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Internal jugular vein fenestration: a rare but possible event. A case report and review of the literature

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Abstract

While fenestration and duplication are relatively common in the arteries, they are extremely rare in the venous compartment: internal jugular vein fenestration has been reported occurring in 0.4% of unilateral neck dissections. Familiarity with these morphological anomalies is important for the radiologist and for the surgeon to prevent neurovascular injury, especially in neck surgery and interventional catheterization. We present the case of a patient harboring a fenestration of the left internal jugular vein, diagnosed by magnetic resonance angiography, and a systematic review of the literature. To our knowledge, from 1985 until 2016 only 36 patients (including the present) were diagnosed as having an internal jugular vein morphological anomaly. Out of 36 patients, only 11 (30,5%) were diagnosed using radiological imaging; the high rate of intraoperative diagnoses (22/36, 62,5%) is likely related to the limited use of diagnostic imaging or to misdiagnosis/misinterpretation of a relatively unknown and rare morphological anomaly. A contrast enhanced computed tomography or magnetic resonance angiography should be considered in case of vascular procedures in a patient with known internal jugular vein anomaly.

Key words

Duplication, fenestration, internal jugular vein, magnetic resonance angiography.

Introduction

The jugular venous system constitutes the primary venous drainage of the head and neck, in a pattern common to human and other species (Williams, 1995; Mancini et al., 2015).

It is known that the internal jugular vein (IJV) presents high variability in its flow rate and cross-sectional area (Cocozza et al., 2016), whereas fenestration and duplication are extremely rare, compared to the arterial compartment: IJV fenestration has been reported occurring in 0.4% of unilateral neck dissections (Prades et al., 2002).

Familiarity with these morphological anomalies is important for the radiologist and the surgeon to prevent neurovascular injury, especially in neck surgery and interventional catheterization.

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We present the case of a patient harboring a fenestration of the left IJV, diagnosed by magnetic resonance (MR) angiography (MRA), and a systematic review of the literature.

Case report

A 35 year-old-man with the diagnosis of Méniere's disease was admitted to investigate the coexistence of chronic cerebrospinal venous insufficiency and eventually planning of IJV percutaneous transluminal angioplasty.

Magnetic resonance imaging was performed with a 1.5 T clinical MR system (Magnetom Essenza Siemens), by using 3D contrast-enhanced MR angiography/venography imaging of the neck (TE 1.49 ms; TR 3.88 ms; flip angle 25°; slice thickness 1.20 mm; FOV 340 mm, FOV PHASE 75%), with maximum intensity projection reconstructions.

Contrast medium administration for time-resolved MR venography was performed using an automatic contrast-injector with intravenous 1 mmol/ml gadobutrol (Gadovist 1.0, Schering AG, Switzerland), 0.1 mmol/kg at 2 mL/s beginning simultaneously with the start of the sequence, followed by 20 mL bolus of saline at the same rate.

Magnetic resonance angiography and maximun intensity projection reconstructions showed that the left IJV emerged as a single trunk from the jugular foramen, then split into two parts, anterior and posterior, after descending for about 6.5 mm from the base of the skull. Both parts then rejoined to form a single trunk in the lower part of the neck, before joining the subclavian vein to form the brachiocephalic vein (Figure 1).

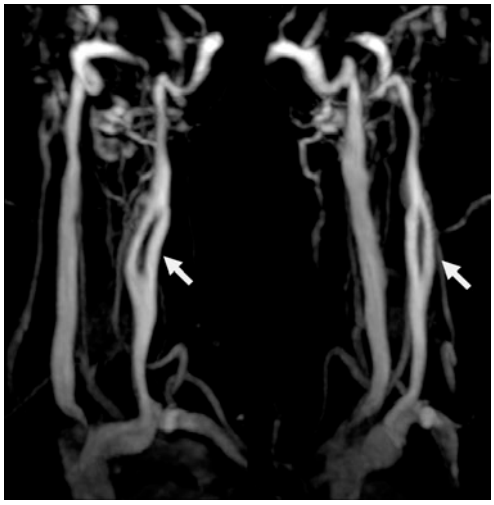

Figure 1. Magnetic Resonance Angiography (oblique views). Splitting of the internal jugular vein into two parts (arrows), then rejoining to form a single trunk in the lower part of the neck before joining the subclavian vein to form the brachiocephalic vein.

