

Challenging surgical management of right internal jugular vein haemangioma: a case report

Özlem Balcioğlu¹*, MD, Serkan Ertugay², MD, Hakan Posacioğlu², MD

¹ Cardiovascular Surgery Department, Near East University, Nicosia, Cyprus

² Cardiovascular Surgery Department, Ege University, Izmir, Turkey

* Corresponding author: balcioglu@neu.edu.tr

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Case report

A 27-year-old female was admitted in January 2021 to the cardiovascular department of Ege University, Turkey with a history of a gradually enlarging swelling in the right supraclavicular area. Physical examination revealed a semi-soft, semi-mobile and painless mass localised at the bottom third of the right lower neck, extending to the clavicle with no distinct inferior border. It appeared to follow upper mediastinal inflow. The overlying skin was normal, and no pulsation or thrill was detected. All laboratory results including infection markers were within normal limits. Sonographic evaluation demonstrated a solid, hypoechoic and lobulated mass with slow flow pattern and slow filling of the right internal jugular vein (IJV). Computed tomography scan illustrated a homogenous oval solid lesion on the right lateral aspect of the neck, originating from the thyroid level and elongating to the infraclavicular area. It measured 33 × 51 × 51 mm, with peripheral interrupted nodules in arterial phase (Fig 1a). The mass was located 180° to the IJV. The border could not be differentiated and the trachea was deviated medially to the left and right common carotid artery posteriorly. Magnetic resonance imaging demonstrated intermediate signal intensity on T1-weighted image (Fig 1b) and very high signal intensity on T2-weighted image of an enhanced solid mass (Fig 1c). The clinical diagnosis was vascular malformation or haemangioma. Surgical excision was planned. After general anaesthesia and positioning of the neck, a neck incision was made parallel and anterior to the right sternocleidomastoid muscle, similar to that performed for a standard carotid endarterectomy. Although the incision was extended through the sternal notch, it was insufficient to enable complete excision. A right-sided mini ‘J-sternotomy’ was performed subsequently to facilitate complete visualisation of the brachiocephalic bifurcation. Exploration and dissection of the mass from peripheral tissue revealed that it arose from the distal part of the right IJV and extended through the brachiocephalic bifurcation.

The proximal part of the right IJV, right subclavian vein and distal part of the right

