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CASE REPORT

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Idiopathic external jugular vein thrombosis

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Abstract

Idiopathic external jugular vein (EJV) thrombosis is a very rare condition. EJV thrombosis usually develops as a result of injury, intravenous cannulation, intravenous injections, inflammatory and malignant tumours of the neck or haematologic diseases. This article presents a case of a 29-year-old otherwise healthy male admitted to the Department with left EJV thrombosis manifesting as a new onset of swelling of the left cheek and side of the neck which he noticed upon awakening in the morning. The oedema resolved spontaneously a few hours later. Clinical examination only revealed a mild bulge in the left supraclavicular area. Doppler ultrasound (DUS) and computed tomography angiography (CTA) revealed an oval, heterogeneous mass with a smooth contour and well-circumscribed margins within the terminal segment of the left EJV, adjacent to the vascular wall. The mass did not occlude the vessel completely, but only caused its significant dilatation above the lesion, and diagnostic imaging could not unequivocally exclude a tumour. Therefore, the patient underwent surgery whereby the diseased part of the EJV was removed. The histologic evaluation confirmed a typical structure of a thrombus. The postoperative course was uneventful. To date, over the 4-year follow-up, there was no recurrence confirmed either clinically or in duplex US. The case has been discussed in the context of the available literature.

Keywords: external jugular vein, EJV, thrombus, idiopathic external jugular vein thrombosis

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Case report

A 29-year-old otherwise healthy male was admitted to the Department with left EJV thrombosis. It presented as a new onset of swelling of the left cheek and side of the neck which he noticed upon awakening a few hours earlier. He did not report pain or any other symptoms. The patient denied any recent injury or trauma to the affected area, respiratory tract infection, recent dental treatment or substance use (prescribed or recreational). The oedema resolved spontaneously while the patient attended the emergency department. Clinical examination only revealed a soft bulge sized 1.0–1.5 cm in diameter in the left supraclavicular area, which was painless on palpation (Fig. 1). The biochemistry blood test results were normal. The duplex ultrasound (DUS) revealed an oval, heterogeneous mass sized 11 × 5 mm with a smooth contour within the terminal segment of the left EJV, with a 3–4 mm peduncle attached to the vascular wall (Fig. 2). The radiology report suggested thrombus, with intravascular tumour to be included in the differential diagnosis. The computed tomography angiography (CTA) revealed an oval intraluminal lesion within the terminal segment of

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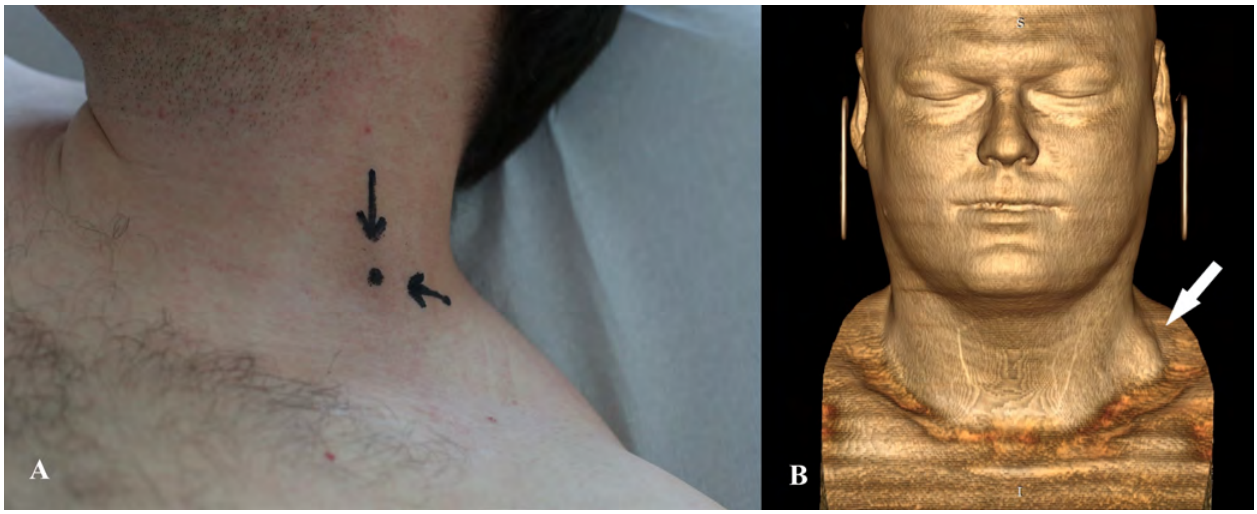

