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Internal Jugular Phlebectasia; A Challenging Neck Mass in Children

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Abstract

Cystic neck masses are uncommon in children, and a minority of them are soft. Internal jugular vein phlebectasia (IJVP) is a rare cause of soft neck cysts. It presents usually as a unilateral soft neck mass of changing size. In most cases it is unilateral, right sided and predominantly in males. Imaging study modalities are diagnostic and helpful for observation and follow up. In the vast majority of cases, it decreases in size with time requiring only conservative treatment. However, surgery might be necessary in large or complicated phlebectasia. IJVP is underdiagnosed or misdiagnosed especially in pediatrics, with few cases documented in medical literature. To improve awareness of presentation and management-plan of this rare case, the authors present an extremely rare case of huge left internal jugular phlebectasia in a 14 year-old boy worsened and complicated over years of wait and see approach that needed surgical treatment.

Introduction

Jugular vein ectasia is a localized fusiform dilation of the jugular venous system, usually the internal jugular vein. It is usually on the right side; affects males twice more than females and presents early in life. It is usually of unknown origin. The incidence of IJVP is rare; 247 cases have been reported in total in medical literature;150 cases of them are children. (1) due to this scarcity, no clear etiology or pathophysiology could be universally agreed on (2). Rossi A et al. (3) and Yu-Tang Chang et al. (4) believe that the most likely cause for this pathological entity appears to be a congenital defect of the internal jugular vein wall structure which leads to gradual decrease in the elasticity of the venous wall. Other congenital etiology like increased scalenus anticus muscle tone, anomalous reduplication of the IJV, compression of the vein between the head of the clavicle and the cupola of the right lung have been hypothesized as causative or contributing factors for the development of IJVP. (5) El Fakiri et al. hypothesized that internal jugular phlebectasia develops as a result of the superior vena cava

hypertension during inspiration, while Paleri et al believes that the distribution of the valves in the superior vena cava or anomalies of the vein wall is the main etiology. Some acquired cases of adult-hood phlebectasia developed as a form of post traumatic sequela, for example in the case of accidental clavicle fractures. (6,7,8). In addition, association with Neurofibromatosis Type 1 and Ehler-Danlos Syndrome have been documented in the literature. Histopathologically IJVP show a normal venous wall structure. Smooth muscle and elastic fibers disarray have been reported in a minority of cases. (9)

The authors here believe that IJVP is a multifactorial abnormality; in addition to the possible congenital defect in vein wall elasticity as proposed by many authors, there is a constant rule to increased venous pressure due to anatomic reasons like clavicle fractures and upper mediastinal tumors, or due to habits or jobs associated with severe chronic straining.

diagnosis of this rare cause of childhood neck mass is a big challenge to the pediatrician and needs a high level of clinical awareness of its typical clinical manifestation as a soft, nontender, nonpulsatile neck mass with changing size (enlarges by Valsalva maneuver or straining and vanishes rapidly at rest). These clinical characteristics help to differentiate it from other causes soft neck lumps. JVP is a fusiform dilatation unlike venous aneurysm which is acquired segmental saccular fixed dilatation in adults. The term phlebectasia indicates abnormal outward dilatation of the vein without tortuosity and differs from the term varix, which implies dilatation plus tortuosity.

The differential diagnosis should include: cystic hygroma, branchial cyst, laryngocele, cavernous hemangioma, dermoid cyst and superior mediastinal mass.

Diagnosis can be confirmed by imaging study. Doppler ultrasonography (USG) of the neck is the gold standard diagnostic investigation in the developing countries. Chest and neck computerized tomography with contrast, CT- angiography, or MRI give better diagnostic accuracy and help to

Disease	Main symptoms	Site	Incidence
CIJP	Soft, compressible, painless, more evident under straining. Usually asymptomatic, voice change, slight discomfort, or pain during deglutition	Lateral, anterior to the sternocleidomastoid muscle	Non-reported
Laryngocele[4]	Compressible mass that increases in size with intralaryngeal pressure (external). Voice change, hoarseness, airway obstruction, hoarseness, foreign body sensation, or asymptomatic	Upper lateral	1:2.5 million
Hemangioma[5]	Red or bluish soft mass. Usually asymptomatic	Variable	1.64:100
Lymphatic malformation[6]	Soft mass. Asymptomatic or symptoms associated with airway obstruction	Variable	1:250–4,000
Branchial cleft cyst[7]	Cystic or tender mass. Pain, dysphagia, itching skin, or asymptomatic	Lateral, anterior board of the sternocleidomastoid muscle	1:1 million
Superior mediastinal cysts and tumors[8]	Airways obstruction, dysphagia, venous return obstruction, symptoms related to the underlying disease	Superior mediastinum	1:769,000–100,000

exclude intrathoracic masses. These imaging modalities are helpful to exclude complications and for evaluation before surgical intervention. (10,11)

