Jugular Phlebectasia: Clinical Scenario in India

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Abstract Jugular phlebectasia is fusiform dilatation of internal jugular vein seen in children. It usually occurs in children but can occur at any age. It usually presents as unilateral swelling in the neck found during coughing and shouting. It is usually asymptomatic, so it is often ignored or misdiagnosed. Differential diagnosis include laryngocele, cavernous hemangioma, cystic hygroma, branchial cyst. Treatment is usually conservative as complications like thrombosis and horner's syndrome are very rare. Non invasive investigations like Doppler sonogram and are usually done to diagnose this condition. We report two cases of jugular phlebectasia, who presented to our out patient department with history of swelling in the right side of neck which increases on crying and shouting. Doppler sonogram and ultrasound was done to diagnose the condition. There seems be lack of knowledge among medical fraternity in various specialities. So a questionnaire was prepared to assess the knowledge among resident doctors, general practitioners, Otorhinolaryngologists, paediatricians. Questionnaire was posted in social networking sites and the feedback was obtained. Out of the 150 interns in our hospital, only one (0.66 %) was able to explain about the condition. In general practitioners group containing 369 respondents, only 4 (1.08 %) knew the condition and have seen patients. A pediatricians group containing 312 respondents only 23 (7.37 %) pediatricians knew the condition, 289 (92.62 %) persons had no idea about the

treatment. Otorhinolaryngologists group containing 328 respondents. 21 (6.40 %) knew the condition, 293 (89.3 %) have no idea. The Chi square test shows that the specialty of the respondent like pediatrician and ENT practitioner have more knowledge than the other for jugular phlebectasia at $p < 0.0001$.

Keywords Jugular phlebectasia · Jugular ectasia · Laryngocele · Cyst

Introduction

Phlebectasia is a term which denotes abnormal saccular or fusiform dilatation of veins [1]. Internal jugular phlebectasia is fusiform dilatation of the jugular vein which is thought to be of congenital in origin. It usually occurs in children, but can occur at any age and affects both genders. It usually presents as unilateral neck swelling which increases on valsalva manoeuvre. It is usually asymptomatic, so it is often ignored or misdiagnosed. Differential diagnosis include laryngocele, cavernous hemangioma, cystic hygroma, branchial cyst. Treatment is usually conservative as complications like thrombosis and horner's syndrome are very rare. Non invasive investigations like Doppler sonogram and ultrasonogram are usually done to diagnose this condition.

Case 1

A 6 year old boy was accompanied by his mother with history of swelling in the right side of neck for 6 months, found during crying (Fig. 1a). There was no history of trauma. Physical examination was normal at rest but showed a soft, non tender compressible swelling of size 3×3 cm in

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Fig. 4 Child at rest and during valsalva

2a No of cases seen

(a)1–2 (b) 2–5 (c)more than 5

Have you ever seen a operated case?

Have you seen any complications associated with condition- yes or no. If yes,

(a) Rupture (b) Horner's syndrome (c) Thrombosis

Results

We enquired 150 interns in our hospital, all of them participated in the study and the questionnaire was posted in a social networking site. General practitioners group containing 932 members were requested to participate in the study and questionnaire was presented to them. Of which, only 369 responded. A pediatricians group containing 654 members was selected, of which only 312 (47.7 %) responded. Otorhinolaryngologists group containing 721 was requested, of which 328 (52.8 %) responded. The Chi square test shows the specialty of the respondent is independent of response rate at $p < 0.0001$ (as represented in Table 1; Fig. 5).

We presented the questionnaire to the interns in our hospital, out of the 150 interns, only one (0.66 %) was able to explain about the condition. In General practitioners group containing 369 respondents only 4 (1.08 %) knew the condition and have seen patients. A pediatricians group containing 312 respondents only 23 (7.37 %) pediatricians knew the condition, 289 (92.62 %) persons had no idea about the treatment. Otorhinolaryngologists group containing 328 respondents. 21 (6.40 %) knew the condition, 293 (89.3 %) have no idea. The Chi square test shows that the specialty of the respondent like pediatrician and ENT practitioner have more knowledge than the other for jugular phlebection at $p < 0.0001$ as shown in Table 2.

The Chi square test shows that the specialty of the respondent like pediatrician and ENT practitioner have seen jugular phlebection at $p < 0.005$ (Tables 3, 4).