Discussion

The true incidence of IJV fenestration is difficult to assess since most duplicated or fenestrated IJVs are discovered as incidental findings (Cvetko, 2015) and because the terms duplication, partial duplication, and fenestration have been used interchangeably, although a duplication is defined as two distinct vessels with separate origins and no distal convergence, while fenestration is a division of a vessel into distinctly separate channels, each with its own endothelial and muscularis layers, while the adventitia may be shared (Parmar et al., 2005): it refers to a 'window-like opening' in the IJV, with a typical 'eye-of-the-needle' appearance (Dwonie et al., 2007). IJV fenestration are extremely rare, occurring in a recent study in 0.4% of unilateral neck dissections (Prades et al., 2002).

The primary blood vessels of head and neck consists embriologically of close meshed capillary plexus, drained on each side by the precardinal (anterior cardinal) vein, at first continuous cranially with a transitory primordial hind brain vein which is soon replaced by the primary head vein to become continuous with the precardinal vein.

The etiology of IJV duplication-fenestration is still unclear. Three hypothetical explanations have been suggested. The vascular theory, most commonly adopted, is based on the paucity or absence of the IJV muscular layer. The neural hypothesis assumes that the IJV anomaly depends on the altered position of the spinal accessory nerve in relation to the transverse process of the atlas, which can lead to the duplication of the developing IJV. The bony hypothesis suggests that variation in the ossification of the bony bridges of the jugular foramen causes venous duplication; this theory does not explain the relation of the spinal accessory nerve to the duplicated IJV - see below (Sylaidis et al., 1997; Guerra et al., 2000; Gardiner et al., 2002; Alaani et al., 2005; Striano et al., 2005).

To our knowledge, analyzing the literature using Medline database (www.ncbi.nlm.nih.gov/pubmed), from 1985 until 2016 only 36 patients including the present were diagnosed as having a IJV morphological anomaly (Table 1). Out of these 36 patients, 32 (about 89%) had unilateral anomaly, 4 (11%) bilateral anomaly. Out of 40 IJV anomalies, 21 (about 52,5%) were duplication, 19 (47,5%) were fenestration; 25 (62,5%) were on the left, 13 (32,5%) were on the right, 2 were not defined in the report. Out of 36 patients, 22 (about 61%) were diagnosed during neck surgery, only 11 (30,5%) using imaging, and 3 (8,5%) at cadaveric dissection.

Overall, morphological anomalies of the IJV are described more often as unilateral, on the left side, and in almost equal percentage in terms of fenestration and duplication.

Most duplications and fenestrations occur in the upper third of the IJV (Bachoo and Evans, 2014); in the case presented here, the fenestration was observed in the middle of the IJV.

Regarding to the way of detection, the high rate of intra-operative diagnoses (22/36, 62,5%) is likely related to the limited use of diagnostic imaging or to misdiagnosis/misinterpretation of a relative unknown and rare morphological anomaly (Caranci et al., 2015). Colour-Doppler ultrasonography and computed tomography angiography were the technique most often used for the diagnosis, while MRA was used only in 1 paper (Rossi et al., 2001) before our report. Only 3/36 (8,5%) were found in cadaveric dissections.

Table 1. Internal jugular vein anomalies reported in cronological order.

Authors	N.° of patients and anomalies	Morphological anomaly	Side	Diagnosis method
Som et al., 1985	1 unilateral	duplication	right	Imaging
Sylaidis et al., 1997	1 unilateral	fenestration	right	surgery
Guerra et al., 2000	1 unilateral	duplication	right	surgery
Rossi et al., 2001	1 bilateral	duplication fenestration	right left	Imaging
Gardiner et al., 2002	1 unilateral	fenestration	left	surgery
Prades et al., 2002	3 unilateral	fenestration fenestration fenestration	left left right	surgery
Towbin et al., 2004	2 unilateral	fenestration fenestration	left left	Imaging
Turan-Odzemir et al., 2004	1 unilateral	duplication	right	Imaging
Alaani et al., 2005	1 unilateral	fenestration	left	surgery
Nayak 2006	1 unilateral	fenestration	left	cadaveric dissection
Downie et al., 2007	1 bilateral	duplication duplication	left right	cadaveric dissection
Gonzalez-Garcia et al., 2007	1 unilateral	duplication	left	surgery
Iseri et al., 2007	1 unilateral	fenestration	left	surgery
Uecker et al., 2007	1 unilateral	duplication	left	surgery
Coca Pelaz et al., 2008	1 bilateral	duplication duplication	left right	surgery
Colella et al., 2008	1 unilateral	duplication	right	surgery
Wong et al., 2010	1 bilateral	duplication duplication	left right	Imaging
Atalar et al., 2012	1 unilateral	fenestration	left	Imaging
Kapre et al., 2012	1 unilateral	fenestration	left	surgery
Radak et al., 2012	1 unilateral	duplication	right	Imaging
Thakur at al, 2012	1 unilateral	fenestration	not reported	surgery
Kayashima et al., 2013	1 unilateral	duplication	left	Imaging
Ayoub et al., 2014	1 unilateral	duplication	left	surgery
Bachoo et al., 2014	1 unilateral	duplication	left	surgery
Torres et al., 2014	1 unilateral	fenestration	left	Imaging
Cvetko et al., 2015	1 unilateral	fenestration	left	cadaveric dissection
Moreno-Sànchez et al., 2015	1 unilateral	fenestration	1 right	surgery
Pegot et al., 2015	1 unilateral	fenestration	left	surgery
Contrera et al., 2016	3 unilateral	2 fenestration, 1 duplication	2 left 1 right	surgery
Sidana et al., 2016	1 unilateral	duplication	not reported	surgery
Present report 2017	1 unilateral	fenestration	left	Imaging