Due to its benign nature, treatment is usually conservative. Long term clinical observation and serial USG comparative measurements are highly recommended. most case regress spontaneously. However, massive enlargement and longitudinal extension may occur which might lead to a considerable cosmetic and psychological distress.

Complications like dysphagia, dysphonia, Horner syndrome, thrombosis, and rupture of the vein wall after minor trauma are possible. Such events necessitate surgical treatment after accurate evaluation to exclude any laryngeal and mediastinal etiology. (12,13,14)

Case presentation

A 14 years old boy presented to the outpatient pediatric clinic with the chief complaint of a left sided neck lump increasing in size over the last 5 years. It was non-painful and caused no discoloration of the overlaying skin. The volume and intensity of this lump increased during cough, sneeze, shouting and similar activities. The left sided neck mass measured around 3cm at rest, 5 cm with speaking/shouting and 7cm with valsalva maneuver, respectively.

Palpitation showed a soft pressable fusiform swelling on the left side of his neck, anterior to the sternocleidomastoid muscle, extending to the anterior triangle of the neck and the left clavicle. On Auscultation there was no palpable thrill or audible bruit. The regional lymph nodes were not enlarged. The Patient suffered no orthopneic dyspnea.

It is worth noting that the contralateral right sided internal jugular vein appeared to show similar but significantly smaller phleboectastic changes with maximum diameter of 1,5 to 2cm. The rest of the physical exam, including oropharyngeal and systemic examination, were completely unremarkable Fig1 (a, b).

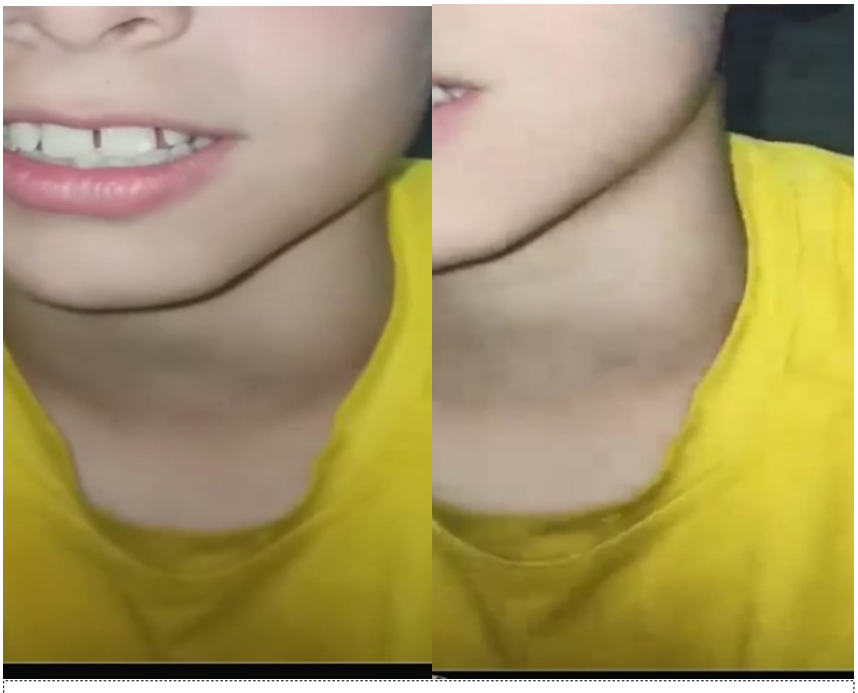

Figure 1. (a Left, b Right)
(a) obvious neck mass on regular speech, (b) huge mass during Valsalva maneuver

