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CASE REPORT

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Fenestration and duplication of contralateral internal jugular veins with concomitant cerebral arteries malformations

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Abstract

Duplication and fenestration of IJV are rare malformations that are usually accidental findings during neck surgeries and anatomical dissections. Their prevalence is estimated at 1–2%, while the prevalence of IJV duplication alone is around 0.4%. This is the first reported case of both fenestration and duplication of the contralateral IJVs and one of few that confirm the coexistence of vascular malformations in other vessels — in this case, cerebral arteries. The awareness of the presence of IJV malformations can prevent complications during surgeries in the neck area and vascular procedures such as IJV catheterization.

Key words: internal jugular vein; duplication; fenestration; vascular malformation; aneurysm

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Introduction

The internal jugular vein (IJV) is a large, bilateral vessel that drains blood from the dural venous sinuses of the brain as well as smaller veins of the head and neck area. IJVs run downwards along the internal and common carotid arteries in the carotid sheath to join the subclavian veins, forming the brachiocephalic veins. The Best-known anatomical variants of the IJV are fenestration and duplication. Both anomalies occur in circa 1–2% of the population — with IJV duplication prevalence estimated at around 0.4%, and are usually diagnosed intraoperatively or during anatomical dissections [1–6]. Occasionally these malformations can be found in diagnostic imaging examinations, mainly CT or MRI with intravenous (i.v.) contrast administration [7–9]. IJV malformations are likely the result of developmental abnormalities appearing in neuronal, vascular, or bone structures of the neck area and are of little clinical significance, except for local surgical and vascular procedures [10].

Case report

A 74-year-old woman with a history of arterial hypertension and pre-diabetes was admitted to the hospital with the suspicion of transient global amnesia (TGA) or transient ischemic attack (TIA). Non-contrast CT of the patient's brain showed mild neurodegenerative changes and a lesion suspected of a meningioma near the cerebral falx in the right frontal lobe. Subsequently, brain MRI (Siemens Magnetom Skyra 3T) followed by angio-CT (Siemens Somatom Definition Edge, 80 ml of Omnipaque 350 i.v.) of the head and neck was performed that excluded the meningioma and confirmed the unruptured, saccular aneurysm in the distal segment of the right anterior cerebral artery (ACA) measuring 7.3 × 7.8 × 6.5 mm (AP × SD × CC). In addition, four more vascular malformations were revealed: 1) additional left

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ACA departing from segment A1/A2, 2) fetal left posterior communicating artery (PComA) with a hypoplastic P1 segment, 3) the presence of right IJV duplication and 4) fenestration of the left IJV (Figs. 1–3).

The duplication of right IJV occurred at the level of C3 vertebra. The lateral branch of the duplicated IJV then passed through the deep cervical fascia behind the sternocleidomastoid muscle (SCM) to follow the external jugular vein (EJV) posteriorly and join the right subclavian vein (SV) independently of the junction of the medial branch of the right IJV.

Left IJV fenestration began at the level of C2/C3 intervertebral disc to rejoin at the C5/C6 level, measuring 45 mm in length. The left internal carotid artery (ICA) bulged into the fenestration lumen.

The patient was discharged with a recommendation for treatment at a neurological clinic and scheduled embolization of the right ACA aneurysm.

Discussion

IJV malformations are usually asymptomatic but surgically significant [11]. Most of the reported developmental IJV anomalies were diagnosed intraoperatively. In one case, IJV fenestration was diagnosed as a result of an accidental incision of one of its branches, which resulted in profuse hemorrhage [6]. The awareness of the presence of IJV malformations can prevent complications during surgery and vascular procedures such as inserting central venous catheters (CVC). Pre-operative radiological evaluation of vascular structures of the neck could be beneficial, although in the case of pathological neck masses additional IJV branches might be compressed and unnoticeable in the imaging examinations [12].

In order to better understand the morphology of IJV duplications and to assess their complexity, a classification was proposed by Nayak et al. [10]. This classification introduces three types of duplications based on the level of IJV duplication and eventual reconnection of IJV branches in relation to the anatomical structures of the neck. Type A, estimated to occur in 75% of cases, refers to the malformation pattern in which IJV begins with two separate branches which then reunite and drain into the subclavian vein as a single vessel. The presented case represents an uncommon type C duplication of the right IJV, the morphology of which is inverse to type A, and may be associated with a higher risk of complications from surgery. Information about IJVs malformations in this patient is particularly important due to the planned right ACA aneurysm embolization — a procedure performed under general anesthesia and requiring insertion of a CVC.