Out of 1,159 medical practitioner of our study, total number of patients seen by them were 25 (2.16 %), which shows that prevalence of the disease was 2.16 %. Out of the 25 patients most of the patients by the Otorhinolaryngologists 15 (60 %) and Pediatricians 9 (36 %). The General Practitioner had a small number, only one (4 %).

Table 1 Frequency and percentage distribution of medical practitioner who have participated in the study of knowledge of jugular phlebection

Speciality	Response	Non response	Total	χ^2 value	Significant
Interns	150 (100 %)	0 (0 %)	150	193.14	$p < 0.0001$
General practitioners	369 (39.6 %)	563 (60.4 %)	932		
Paediatricians	312 (47.7 %)	342 (52.3 %)	654		
Otorhinolaryngologists	328 (52.8 %)	293 (47.2 %)	621		
Total	1,159 (49 %)	1,198 (51 %)	2,357		

Fig. 5 Response by various categories of doctors **Table 2** Frequency and percentage distribution of medical practitioner having the knowledge regarding the jugular phlebectasia

Specialty	Knowledge of the jugular phlebectasia yes	No knowledge of the jugular phlebectasia	Total	χ^2 value	<i>p</i> value
Interns	1 (0.66 %)	149 (99.37 %)	150	25.15	<i>p</i> < 0.0001
General practitioners	4 (1.08 %)	365 (98.92 %)	369		
Paediatricians	23 (7.37 %)	289 (92.63 %)	312		
Otorhinolaryngologists	21 (6.40 %)	307 (93.60 %)	328		
	49 (4.23 %)	1,110 (95.77 %)	1,159		

Table 3 Frequency and percentage distribution of medical practitioner who have seen jugular phlebectasia patients

Specialty	Yes	No	Total	χ^2 value	<i>p</i> value
Interns	0 (0 %)	150 (100.00 %)	150	13.26	<i>p</i> < 0.005
General practitioners	4 (1.1 %)	365 (98.9 %)	369		
Paediatricians	9 (2.9 %)	303 (97.1 %)	312		
Otorhinolaryngologists	15 (4.6 %)	313 (95.4 %)	328		

Table 4 Frequency and percentage distribution of medical practitioner who either follow up or do surgery for jugular phlebectasia patients

Speciality	Follow up	Surgery	Total
General practitioners	1 (0.27 %)	–	1 (0.09 %)
Paediatricians	7 (2.24)	2 (0.64 %)	9 (0.78 %)
Otorhinolaryngologists	8 (2.43 %)	7 (2.13 %)	15 (1.29 %)
Total	16 (1.38 %)	9 (0.78 %)	25 (2.16 %)

Discussion

Internal jugular phlebectasia first described by Zusschwerdt [2]. Though the exact etiology is not known, a number of factors is proposed which include anomalous reduplication of IJV, mechanical compression of right or left brachiocephalic vein, idiopathic [3, 4]. The most common cause of a neck mass that becomes visible or

increases in size with valsalva manoeuvre is laryngocele, followed by jugular ectasia and superior mediastinal tumour cysts. Other differential diagnoses include, pharyngeal pouch, external laryngeal diverticulum, cavernous haemangioma.

The IJV is the most commonly affected and in most cases it is unilateral and right side. Though there are reports of association with childhood asthma [5], both of our

patients were non asthmatic. Diagnosis can be done by using ultrasonography with Doppler, CECT, CT Angiography and MR venography [6]. Though ultrasound gives a accurate extent of the lesion and its relationship to the underlying structures, Doppler ultrasonogram done during valsalva manouver gives a accurate diagnosis. CECT has obvious advantage of delineating the adjacent structures.

Surgical intervention was done in the past with significant mortality and morbidity. Since it is a benign condition, conservative management with regular follow up is advised. Surgical management is planned only if it presents with complications like thrombosis, impending pulmonary embolism, horners syndrome. Congestive cardiac failure, spontaneous rupture which is very rare [7]. There is a rare chance that a thrombosed jugular phlebectasia can mimic a cervical adenitis and attempt to excise or draining it can lead to disaster.

Conclusion

Though jugular phlebectasia is a rare condition, it is not mentioned in any of the standard otolaryngology, pediatrics, pediatrics surgery textbooks. This makes the situation much worse, leading to misdiagnosis or wrong treatment.

So it is highly recommended that this condition should be mentioned in undergraduate and postgraduate textbooks in relevant fields. This will translate into correct diagnosis and unnecessary mismanagement.

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