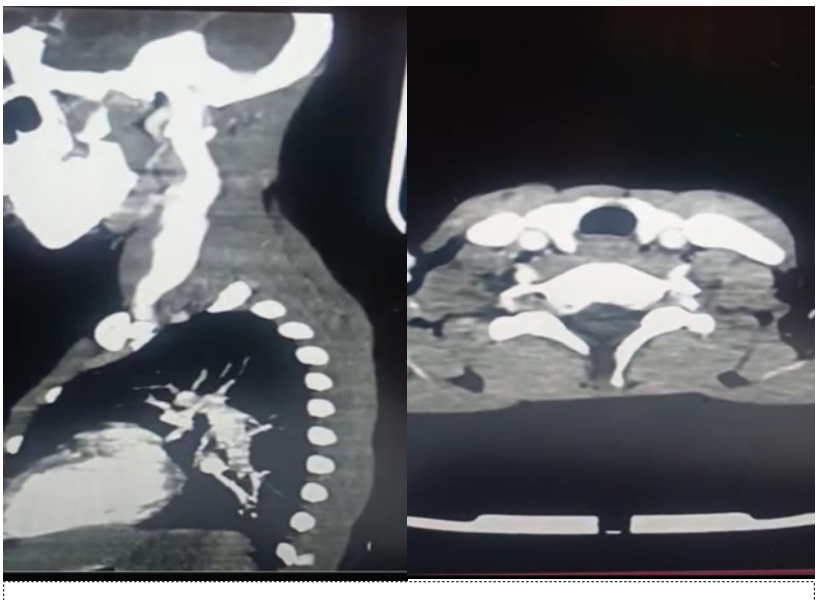

Figure 2. (c Left,d Right) (c) sagittal CT-image shows well-defined (d) axial CT-image showing dilated left IJV with homogenous huge left IJV

Recently his voice quality and tone changed with development of hoarseness as well as increased speech-effort.

3 years ago he was examined and reassured; unfortunately, his lump has grown considerably in size and become psychologically distressing and obvious even with regular speech. The Patient displayed no fever, dyspnea, weight loss or cough. In his past medical history 2 years ago, the patient suffered episodic gross hematuria that was as attributed to benign urethrorrhagia of childhood. His younger sister had a neck surgery at age 2 years for branchial cyst with complete recovery. In addition, one sibling has died at age of 18 months as a result of a complex congenital heart disease.

Ultrasonography (USG) of the neck showed fusiform dilatation of the lower segment of the IJV with a caliber of 4cm increasing to 7cm on Valsalva maneuver. In a previous USG imaging 3 years ago, it was about 2cm and 4 cm consecutively. Contrast-enhanced computed tomography (CT) of his neck and mediastinum excluded any mediastinal mass or anomaly and confirmed the previously known dilatation of the left internal jugular vein without thrombosis (Fig2).

Referral consultation was arranged for evaluation by as vascular surgeon. considering the remarkably growing dilatation complicated by dysphonia (hoarseness and increased work of speech), the vascular surgery team recommended a surgical intervention. The exact surgical technique was kept undetermined at this point as intraoperative evaluation of the anatomy is essential for the choice of correct technique.

Under general anesthesia, incision along the anterior border of the left sternocleidomastoid was made, the IJV was seen and isolated, compressible dilatation as mentioned above was seen with intact wall without aneurysm nor thrombosis. keeping in mind the child age, bilateral internal jugular vein dilation, and anatomical nature intraoperatively, vein encapsulation appeared the preferred method rather than vein ligation or resection with end to end Anastomosis. The left internal jugular vein was isolated along its path, longitudinally constricted and wrapped with a Dacron graft along its path to keep a diameter of 10 mm. Fig3(f,g)

