Case Report

Unusual case of focal neck swelling: Phlebectasia of internal jugular vein with intracranial extension

Virender Malik, Abha Kumari, TVSP Murthy
Departments of Radiology, ENT and Anaesthesiology, Military Hospital Gwalior, Morar Cantonment, Gwalior, Madhya Pradesh, India

Abstract

Internal jugular vein (IJV) phlebectasia is rare in occurrence and is frequently misdiagnosed and managed inappropriately. It commonly presents as a unilateral neck swelling which typically increases in size with valsalva maneuver. Although, the most common cause of a focal neck swelling, which increases in size with valsalva maneuver is laryngocele, the possibility of phlebectasia of IJV should always be borne in mind, especially in child. Owing to the rarity of this condition, a high index of suspicion is required to recognize the same and managed appropriately. We present a case of phlebectasia of the right IJV with intracranial extension and discuss its management. The case is being reported in view of its clinical rarity (the intracranial extension being extremely rare) and to highlight the available management strategies.

Key words: Intracranial extension, internal jugular vein, phlebectasia
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Introduction

Internal jugular vein (IJV) ectasia commonly presents as a unilateral, soft, compressible neck swelling, which typically increases in size with straining, valsalva manoeuvre, sneezing, bending or coughing. The condition is rare in occurrence and is frequently misdiagnosed and managed inappropriately. Although, the most common cause of a focal neck swelling which increases in size with valsalva maneuver is laryngocele, the possibility of phlebectasia of IJV should always be borne in mind, especially in child. Although the exact etiology is still controversial, many hypothesis have been proposed.1 Due to the rarity of this condition, a high index of suspicion is required to recognize the same. We present a case of phlebectasia of the right IJV with intracranial extension and discuss its management. The case is being reported in view of its clinical rarity (the intracranial extension being very rare) and to highlight the available management strategies.

Case Report

An 8-year-old boy, presented with intermittent swelling on the right side of the neck for last 2 years. The swelling was insidious in onset, gradually progressive with the prominence noted on speaking for longer duration, crying or straining. There was no history of change in voice, chronic cough, difficulty in breathing or swallowing, trauma or previous surgery.

Physical examination at rest revealed a small soft tissue swelling on the right side of the lower third of the neck, anteromedial to the sternocleidomastoid muscle. The swelling markedly increases in size and was fusiform in shape on performing valsalva maneuver.1 The swelling was soft, compressible, nontender, nonpulsatile and non transilluminant. Fibreoptic laryngoscopy revealed no abnormality in larynx and hypopharynx.

Radiograph of the neck showed a well-defined, soft tissue swelling on the right side of the neck with no evidence of air, calcification or phlebolith within. Ultrasonography of the neck showed mild prominence of the right internal jugular vein at rest, which increased markedly on performing valsalva. Marked turbulence of the flow was seen both on color Doppler and spectral Doppler waveform.
Contrast-enhanced computed tomography (CECT) neck showed mildly dilated right IJV at rest, markedly increasing in size with valsalva maneuver. The focal fusiform swelling on valsalva measured 24 and 35 mm as max. AP and transverse diameter respectively [Figure 2a]. The swelling extended from C4 vertebral body level to clavicle for cranio-caudal length of 7 cm [Figure 2b]. However, on performing valsalva maneuver, the entire right IJV [Figure 2c], ipsilateral sigmoid and transverse sinus [Figure 2d] were noted to be very prominent, compared to the normal left side. Screening CECT of the brain revealed no additional abnormality.

The cardiovascular examination was essentially normal and the patient was otherwise asymptomatic. The parents were explained about the disease, reassured, nonsurgical treatment offered and put on regular follow-up with advise of review at the earliest if there is any change in size of swelling and development of any symptoms.

**Discussion**

Internal jugular ectasia was first described by Zukscherdt[1] and subsequently characterized by Gerwig. Phlebectasia of the IJV has been described in the literature by various terms, including, venous aneurysm, venous cyst, aneurysmal varix and venectasia.[2] The term phlebectasia indicates abnormal outward dilatation of the vein without tortuosity and differs from the term varix, which implies dilatation plus the tortuosity.[2] Many hypotopian have been proposed as a cause for phlebectasia, including increased scalenus anticus muscle tone,[3] anomalous reduplication of the IJV,[1] compression of the vein between the head of the clavicle and the cupola of the right lung, trauma and congenital origin.

Histopathological studies have shown loss of the elastic layer and the hypertrophy of the connective tissue with focal intimal thickening.[4]

Typical clinical presentation is a child with soft, round or fusiform neck swelling located in the lower third of the neck, at the anterior border of the sternocleidomastoid muscle that increases in size with straining, coughing, bending, sneezing, valsalva manoeuvre, or after exertion.

The phlebectasia is seen more commonly on the right side because the right innominate vein lies in contact with the right apical pleura. Therefore, any increase in the intrathoracic pressure could be directly communicated to the right IJV. The left vein, being placed more medially, is not subjected to such stress.[5] Furthermore, the right IJV valves are placed at a higher level than the left sided valves. The valves play an important role in preventing retrograde blood flow. Valves are almost never seen in the right brachiocephalic vein, but the incidence of the valves in the left brachiocephalic vein are 4–8%, most being competent.[2]

The differential diagnosis for similar neck swelling includes a laryngoecele, branchial cyst, cystic hygroma, cavernous hemangioma and superior mediastinal cyst.[6] The thyroglossal duct cyst, dermoid cyst, bronchogenic cyst, cervical adenitis and metastatic adenopathy also needs exclusion.

An association of this condition with Menkes disease has been suggested.[7] The only complications reported to date are thrombosis, Horner’s syndrome,[4] congestive cardiac failure and massive hemorrhage secondary to trauma.[8] The spontaneous rupture has never been reported.

Ultrasonography, and color Doppler are the investigation modalities of choice. Ultrasonography is the best screening method. However, CECT gives exact extent of the venous abnormality, specially the details of intracranial extension, as in our case.

No treatment is indicated for this benign self-limiting condition in asymptomatic or patients who have minor symptoms.
Treatment is recommended in patients who have phlebitis, thrombus formation, Horner’s syndrome, intractable cough or for cosmetic reasons. The various treatment options are regular follow-up, ligation of the affected vein, resection of the dilated segment, sheathing of the affected segment in a poly tetra fluoro ethylene (PTFE) tube graft. Some authors believe that ligation of the jugular vein may produce effects of venous congestion in few patients resulting in cerebral edema.

Jugular vein ligation is too radical procedure for such a benign condition, and this definitely cannot be applied in cases with bilateral affliction. In symptomatic patients or when complications are present, resection of the dilated segment or covering with muscular segment is also recommended. (Some authors believe that ligation of the jugular vein may produce effects of venous congestion in few patients resulting in cerebral edema.)

Potency of contralateral IJV should be confirmed before excision in cases of unilateral lesion. The whole affected vein can be sheathed in an 8 mm PTFE tube graft from the site of its emergence to the point where it disappears behind the right sternoclavicular joint.

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