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Aneurysmal Dilatation of Right Internal Jugular Vein – A Rare Case

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Abstract:

Internal jugular phlebectasia (IJP) is a rare disease in which there is a fusiform dilatation of internal jugular vein, usually presents as an intermittent neck mass in children. Accurate diagnosis can be made from history, physical examination, ultrasonography and CT Angiography. We report a case of a 10-year-old boy with history of swelling appearing on right side neck only on straining, coughing or on valsalva. Diagnosis of IJP was made after doing ultrasound (USG). Because of its rarity, this entity is frequently ignored or misdiagnosed. IJP should be kept as a differential diagnosis when a patient comes with intermittent neck swelling.

Keywords: Aneurysmal dilatation; Internal Jugular Vein; Phlebectasia

INTRODUCTION

Internal jugular phlebectasia (IJP) is a rare congenital variation often diagnosed during childhood.[1-2] It occurs either unilateral or bilateral affecting the internal jugular vein. It usually presents with a benign swelling over lateral side of neck on the affected side, seen on exertion. Valsalva maneuver is most important and useful clinical sign used to diagnose the dilatation of internal jugular vein.[2-4] The differential diagnosis for a neck mass that increases in size with valsalva include laryngocele, cystic hygroma, external laryngeal diverticula, superior mediastinal cysts. We present a rare case of phlebectasia of internal jugular vein in a 10-year-old boy.

CASE REPORT:

A 10-year-old child comes with a right sided intermittent neck swelling for last 2 years. The swelling was intermittent in nature, insidious in onset, initially small in size and it gradually increased to its present size. The swelling was not prominent on right side unless the patient is asked to strain. The swelling became prominent on coughing, straining and performing Valsalva maneuver. The swelling continued to appear till the child was straining and got back to normal once child stopped doing the maneuver. There is no history of pain or difficulty in swallowing or difficulty in breathing. There is no history of change in voice.

The child was absolutely normal when the child was not straining. However, on Valsalva, a diffuse large swelling was noted on the right-side neck, seen right from below the angle of mandible to the clavicle and was palpable. The swelling measured 6x2 cm, was soft and compressible. There was no local rise of temperature. It was non tender. Cardiovascular and respiratory examination were within normal limits.

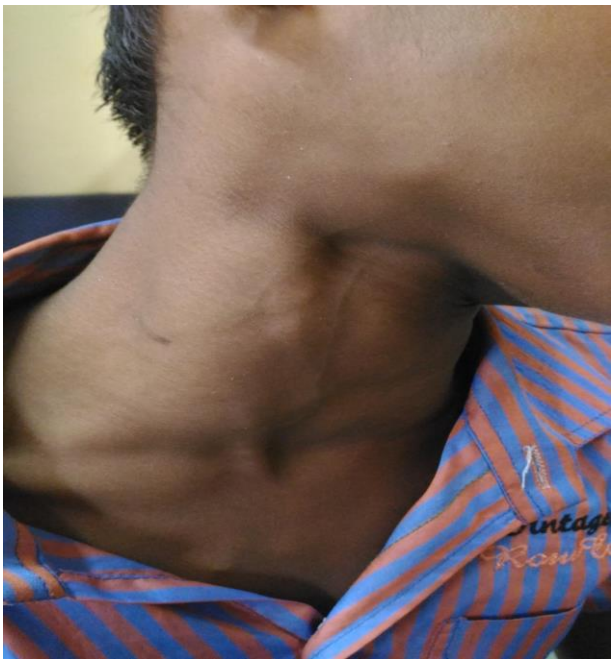

Figure 1: Clinical image showing a diffuse large swelling on the right side of neck

Chest x ray was normal. X ray neck revealed no widening or air at the region of the mass thus excluding laryngocele. The differentials could be Internal jugular phlebectasia, cystic hygroma, laryngocele. Fibreoptic laryngoscopy (FOL) was done and revealed all structures within normal limits. Fine needle aspiration cytology (FNAC) was done. However, smears showed only red blood cells, suggesting possibility of a vascular lesion.

Ultrasound with doppler confirmed the diagnosis. A 4.1x4.1x2.1 cm swelling was seen lateral to the right common carotid artery. Sluggish flow was seen within compressible swelling. Aneurysmal dilatation of internal jugular vein was seen. CT Angiography of neck was done. There was significant differential fusiform dilatation of right IJV in its entire course with no definite stenosis or abnormal connection, suggesting the diagnosis internal jugular phlebectasia.

The child was asymptomatic and there has been no significant increase in size of swelling for the last 2 years. Also, there were no complains of hoarseness, stridor or dysphagia so decision of conservative follow up was taken. The child was kept on follow up for 6 months and he had no increase in size of swelling of the neck and no complications whatsoever.

DISCUSSION

IJP is a rare and benign condition occurring as a congenital variation. About 50 cases have been reported so far in literature.[1-4] It is usually diagnosed during childhood. Because of its rarity there have been cases that go unrecognized. The exact etiology remains unclear because of sporadic reports of venous ectasia.[2-5] The possible causes are gross anatomic abnormality, mechanical compression or trauma, congenital structural defects in the wall of vein or idiopathic.

Phlebectasia and Varix must be differentiated. Phlebectasia indicates abnormal outward dilatation of vein without tortuosity, however varix implies dilatation plus tortuosity. Ectasia can affect any neck vein especially internal jugular vein, external jugular vein, anterior jugular vein, superficial communicans.

Histologically it shows loss of elastic layer, hypertrophy of connective tissue and focal intimal thickening. IJP is usually benign and asymptomatic. The most useful clinical sign is Valsalva.[2-4] During a Valsalva, diameter of affected vein may increase up to 2.2 times compared with its measurement at rest. Men are most affected in the ratio 2:1 to 3: 2. It is more commonly seen on the right side. The possible differential diagnosis includes IJP, laryngocele, AV malformations, cavernous hemangioma, branchial cyst and cystic hygroma.

Laryngoscopy directly rules out the possibility of a laryngocele and a thoracic CT scan rules out the possibility of a mediastinal cyst or tumour. Colour

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Doppler at rest and during valsalva maneuver is the investigation of choice.[2-4] It defines the extent of the lesion and its relationship with the surrounding structures in the neck. Colour Doppler imaging confirms the presence or absence of blood flow and thrombus formation in the lumen of the vein and is the gold standard for diagnosis of jugular vein phlebectasia.

In literature, the management is described as both conservative and surgical by different authors. Since IJP is not known to progress rapidly and there are no instances of rupture of swelling or other serious complications, a conservative approach can be adopted with close follow up.[3-7] Also ligation of jugular vein may produce effects of venous congestion in a small subset of patients resulting in cerebral oedema.

Surgical management has been advocated by some for few reasons.[5-6] Since hemodynamics in dilated vein are markedly changed, it is prone to form intramural thrombus. Secondly involved vein has a potential to rupture. Hence "ligation and resection of dilated segment of vein" can be done as per few authors.[4-7]

We went for a conservative approach owing to its benign nature and no complications in the child. Reassurance to the child was given and close follow up was advised. After a follow up period of 6 months, swelling in the neck was of same size and there were no complications.

CONCLUSION:

IJP is not known to progress rapidly and no instances of rupture of swelling or other serious complications have been seen. There is some conflict of interest regarding management of IJP. Though some advocate surgical management of phlebectasia which is ligation and resection of dilated segment of vein. Some are of the opinion that IJP usually does not require any active intervention. Only reassurance to patients owing to benign nature of phlebectasia is required.

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