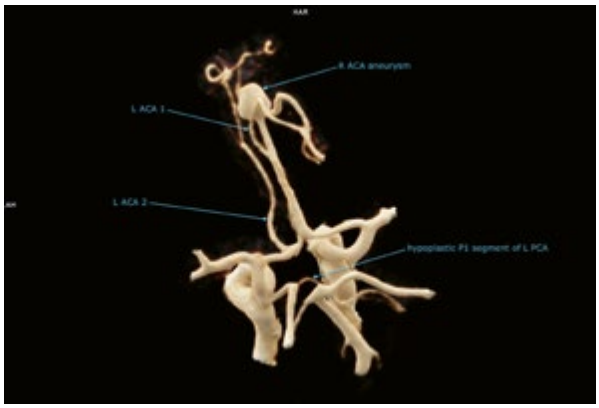

Figure 1. Angio CT in 3-dimensional reconstruction showing right anterior cerebral artery (R ACA) aneurysm, two left anterior cerebral arteries (L ACA 1 and 2), and a hypoplastic P1 segment of the left posterior cerebral artery (L PCA)

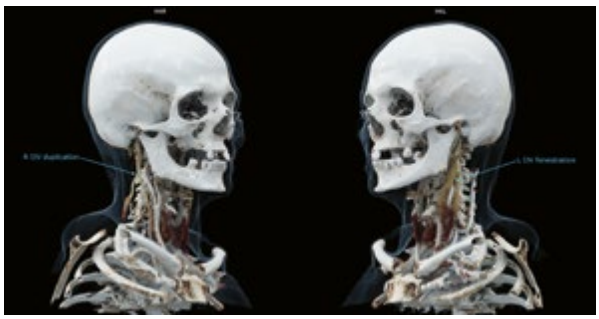

Figure 2. Angio CT in 3-dimensional reconstructions showing right internal jugular vein (R IJV) duplication and left internal jugular vein (L IJV) fenestration

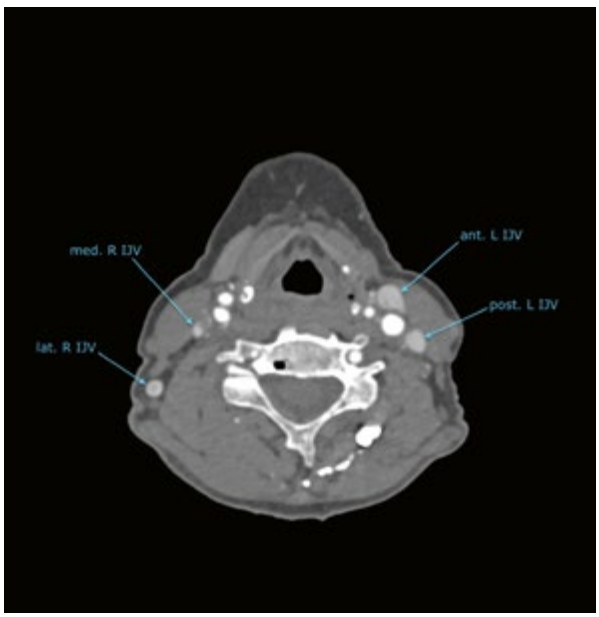

Figure 3. Angio CT in axial plane showing lateral branch (lat.) and medial branch (med.) of right internal jugular vein (R IJV) and anterior branch (ant.) and posterior branch (post.) of left internal jugular vein (L IJV). The medial branch of the right IJV is passing through the deep cervical fascia behind the sternocleidomastoid muscle (SCM)

The available data suggest the increased prevalence of other vascular malformations coexisting with IJV duplications and fenestrations. However, only a few studies reported other concomitant vascular findings such as IJV phlebectasia and bovine aortic arch [2, 4]. The presented case report supports the thesis that both duplication and fenestration of IJV accompany other vascular anomalies, in particular involving the cerebral arteries. This correlation may be of importance in patients initially undergoing interventional radiology procedures on cerebral vessels i.e. digital subtraction angiography (DSA), thrombectomy or aneurysm embolization. In the case of diagnosed pathologies in the cerebral vessels paying more attention to the detection of possible other vascular anomalies, including those in the venous system, could translate into a reduction in complications during the insertion of the CVC and the procedure itself.

Conclusions

This is the first reported case of both fenestration and duplication of the contralateral IJVs and one of the few that confirm the coexistence of vascular malformations in the IJV and other vessels — in this case, the aneurysm of the right ACA, accessory left ACA and fetal left PComA with a hypoplastic P1 segment.

The awareness of the presence of IJV malformations can prevent complications during surgeries in the neck area and vascular procedures such as IJV catheterization.

Conflict of interest

None.

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