JUGULAR PHLEBECTASIS AND ACHALASIA CARDIA: A case report and review literature

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ABSTRACT

Internal jugular vein phlebectasis and achalasia cardia were diagnosed in a 10 year -old boy. Chest film revealed posterior mediastinal mass. Ultrasonography of the neck showed fusiform dilatation of internal jugular vein and dilated esophagus. Esophagogram showed dilated esophagus with "Bird beak" or "Rat tail" appearance of distal part. The treatments of achalasia cardia were Heller's operation and esophageal dilatation and there was no treatment for phlebectasis.

Keywords: Phlebectasis, internal jugular vein, achalasia cardia, ultrasonography and esophagography.

INTRODUCTION

Internal jugular vein phlebectasis is an uncommon condition, defined as fusiform dilatation of internal jugular vein, usually idiopathic and asymptomatic. In this case we report internal jugular vein phlebectasia, which was occurred with achalasia cardia. The chest film, ultrasonography and esophagography were illustrated.

A CASE REPORT

A 10 year-old boy present with chronic vomiting and painless right neck mass for 2 years. This mass was increasing on straining or cough. The vomiting occur more prequently during swallowing liquid food than solid meal.

Physical examination showed a thin boy, with non-expansile cystic mass at right lower neck, anterior to sternocleidomastoid muscle. It was measured about 4 cm in diameter, and precipitating by cough, straining and Valsalva maneuver (Figure 1). Otherwise, it was unremarkable.

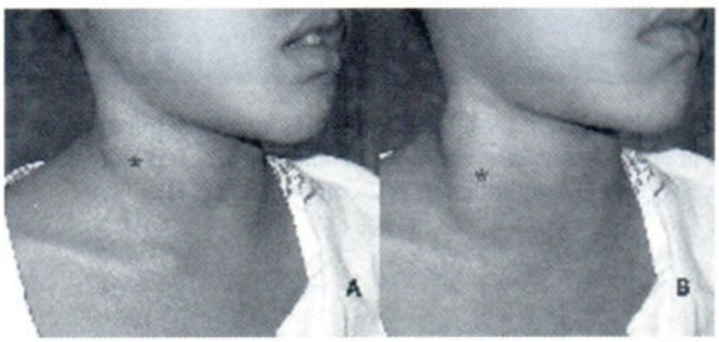

Fig.1 Photograph of a boy with right neck mass (*) **A:** Resting phase, **B:** The mass is increasing in size during straining.

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Chest film showed posterior mediastinum mass (Figure 4A).

Ultrasonogram revealed dilated right internal jugular vein, and increased more in size on Valsalva maneuver. (Figure 2) There was also dilated esophagus (Figure 3).

Esophagogram showed dilated entire esophagus, with gradual tapering end at distal part, seen as "rat tail" or "bird beak" appearance (Figure 4B and 5A-B)

The treatment was Heller's operation. The patient was slightly improved in a short time after operation, and there was recurrent vomiting, so dilatation of oesophagus have to be performed repeatedly.