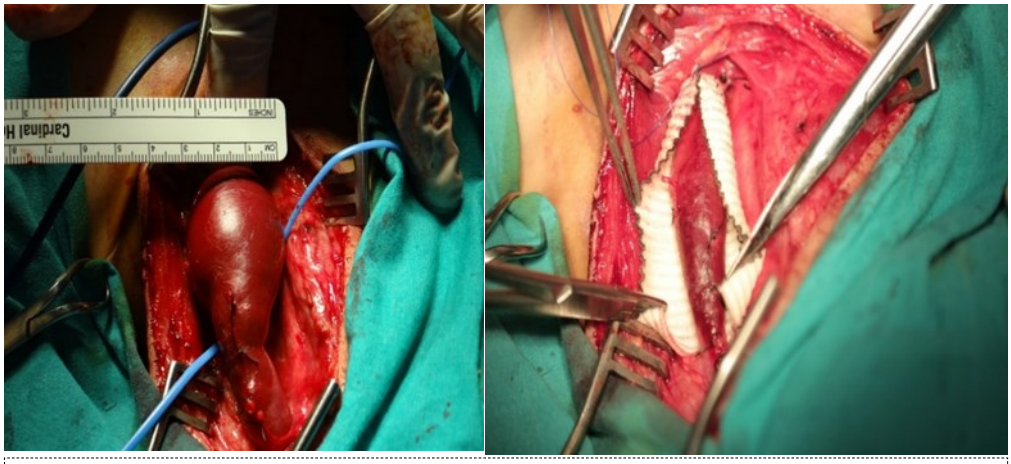

Figure 3. (f) intraoperative photograph shows a fusiform compressible dilatation IJVP. (g) intraoperative photograph shows the left vein wrapped in Dacron tube graft.

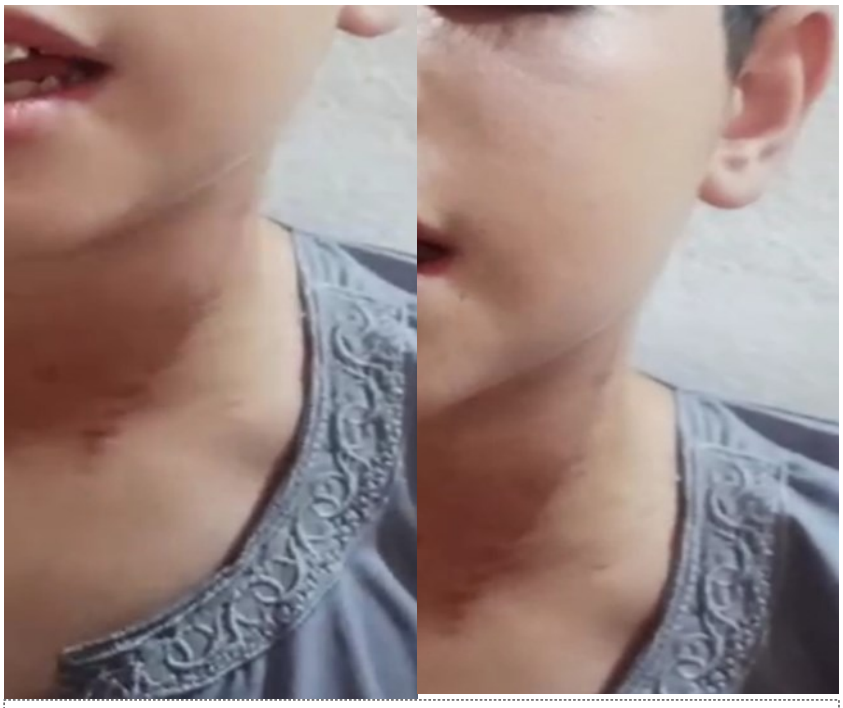

Figure 4. Complete recovery of neck mass with and without straining.

No drainage or anticoagulant were needed, and he was discharged home next day. On follow up visit one month later, the child was healthy and asymptomatic with complete resolution of voice hoarseness, speech effort as well as neck swelling even with Valsalva maneuver Fig4.

Discussion

67% of neck masses in children are solid, 27% of them are cystic, and 5% are mixed. (11) Clinicians are familiar with solid neck masses which are mostly infectious or neoplastic in origin, but it is not uncommon to see a child with a cystic lesion. Neck cysts in pediatrics are mostly congenital or developmental in origin; the most common cystic lesion is thyroglossal cyst, followed by branchial cyst; other less frequent causes include (cystic hygroma, ranula, epidermoid cyst, laryngocele, cervical bronchogenic cyst hemangioma, thymic cyst, and phlebectasia).(11,15)

Neck abscess is usually surrounded by severe inflammation, firm, extremely painful, and can be recognized from neck masses. however, it can be a complication of an infected latent neck mass or cyst (16,17). Thus, neck abscess in children need a specific evaluation before any surgical intervention. neck ultrasonography and fine needle aspiration have been recommended by many authors as the best diagnostic investigation for cystic neck masses (18).

