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CASE REPORT

Congenital internal jugular phlebectasia

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KEYWORDS

Jugular vein;
Ectasia;
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Summary

Introduction: Congenital internal jugular phlebectasia corresponds to congenital dilatation of the vein without tortuosity. More than one hundred cases of phlebectasia involving the neck veins have been reported in the literature. The authors describe the clinical features, treatment and outcome of this anomaly.

Case report: A six-year-old child with no particular past medical history presented with a two-year history of progressively enlarging mass on the right side of the neck. Physical examination revealed a non-pulsatile mass that was increased in size by all manoeuvres increasing intrathoracic pressure. Neck computed tomography confirmed the diagnosis of internal jugular vein phlebectasia. In the absence of complications, simple surveillance was advised with a follow-up of 20 months with no complications.

Discussion/Conclusion: Congenital internal jugular phlebectasia is a rare condition. Management must include imaging to confirm the diagnosis. Long-term surveillance is recommended and surgery is only required in the presence of complications.

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Introduction

Congenital internal jugular phlebectasia is an entity that has been reported more frequently over recent decades. It corresponds to congenital dilatation of the jugular vein without tortuosity [1]. About 100 cases of phlebectasia in the neck have been reported in the English language literature [2]. This case report illustrates the clinical features, imaging, and treatment of this disease.

Case report

A six-year-old child, A.E., with no history of trauma, presented with a mass on the right side of the neck slowly enlarging over a period of two years. Manoeuvres raising intrathoracic pressure demonstrated a non-pulsatile, right cervical mass situated medially to the right sternocleidomastoid muscle, four centimetres in diameter, with a soft consistency, and accompanied by neck pain following these manoeuvres (Figs. 1 and 2). Nasal endoscopy was normal. Postcontrast computed tomography of the neck showed right internal jugular phlebectasia (Fig. 3). In the absence of complications, quarterly surveillance of this case of congenital internal jugular phlebectasia was recommended. Follow-up is now 20 months with no complications.

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Figure 1 Photograph at rest.



Figure 2 Appearance of a right lateral neck mass when increasing intrathoracic pressure (inflating a balloon).

Discussion

Between 1996 and 2001, only 31 cases of internal jugular phlebectasia in children were described in the United Kingdom, mostly occurring on the right side [2].

Congenital internal jugular phlebectasia is due to dilatation of the jugular sinus. This dilatation has a multifactorial origin: hypertension of the superior vena cava system during inspiration and the particular distribution of valves in the superior vena cava system, as well as anomalies of the vein wall. Several anatomical factors also predispose to hypertension of the right brachiocephalic vein compared to the left brachiocephalic vein: the right brachiocephalic vein



Figure 3 Axial CT scan of the neck showing right internal jugular phlebectasia.

travels in the same direction as the superior vena cava; the diameter of the right brachiocephalic vein is greater than that of the left brachiocephalic vein and the right brachiocephalic vein is shorter than the left brachiocephalic vein; finally, valves are present in the left brachiocephalic vein in 4 to 8% of cases [2]. These various factors would account for the higher incidence of phlebectasia of the right internal jugular vein.

Clinically, patients report neck pain and heaviness, unstable voice or even aphonia with no history of neck trauma. Physical examination demonstrates a non-pulsatile, lateral cervical mass with a fluid consistency, evolving since childhood and increasing in volume on Valsalva manoeuvre [1,2].

Complementary examinations are performed to confirm the diagnosis of congenital internal jugular phlebectasia, demonstrate complications such as thrombosis and for follow-up. Doppler ultrasound can demonstrate venous flow. Postcontrast computed tomography of the neck can distinguish between a solid tumour and a vascular lesion. Venography may show a normal or thinned venous wall in the zone of phlebectasia [1].

The differential diagnosis comprises other non-pulsatile masses of the neck, such as laryngocele, superior mediastinal cysts and tumours and hypertrophy of the pulmonary apex [2,3].

Complications are rare, such as thrombosis [3,4] and Horner's syndrome [5], and the main complaint is essentially cosmetic. No case of rupture has been reported in the literature [2].

In the absence of complications, recommended management is surveillance [1]. Surgery is required in the presence of a complication, and consists of resection of the phlebectasia or venorrhaphy depending on the type of dilatation. However, this type of operation is associated with a high surgical risk: vessel or nerve injury (vagus nerve injury, phrenic nerve injury or brachial plexus injury), air embolism or venous thrombosis [1].

Conclusion

Congenital internal jugular phlebectasia is a rare entity. The diagnosis should be considered in the presence of a neck mass present since childhood and enlarged by manoeuvres increasing intrathoracic pressure (such as Valsalva manoeuvre). The diagnosis must be confirmed by imaging. Surgery is only indicated in the presence of complications.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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