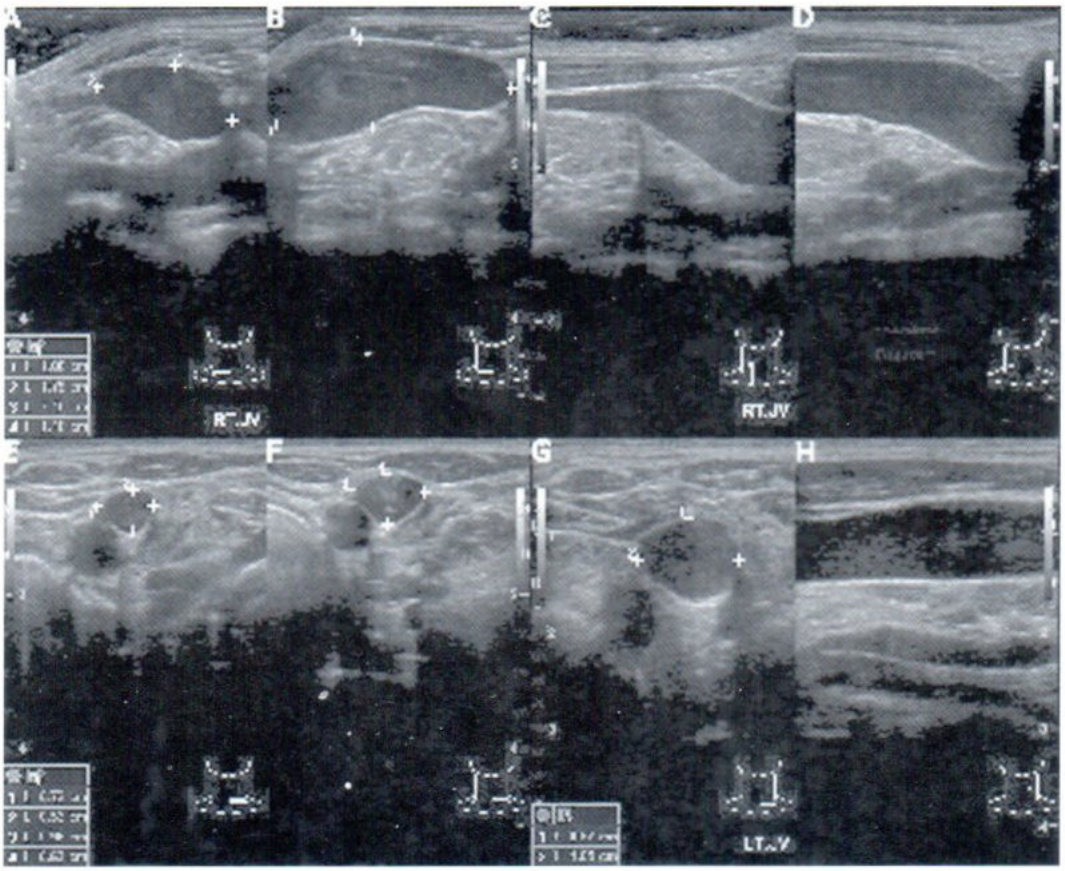

Fig.2 Ultrasonography of right (A-D) and left (E-H) internal jugular veins (caliper). Size of right IJV at rest (A-transverse, and C-longitudinal plane). It is more dilated during Valsalva maneuver (B and D), with the left IJV comparison (E-H).

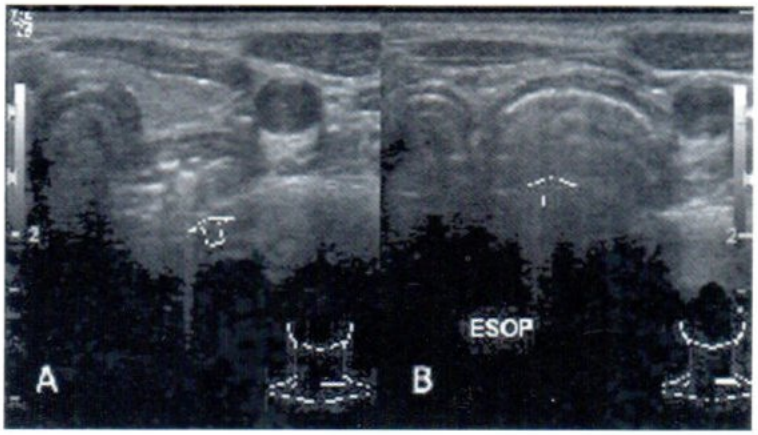

Fig. 3 At rest, air filled esophagus is noted at left side of trachea (arrow in **A**) and more dilated air-filled esophagus during Valsalva meneuver (arrow in **B**).

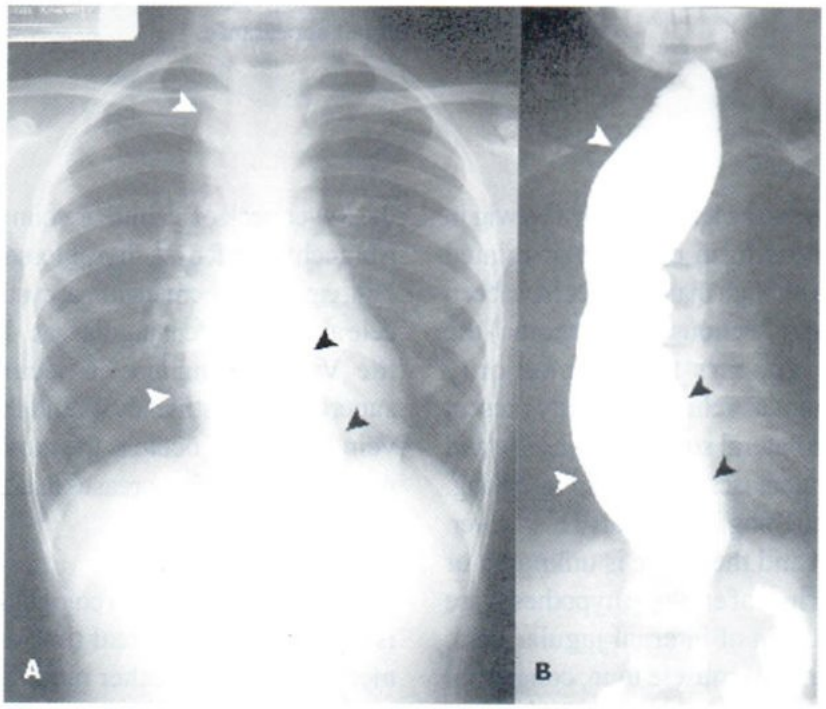

Fig. 4A Chest film shows posterior mediastinal mass (arrow head). Correlated frontal esophagogram **(B)** reveals dilated entire esophagus (arrow head).