The IJV is also a radiological landmark (Pegot et al., 2015). It is important for the radiologist to be familiar with the anatomical variations of these veins, in order to avoid misinterpretation and misidentification [31]: IJV anomalies have been sometimes mistaken as laryngoceles or branchial cleft cysts (Caranci et al., 2015).

Also, their knowledge is essential before performing head and neck surgery, oncological surgery, percutaneous catheterization or when planning IJV percutaneous transluminal angioplasty for chronic cerebrospinal venous insufficiency (Briganti et al., 2004, 2013, 2016), in order to avoid severe clinical consequences. In neck dissections, IJV anomalies could greatly increase the risk of bleeding, or make complete clearance of lymph nodes impossible, particularly if the patient has previously been treated with radiation. The IJV is also often used as a recipient vein for a free flap and in some cases fenestrations could increase operative difficulties and morbidity (Pegot et al., 2015).

The IJV is a common site for insertion of a central venous line; in case of a fenestration, difficulties in insertion of the catheter could cause vascular injury with cervical bleeding or hematoma. The anatomical relation of the IJV to the contiguous structures, particularly the accessory nerve, can change depending of the presence or absence of a fenestration or duplication. In fact, the spinal accessory nerve usually passes superficial to the IJV, so any tissue that is superficial to this landmark can be dissected during neck surgery with no risks. In duplicated or fenestrated IJV, the spinal branch of the accessory nerve is always reported to pass deep to the anterior branch of the duplicated internal jugular vein and superficial to the posterior branch (Parmar et al., 2005), or rarely deep to both posterior and anterior branches of the vein (Alaani et al., 2005).

Consideration of contrast enhanced computed tomography (CTA) or MRA is warranted if a patient with known IJV anomaly is undergoing interventional procedures involving major vascular structures (Caruso et al., 2002; Thakur et al., 2012; Torres et al 2014).

Patients with IJV bifurcation may have a higher prevalence of additional vascular abnormalities. Like in arteries, where duplication is associated with aneurysm formation, duplication of the IJV is usually reported in association with phlebectasia, a congenital dilation of the jugular venous system. Phlebectasia is a local fusiform, soft, non-pulsatile swelling in the cervical region which increases during Valsalva maneuver (Som et al., 1985; Rossi et al., 2001; Prades et al., 2002). IJV duplication associated with phlebectasia was reported in a 2 year old girl studied by color-Doppler ultrasound and MRA (Rossi et al., 2001).

In our patient no phlebectasia was assessed; moreover, he didn't find symptoms related to IJV anomaly. This is consistent with past reports, although there have been cases of patients presenting with neck swelling, dyspnoea, and dysphagia (Wong et al., 2010); moreover, IJV anomalies raises the possibility for deep venous thrombus formation secondary to changes in flow velocities (Cvetko, 2015).

In conclusion, IJV is a rare vascular anomaly that may have significant clinical consequences. Familiarity with these morphological anomalies is important for the radiologist and for the surgeon to prevent neurovascular injury, especially in neck surgery and interventional catheterisation. Given the advances and wider availability of imaging examinations worldwide in the last decades, it is expected that a growing number of cases will be identified (Torres et al., 2014; Cocozza et al., 2016).

Disclosures

All authors declare that they have no involvement, financial or otherwise, that might potentially pose conflict of interest.

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