This approach could be true in solid masses. on the contrary, USG alone may show only the tip of the iceberg when used to evaluate a neck cyst associated with intrathoracic extension or primary mediastinal mass. furthermore, needle aspiration could be hazardous in hemangioma or phlebectasia. (10,19)

Phlebectasia may affect any vein, and is rarely symptomatic. Internal jugular vein phlebectasia (JVP) most commonly presents in childhood and adolescent, age rang (7months-15years), mean age of 7.5 years. In 85% of cases the right side is involved (13). This entity presents as soft compressible fusiform neck mass with intermittent rapid increase in volume during straining, Valsalva maneuver, cough, and sneeze. It is usually nonpainful, nontender, non-pulsatile, and exhibits no audible bruit (16). Cosmetic impact is usually the only complain. A minority of complicated cases may have Horner syndrome, voice hoarseness, dysphagia, thrombosis, phlebitis, or vein rupture (20).

Jugular system phlebectasia is usually benign, but might be associated with intrathoracic and /or intracranial extension without any central nervous system involvement features. (2). Differential diagnosis may include all types of neck cystic masses as mentioned above. However, the above emtnioned unique clinical features and its changeable size with straining, crying, or Valsalva maneuver shrink the possibilities to very few cases: laryngocele ,superior mediastinal herniation, and cystic hygroma.(15)

Dopler sonography imaging, simple chest x ray, and laryngeal assessment are usually more than enough as a primary investigation plan to confirm the diagnosis and evaluate mediastinum. Fine needle aspiration should be avoided. MRI or CT with contrast give complete view of the vein dilatation extension, dimensions, and its relationship with surrounding organs and mediastinal structure. It is recommended in huge deteriorating phlebectasia ,symptomatic cases, and before surgery (10,19).

Since IJVP is benign, conservative observation with regular clinical and imaging follow up tests is advised (13). The patient should be instructed to avoid risky activities which might lead to injury, bleeding, thrombosis, and infection.

Surgery is indicated for complicated cases and in cases with large-sized Ectasia for cosmetic reasons (1). Surgical options include:

a. ligation of the jugular vein (endovascular such as transcutaneous cervical or trans axillary ligation, or open surgical),

b. longitudinal constriction/reconstruction suture venoplasty plus encapsulation/sheathing of the affected vein in a polytetraGuoroethylene graft.

c. primary resection of the affected dilated venous section with direct end to end Anastomosis.

In small number of cases ligation might be complicated by brain venous oedema due to venous drainage obstruction of the cerebral venous system, this is especially risky the case in Ligation of bilateral internal jugular Vein Phlebectasia, or occassionally in isolated right jugular venous ligation, as the right jugular vein is responsible for 60-70% of the cerebral venous Drainage (6, 7, 8). Thus, in Patients with bilateral IJVP, ligation of the IJV is contraindicated, and venoplasty and encapsulation are

more favored (9, 13, 20).

the method of constriction suture venoplasty plus encapsulation has been preferable in last two decades due to preservation of normal physiology and flow through the IJV. In addition, this surgery also represents the only choice for bilateral internal lesions. Both Dacron cloth and the PTFE artificial vessel patch are suitable for strengthening the venous wall (11, 13).

In instances where full mobilisation of the IJV as well as easy achievement of proximal and distal vascular control, surgical treatment with direct primary resection with end to end Anastomosis can offer another effective and safe surgical treatment technique, which similar preserves the physiological function of the IJV (21).

To determine the best surgical procedure among these various options, further studies will have to be conducted with longer-term follow-up.

Conclusion

Soft neck masses in children are uncommon and a source of diagnostic challenge. Internal jugular phlebectasia is a rare type of them with characteristic features. It increases in size during crying, straining and Valsalva maneuver. Diagnosis of this case needs high level of clinical suspension; and can be confirmed by Doppler ultrasonography and chest x ray. Needle aspiration should be avoided. It is usually a benign condition. However, it may extend to intracranial or intrathoracic veins. MRI and CT with contrast are recommended in complicated cases and before surgery. Conservative treatment is the norm as most cases regress spontaneously. Surgery is recommended for complicated cases and for cosmetic and psychogenic reasons. The method of constriction suture venoplasty plus encapsulation has been preferable in last two decades due to preservation of normal physiology and flow through the IJV. To determine the best surgical procedure among these various options, further studies will have to be conducted with longer-term follow-up.

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