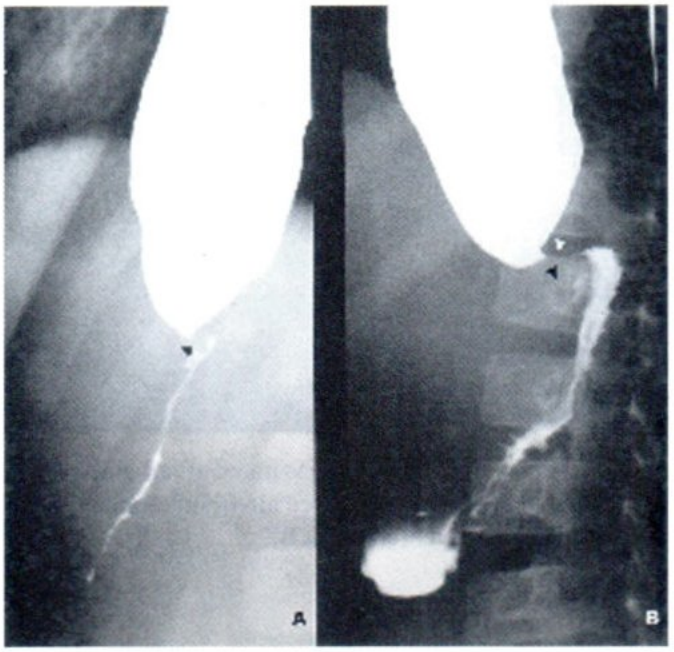

Fig. 5A, B Spot films of esophagogram show smooth tapering end of distal esophagus (arrow head) with bird beak or rat tail appearance.

DISCUSSION

Phlebectasis was the term used by Gerwig in 1952[1] to describe a fusiform or saccular dilated segment of the vein. Although many names have been used (venous aneurysm, venous cyst, venoma) the term phlebectasis appears to be commonly accepted.[2,3] Internal jugular vein is frequently present as a asymptomatic unilateral soft, non-palsatile and non-tender mass that increases with straining, crying, bending, sneezing, coughing and Valsalva maneuver. It is a rare condition and the cause is unknown or idiopathic.[2,4,5] The variety of etiology hypotheses are anomalous reduplication of internal jugular vein, increased scalenus anticus muscle tone, congenital origin, compression of the vein between the head of clavicle and the cupula, superior mediastinal irradiation, trauma, increased intrathoracic pressure and a congenital muscular defect in the wall of the vein.[2,3,5,6,7]

The differential diagnosis of a cystic mass in the lower neck of a child or young adult must include a branchial cleft cyst, thyroglossal duct cyst, dermoid cyst, cavernous hemangioma, cystic hygroma, laryngocele and a persistent jugular sac. The association with the Valsalva maneuver occurs in laryngocele, superior mediastinal cysts and the ectasia of jugular vein.[2,4,7] Laryngocele is the most common cause of neck mass, which increases in size with the Valsalva maneuver.[4,8]

Ultrasound with color Doppler flow imaging is the non-invasive, real time and best diagnostic method.[2,4,5,6,7] The other modalities used included computed tomography, venography, arteriography, nuclear scintigraphy and MRI.[2,6,7,8] Conservative management is generally recommended, but surgery may be advocated in cases of phlebitis, thrombus formation, rupture of the lesion or cosmetic deformity.[10]

Achalasia cardia was first described by Thomas Willis in 1674 . It is a condition, characterized by failure of relaxation of distal esophagus and esophagogastric junction. It could be primary, which there is absence of ganglion cells in the myenteric plexus or it could be secondary, which there is progressive degeneration of ganglion cells such as Chaga's disease. It is usually found in middle aged female, but uncommon in children.[12,13,14,15,16] Less than 5% of patients present before the age of 15 years.[11] Dysphagia is the most common presenting symptom in the patients with achalasia cardia. The child commonly presents with regurgitation, vomiting, recurring chest infection and rarely with respiratory distress and near miss sudden infant death syndrome.[11,14,16,17,18]

Diagnosis of achalasia cardia is based on history, barium swallowing, upper endoscopy and esophageal manometry. The esophagogram reveals dilated esophagus with smooth narrowing of the distal esophagus and esophagogastric junction, described as "bird beak" or "rat tail" appearance.[11, 12, 15, 16] As the degenerative neural lesion of this disease can not be corrected, the treatment is palliation of symptom and prevention of complications. The aims of treatment are improving of esophageal emptying time and decreasing in diameter of esophagus. The palliative treatments are pneumatic dilatation, Heller myotomy, pharmacotherapy and botulinum toxin injection.[11, 12, 17,19,20 ,21]

In the review literature, there is no previous report, mentioned that achalasia cardia occurs with jugular phlebectasis. This is the first report case of achalasia cardia, which there is dilated esophagus in the mediastinum and we think that is the cause of venous drainage obstruction, so there is resulting as dilatation of internal jugular vein.

In conclusion, although internal jugular phlebectasis is an idiopathic entity; we should look for the venous pathway obstruction as in this case.

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