all normal and patient underwent surgery. Under GA transverse incision given in neck creases. The lesion resulted in thinning of the platysmal layer. The ansacervicalis dissected out safely. The dissection was tiring because of multilobulated attachments of the lesion which was tethered to the left submandibular gland with loss of intervening fat and left masseter muscle. Prominent vessels were seen traversing through the lesion as mentioned by the CT report and also depicted by per operative hand held doppler USG. All the dissected specimen along with its vascular component was sent for histopathology. Histopathology findings were consistent with radiologic diagnosis.

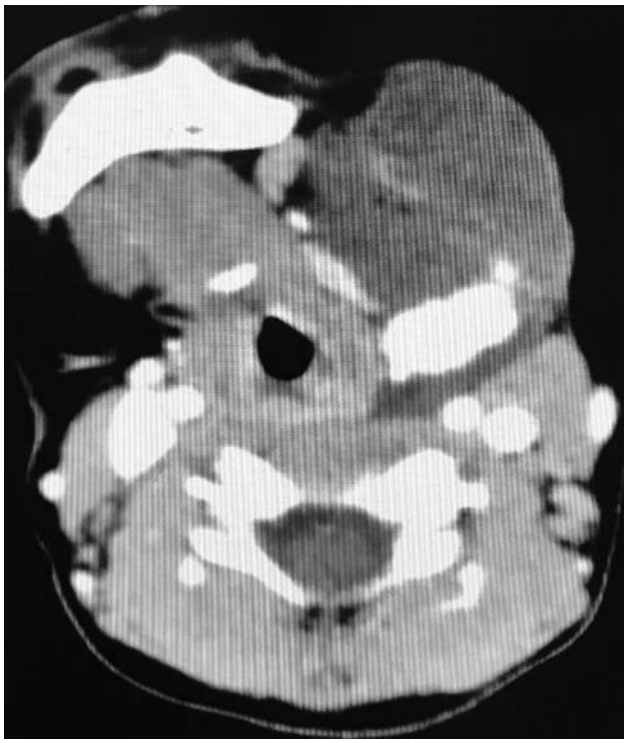

Figure 1: Axial image showing left submandibular cystic hygroma with aneurysm of common facial vein

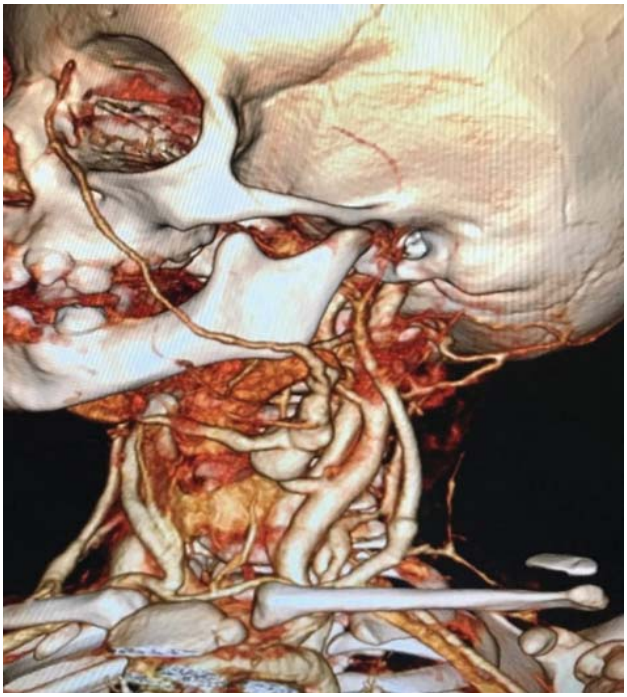

Figure 2: Showing 3D image of common facial vein aneurysm and ectatic lingual vein