Figure 1. A. Slight convexity of the side of the neck (the arrows show the location of the thrombus in the external jugular vein, as determined by ultrasound examination); **B.** Volume Rendering 3D CT of the face and the neck — the arrow shows the convexity of the lateral neck area

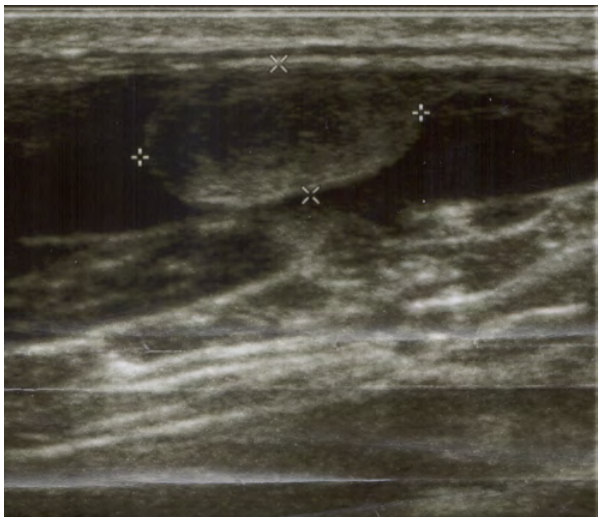

Figure 2. Ultrasound image of an oval, smooth, heterogeneous mass in the distal segment of the left external jugular vein

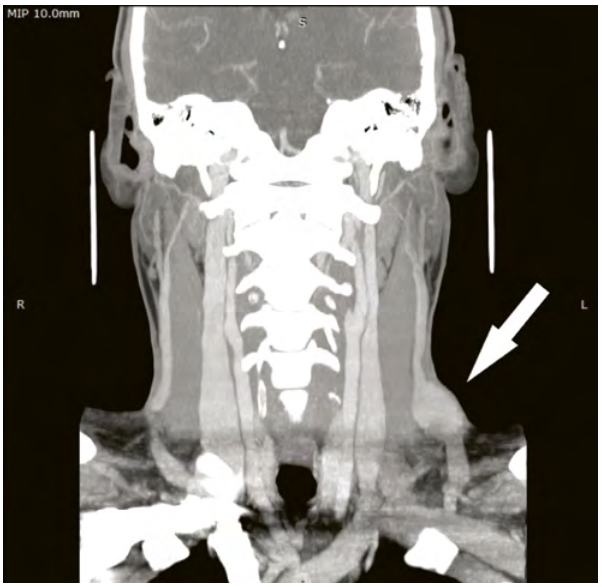

Figure 3. 3D CT angiography reconstruction — the arrow shows a thrombus inside the jugular vein

