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Bilateral internal jugular phlebectasia

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Abstract
Internal jugular phlebectasia is a venous anomaly commonly presenting as a unilateral neck swelling in children. The clinicopathology, aetiology and management are discussed. Bilateral doppler ultrasonography is the diagnostic investigation of choice and should be performed in all suspected cases. Conservative management of the bilateral case is recommended.

Introduction

Internal jugular phlebectasia was first described by Harris (1928) and subsequently characterized by Gerwig (1952). Clinically it is a rare condition in which there is a fusiform or saccular dilatation of the internal jugular vein. Thirty-two cases of unilateral swellings have been reported under a variety of names including venous cyst, venous aneurysm, venectasia, venous ectasia, aneurysmal varix, and venoma but only once has a case presented with bilateral neck swellings (Leung *et al.*, 1983).

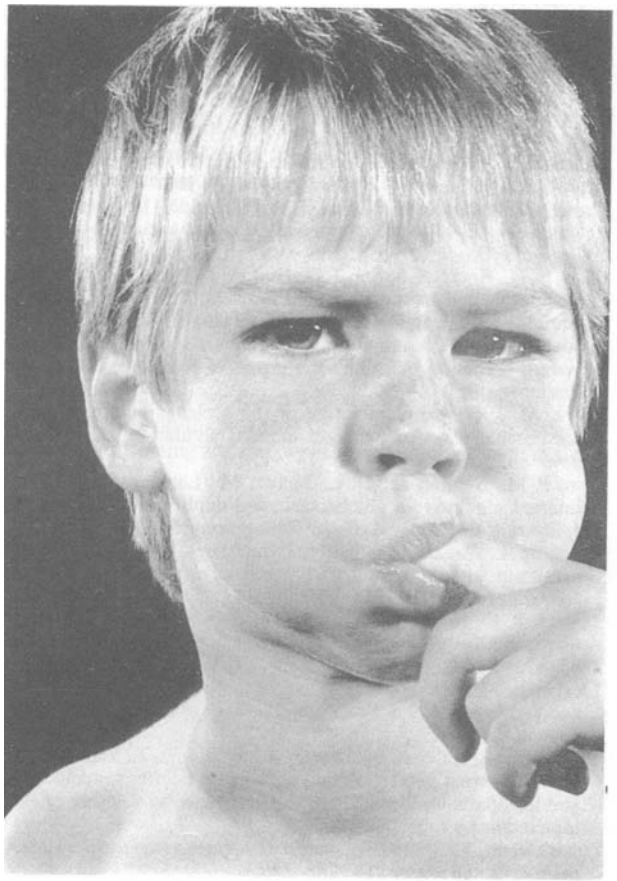

FIG. 1
Cystic swelling in right anterior triangle of neck during the Valsalva manoeuvre.

We report a case of bilateral internal jugular phlebectasia presenting as a unilateral swelling in the neck.

Case report

A 5-year-old boy presented with a three-month history of a painless right neck swelling which had only become noticeable on or after exertion. Examination during a Valsalva manoeuvre revealed a soft non-tender 2 × 3 cm cystic mass in the lower neck located anterior to the anterior border of the sternocleido mastoid muscle (Fig. 1). This cystic mass was obliterated by digital pressure from above downwards but a venous hum was not detected by auscultation. Clinical examination of the left side of the neck was unremarkable.

Ultrasonography investigation of the neck showed a non-tortuous 3 cm diameter dilatation of the distal segment of the right internal jugular vein on Valsalva manoeuvre only (Fig. 2). A similar but less pronounced phenomenon was present in the left internal jugular vein (Fig. 3).

Bilateral internal jugular phlebectasia was diagnosed. In the

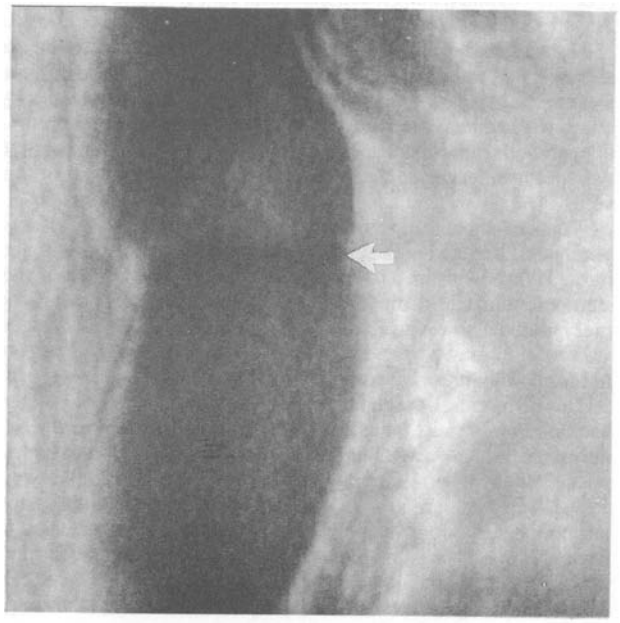

FIG. 2
Ultrasound scan of right neck showing 3 cm diameter dilatation of internal jugular vein.