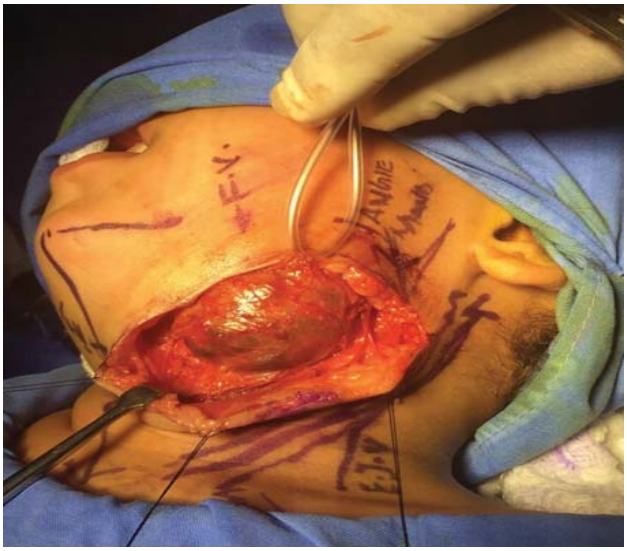

Figure 3: Per operative image

Discussion

Cystic hygromas are developmental anomalies of vasculolymphatic origin. They can arise anywhere along the lymphatic system and in most cases (80-90%) appear by the age of 2 years.[5] Enlargement of these cystic lesions is common and may compress the adjacent organs, causing complications such as respiratory distress, feeding difficulties, or vascular compromise.[6] Radiologic evaluation of the cystic hygromas includes sonography, CT, and MR imaging.[7]

The association of cystic hygromas and vascular malformations is extremely rare.[5] Joseph AE explained thateight out of 15 patients with mediastinal cystic hygroma were found to have abnormal enlargement of neck or thoracic veins. Five of these children had aneurysmal dilatation of the superior vena cava (SVC). Three more had mild enlargement of the SVC. Although the association of venous aneurysms and cystic hygromas has not been previously observed, their coexistence does not appear to be coincidental.[8] Gorenstein documented two cases of cystic hygroma associated with venous malformations.[9] Et al mentioned reported a 4-year-old girl with a known history of cystic hygroma of the neck presented with painful left neck mass enlarging in size over a period of 2 days. An enhanced CT study of the neck, revealed multiloculated cystic lesions on both sides of the neck predominantly involving left side of the neck. Vascular lesion, was seen which was concluded as originating from the left posterior facial vein.[5] In our case left sided submandibular CH with aneurysm of common facial vein was noted with facial vein traversing through the lesion. Lingual vein was also mildly ectatic. Our case was further complicated by the passage of facial and lingual arteries through the lesion.

The most preferred modality of treating cystic hygroma remains complete surgical excision; however, sclerosing agents may also be used such as bleomycin and OK 432, however OK 432 has more satisfactory results and less complications as compared to bleomycin.[1,10] The other treatment modalities include simple drainage, aspirations, radiation, laser excision, radio-frequency ablation and cauterization. Surgical excision of the complex cystic hygromas, involving deep and vital structures, is not an easy task.[10] In our case the surgery was done by a vigilant plastic surgeon who examined the case and discussed it with radiologist before performing surgery. Extreme care has to be takento avoid per-operative complications, such as damage to facial nerve, facial artery, carotid vessels, internal jugular vessels, thoracic duct and pleura and incomplete excision in case of infiltration to the surrounding structures. In about 20 % of cases, there is recurrence even after apparent complete excision of the lesion.[1]

Conclusion

Cystic hygroma is a common and manageable lesion in paediatric population. Knowledge of aneurysmal veins may be helpful to the surgeon planning resection of a cystic hygroma or can result in catastrophic results for the patient and surgeon if no prior knowledge is known. Association of cystic hygroma has not been well established with venous aneurysms or vascular anomalies but cases are reported in the literature and with advanced imaging such as doppler USG and CE CT / MRI, might bring more cases into light.

Conflict of interest: None

Disclaimer: The case report has been presented in the 21st ACOR conference as electronic poster

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