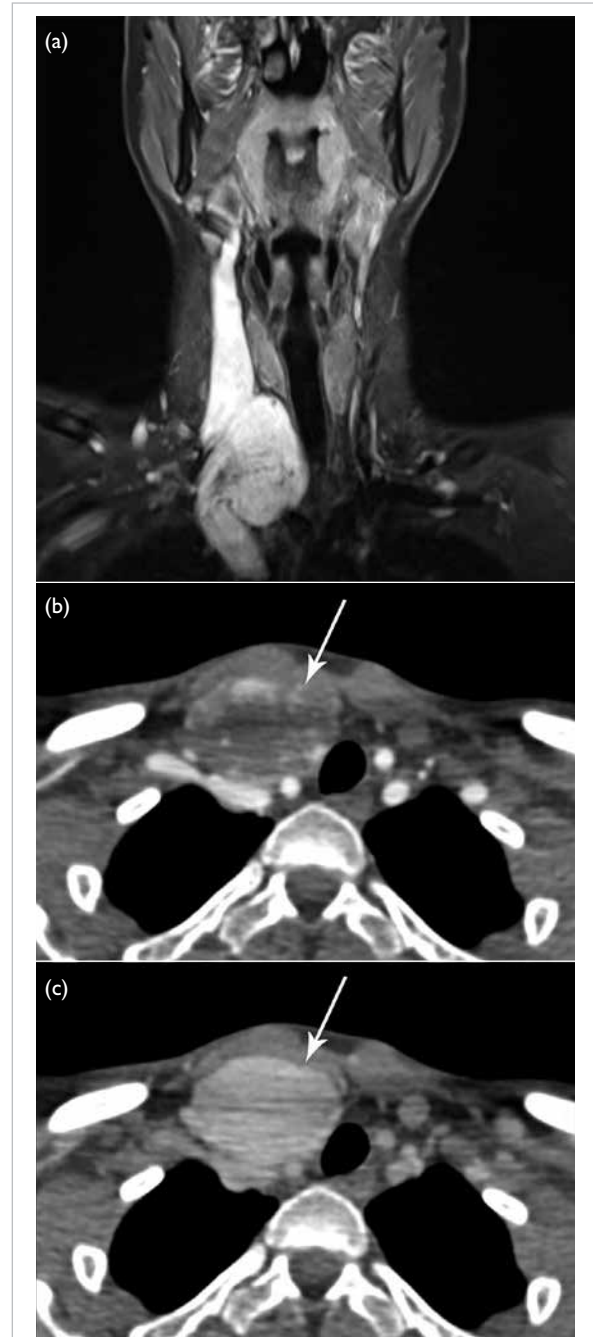


FIG 1. (a) Computed tomography angiography image of the mass with peripheral interrupted nodules in arterial phase. Magnetic resonance imaging scans with (b) intermediate signal on T1-weighted phase and (c) very high intensity signal on T2-weighted phase

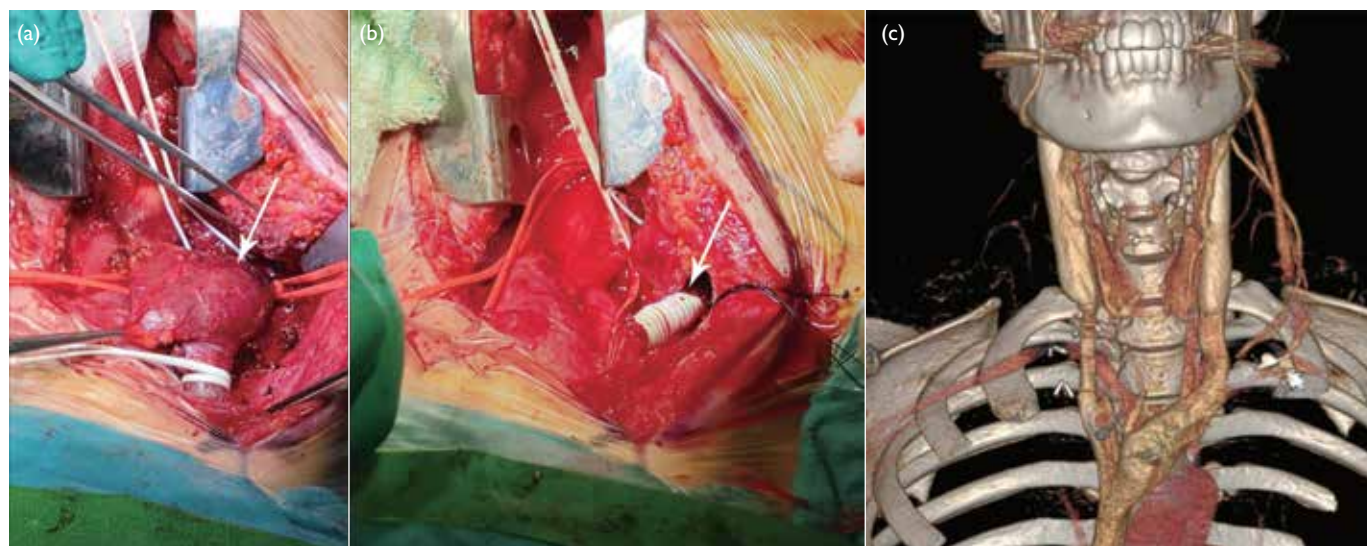


FIG 2. Surgical view showing (a) the mass and (b) reconstruction of the vein with synthetic graft [arrows]. (c) Postoperative computed tomography scan shows patent graft with excellent reconstruction