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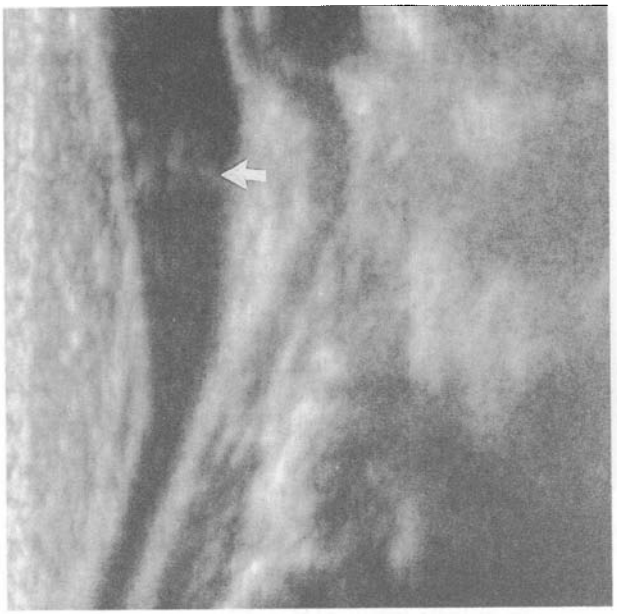

FIG. 3
Ultrasound scan of left neck showing 1.5 cm diameter dilatation of internal jugular vein.

absence of other symptoms surgical excision was not recommended. The patient remains well with the swellings unchanged some 18 months later.

Discussion

The majority of cases are idiopathic but several predisposing factors have been suggested: superior mediastinal irradiation (Harris, 1928); anomalous reduplication of the internal jugular vein (Zukschwerdt, 1929; Som *et al.*, 1985), increased scalenus anterior muscle tone (Rowe, 1946), compression of the jugular bulb between the head of the clavicle and the cupula of the right lung (Gerwig, 1952, Lamonte *et al.*, 1976), trauma (Teoderescu *et al.*, 1978) and a congenital muscular defect within the wall of the vein itself (Yokomori *et al.*, 1990).

The histopathological findings vary. Several authors found no significant abnormality (Gerwig, 1952; Gordon *et al.*, 1976) but Davis (1982) reports elastic tissue dysplasia, focal intimal thickening with an increased amount of connective tissue and prominent smooth muscle cells. Yokomori *et al.* (1990) in a report on two siblings described a muscular defect within the wall of the vein.

Phlebectasia of the internal jugular vein predominantly affects young children and presents as a soft cystic swelling commoner in the right lower third of the neck at the anterior border of the sternocleidomastoid muscle (Bowdler and Singh, 1986). It is precipitated by the Valsalva manoeuvre and disappears at rest. A venous hum may be present and the swelling can classically be obliterated by pressure from above.

The differential diagnosis of a cystic swelling in the lower neck of a child must include a branchial cyst, thyroglossal cyst, dermoid cyst, cavernous haemangioma, cystic hygroma, laryngocoele and a persistent jugular sac (Steinberg and Watson, 1966). The association with the Valsalva manoeuvre occurs in laryngocoeles, superior mediastinal cysts and phlebectasia. A chest X-ray, which includes the lower neck area, performed during Valsalva and at rest may exclude the former two, but further radiological evaluation is required to confirm the diagnosis of phlebectasia.

Historically, percutaneous venography either by direct puncture or by the transfemoral route was the investigation of choice (Okay *et al.*, 1970; Gordon *et al.*, 1976; Passariello *et al.*, 1979). This invasive technique is however potentially dangerous and the non-invasive techniques of ultrasonography (Stevens *et al.*, 1982) and CT scanning (Som *et al.*, 1985) have been advocated. Ultrasonography would appear to be the investigation of choice as it is widely available, comparatively inexpensive, and accurately defines the extent of the lesion and its relationship with the surrounding structures in the lower neck. Magnetic resonance imaging (MRI) can also demonstrate the lesion but should not normally be needed.

The natural history of jugular phlebectasia has not been documented but there would seem to be no reason why spontaneous obliteration should occur. Although rarely discomforting (Bowdler and Singh, 1986) the lesion has on occasions been surgically excised for cosmetic reasons. In the case reported with bilateral internal jugular phlebectasia surgical excision would be hazardous with a high risk of cerebral oedema and its consequences. As there are no reported cases of rupture of the untreated internal jugular phlebectasia we advocate a conservative policy.

Conclusion

Internal jugular phlebectasia can occur bilaterally although it may appear clinically unilateral. Ultrasonography is the investigation of choice and should be performed on both sides of the neck. In bilateral cases of jugular phlebectasia a conservative policy can be adopted.

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