the left EJV, sized 10 × 7 mm (cc, 10 mL) with well-circumscribed margins, adjacent to the vascular wall. The radiology report again suggested a thrombus, with an intravascular tumour to be included in the differential diagnosis (Fig. 3). Above the lesion, there was EJV dilatation up to 25 × 20 mm. The ENT assessment did not demonstrate any other abnormalities, apart from the lesion in question. Since diagnostic imaging findings were inconclusive, suggesting possible malignancy, the authors opted for surgical treatment. Perioperatively, the patient was administered small molecular weight heparin (enoxaparin, 80 mg every 24 h). The surgery was performed under general anaesthesia with the patient lying supine and his head turned rightward 90°. A 3 cm long segment of the EJV was removed including the intraluminal lesion. Having dissected the vein, a solid thrombus sized 1 cm in diameter, firmly attached to the vascular wall, was found within the lumen (Fig. 4). The histologic evaluation confirmed a typical structure of a thrombus. The postoperative course was uneventful and on day 2 postoperatively the patient was discharged home. Haematologic tests carried out 6 weeks later excluded thrombophilia. To date, over the 4-year follow-up, there was no abnormality confirmed either clinically or in duplex US.

Discussion

Jugular vein thrombosis usually affects the internal jugular vein (IJV), which constitutes up to 5% of

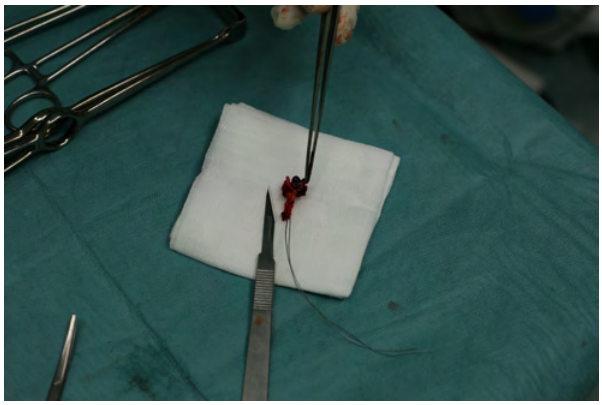

Figure 4. A fragment of the external jugular vein with a thrombus firmly attached to its wall (the tip of the forceps indicates a thrombus)

all thrombosis cases [1]. EJV thrombosis is even less common. It is usually secondary, induced by trauma, central venous cannulation, intravenous injections, inflammatory and malignant tumours of the neck or haematologic diseases [2–4].

Idiopathic EJV thrombosis is, therefore, very rare [5, 6]. A high-grade dilatation of the EJV, which may facilitate intravascular coagulation, is considered one of the possible causes [7]. This is, however, debatable, as EJV may as well get dilated as a result of thrombosis and flow obstruction. The case of the present patient, in whom the thrombus acted like a "stopper" leading to venous stasis, seems to confirm this theory. As a result, he developed swelling of the left cheek and side of the neck, which resolved following his assuming an upright position and ambulation.

It has been further corroborated by diagnostic imaging (DUS and CTA), which confirmed partial EJV obstruction. Most authors think that DUS and CTA are sufficient to inform the diagnosis and post-diagnostic monitoring [8]. In this case, however, there were some diagnostic challenges, as both DUS and CTA failed to provide an unequivocal answer as to the nature of the lesion (thrombus vs intraluminal tumour), which is why the authors opted for surgical treatment.

There are no guidelines on the management of idiopathic EJV thrombosis. Possible progression of thrombosis and subclavian vein involvement leading to pulmonary embolism prompts conventional anticoagulant therapy with low molecular weight heparin and other anticoagulants [8]. However, in EJV thrombosis, where large subcutaneous masses are present, surgical treatment involving the excision of the diseased venous segment is preferred [7, 9].

To sum up, it should be noted that a fairly small thrombus in a superficially located blood vessel can still be a source of significant diagnostic challenges despite considerable advances in medical technologies and knowledge.

Article information and declarations

Ethics statement: The authors' institution does not require ethical approval for reporting individual cases or case series.

Author contributions: The authors confirm their contribution to the paper as follows: study conception and design: WW, MW, SM, KM; analysis and interpretation of data: WW, MW, KM, PM; draft manuscript preparation: WW, MW, SM, KM, PM. All authors reviewed the results and approved the final version of the manuscript.

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