innominate vein were explored and controlled by silicon loop. Following heparin administration, the proximal right subclavian vein was ligated and all other vessels clamped. An incision was made and the mass was observed to extend into the vascular lumen. It was removed en bloc along with the distal segment of the right IJV and proximal segment of the right brachiocephalic vein. Vein reconstruction was performed to prevent venous hypertension in the neck. Because of the large diameter of vascular structures, a synthetic 8-mm polytetrafluoroethylene self-ringed graft was sutured between the distal part of the right innominate bifurcation and right IJV (Fig 2a and 2b). Histopathology revealed CD34(+), endothelial cell(+) haemangioma. The patient experienced no postoperative complications. Both antiaggregant and anticoagulant therapy were commenced on postoperative day 1 with acetylsalicylic acid (100 mg/day) and apiksaban (5 mg/day). Control computed tomography scan 11 months after surgery revealed excellent reconstruction with an intact and patent graft (Fig 2c).

Discussion

The nomenclature of primary tumours of the venous system is based on their origin: lipomas, leiomyomas, haemangiomas, leiomyosarcomas, and angiosarcomas. Leiomyosarcomas are the most common tumours with a malignant course. Until the classification scheme proposed by Mulliken and Glowacki,¹ the terms 'haemangioma' and 'vascular

malformation' were used interchangeably because of the lack of a standardised nomenclature. Although they are classified as benign tumours, accurate diagnosis is possible only by histopathological evaluation following surgical excision. According to their classification, haemangiomas are characterised by rapidly proliferating endothelial cells and frequent mitosis. They are not usually visible at birth but become apparent at the neonatal stage and demonstrate rapid proliferation during the first 2 years, followed by spontaneous involution. In contrast, vascular malformations possess flattened endothelial cells and ectatic vessel formation. Although haemangiomas grow with hyperplasia, vascular malformations expand by hypertrophy.¹ Some authors have noted that trauma, sepsis, hormonal changes or pressure in the venous system may cause expansion of the vascular malformation.² Although cases of external jugular vein haemangiomas and vascular malformations have been reported,² only three cases of IJV haemangioma have been reported. The exceptions were incidentally diagnosed asymptomatic cases; these presented with swelling and may have led to incorrect differential diagnoses of neck malignancy, infection, lymphoma or thrombosis. A multidisciplinary approach with a full physical examination and medical history is required to reach a definitive diagnosis. Advanced imaging techniques play an important role in diagnosis and are helpful when planning treatment. The sonographic and magnetic resonance imaging features of vascular malformations and haemangiomas have been studied and described

in detail. In a case series, Ahuja et al³ reported the radiological features of vascular malformations in the external jugular vein, aiming to help clinicians make treatment decisions. Head and neck vascular malformations typically showed intermediate signal intensity on T1-weighted images, very high signal intensity on T2-weighted images and variable enhancement following intravenous administration of gadolinium.⁴ Similar radiological features were identified in our case. To date, different treatment modalities such as cryotherapy, laser therapy, and steroids have been explored. Excision of a benign vein tumour with the adjacent vein segment en bloc is the most successful curative treatment and enables pathological confirmation of the diagnosis.⁵

In previous case studies of IJV haemangiomas, a supraclavicular approach was applied to access the mass and reconstruction not performed following removal of the vein segment.⁵ In our challenging case, the neck incision was extended to the sternal notch and continued with a right-sided mini ‘J-sternotomy’ to ensure adequate control over the mass. Contrary to other case reports of IJV haemangioma, we performed vein reconstruction to prevent venous hypertension and swelling of the neck. To the best of our knowledge, this is the first reported case where vein reconstruction was performed using a synthetic graft following successful surgical excision of a haemangioma from the IJV via an extraordinary approach.

Author contributions

Concept or design: Ö Balcıođlu, S Ertugay.
 Acquisition of data: S Ertugay, H Posacıođlu.
 Analysis or interpretation of data: All authors.
 Drafting of the manuscript: Ö Balcıođlu, S Ertugay.

Critical revision of the manuscript for important intellectual content: All authors.

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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Ethics approval

The patient was treated in accordance with the Declaration of Helsinki and provided written informed consent for publication of this report.

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