Radiology Case Reports 14 (2019) 452–455

Available online at www.sciencedirect.com

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Internal jugular vein agenesis: a rare vascular abnormality and incidental finding. A case of internal jugular vein agenesis in a 52-years old male

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ARTICLE INFO

Article history:
Received 5 December 2018
Revised 7 January 2019
Accepted 13 January 2019
Available online 29 January 2019

Keywords:
Jugular
Vein
Agenesis
Cannulation
Malformation

ABSTRACT

We report a case of vascular malformation arising from internal jugular vein discovered during radiological investigations for restaging of metastatic colon carcinoma of an adult male patient. Congenital absence of internal jugular vein is extremely uncommon. These developmental anomalies in general population are seen in about 0.05%-0.25%. The awareness of these vascular anomalies is extremely important to avoid unsafe complications, primarily in oncological patients, whom usually require the incannulation of neck veins for diagnostic procedures or intravenous therapy administration.

Introduction

Internal jugular vein (IJV) drains blood from the head and neck district. It originates from the sigmoid sinus after it exits from the cranial cavity through the jugular foramen, then descends in the carotid sheath, and unites within the subclavian vein to form the brachiocephalic vein, which enters the thorax to join vena cava superior [1].

Vascular anomalies are divided into 2 types: vascular tumors (for example, hemangioma, hemangioendotelioma, and angiosarcoma) and vascular malformation result from embryological developmental deformities [2].

Absence of IJV is very rare vascular anomaly and it is asymptomatic.

There are few cases reported in the literature. Only 6 cases of IJV agenesis were reported in published case reports since 2010 [2–7]. Most of the cases were accidental detected on routine check prior to central venous cannulation in patients

Abbreviations: IJV, internal jugular vein; CT, computed tomography; US, ultrasound.

Declarations of interest: None.

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https://doi.org/10.1016/j.radcr.2019.01.008

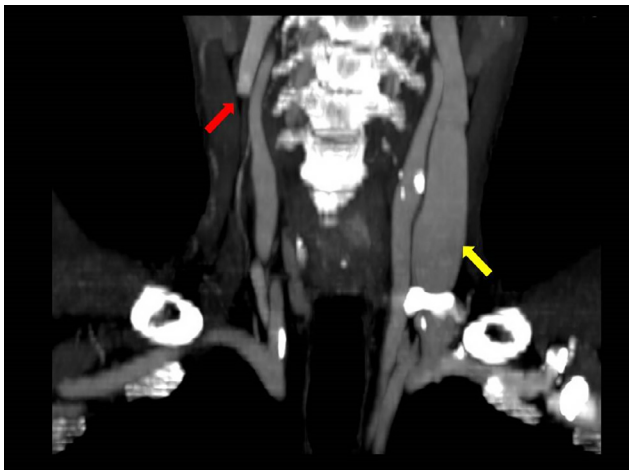

Fig. 1 – The figure shows the vasculature of neck with absence of right internal jugular vein (red arrow) and enlargement of left internal jugular vein (yellow arrow). (Color version available online.)

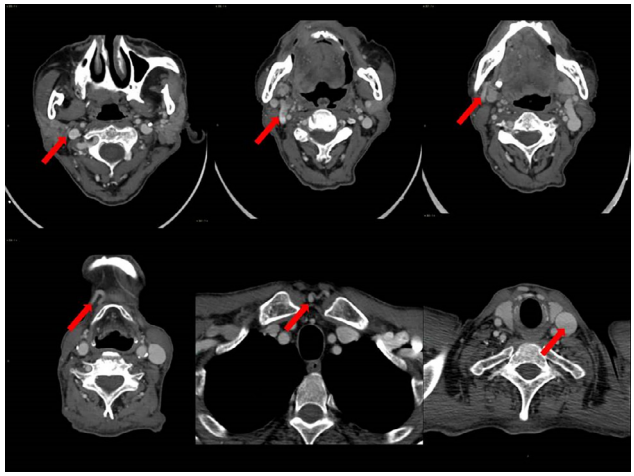

Fig. 2 – The figures shows the collateral venous circulation from the right internal jugular vein to left internal jugular vein, with ectasia of submental vein (red arrows). (Color version available online.)

undergoing major surgical procedures, for example, excision of a non-secretory pheochromocytoma [4] or right hepatectomy [3], and others were discovered during ultrasound guided vascular access [6,7]. One was found during neck dissection for a squamous cell carcinoma of the lip [5], another one during ultrasound examination of the neck for a mass [2].

We report a case of right IJV agenesis in a 52 years old cancer patient.

Case report

A 52 years old male suffered from metastatic colonic carcinoma referred to our Department for restaging.

No specific physical signs and no relevant symptoms were reported.

A subsequent contrast enhanced computed tomography (CT) examination was performed by a 64-slice CT scanner (Lightspeed; General Electric Healthcare, Waukesha, WI). The protocol included a noncontrast CT scan, with 5 mm of thickness, and dynamic acquisition at 35 and 90 seconds after administration of iodine contrast medium (Iobitridolo; volume: 120 mL; flow rate: 2.5 mL/s), slice thickness: 2.5 mm.

Enhanced CT images showed absence of opacification of the right IJV immediately after its proximal portion along the neck, in absence of intraluminal thrombosis (Fig. 1).

A collateral venous circulation vein that drains from the right to the left IJV, with compensatory left IJV phlebectasia, was demonstrated (Fig. 2). In particular, ectasia of the anterior branch of the right retromandibular vein and of the right anterior jugular vein was showed, draining blood toward the left IJV within its distal portion just before to join with the subclavian vein to form the brachiocephalic vein (Fig. 3). No other malformations were reported.

The alteration was attributed to right IJV agenesis.

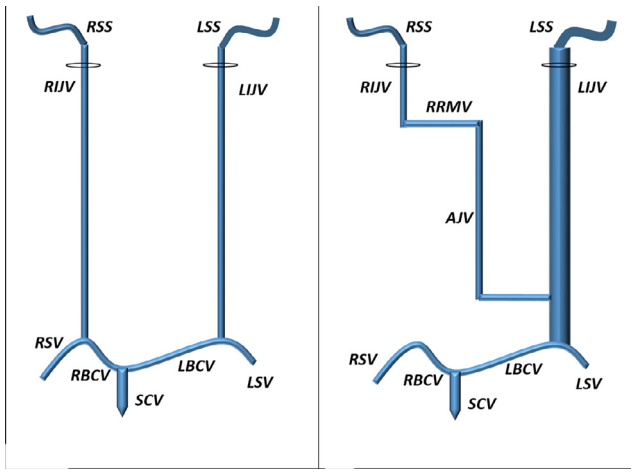

Fig. 3 – The figure shows normal internal jugular vein circulation (3a) and right internal jugular vein agenesis with anomalous venous circulation (3b): the right retromandibular vein and the right anterior jugular vein drain blood to the left internal jugular vein that is dilated. (RIJV, right internal jugular vein; LIJV, left internal jugular vein; RSS, right sigmoid sinus; LSS, left sigmoid sinus; AJV, anterior jugular vein; RRMV, right retro-mandibular vein; SCV, superior cava vein; RSV, right subclavian vein; LSV, left subclavian vein; RBCV, right brachio-cephalic vein; LBCV, left brachio-cephalic vein).

Discussion

In this paper, CT imaging of a case of IJV agenesis is reported. Early discovery of this can be useful to reduce harms during medical management of oncological patients. It is important

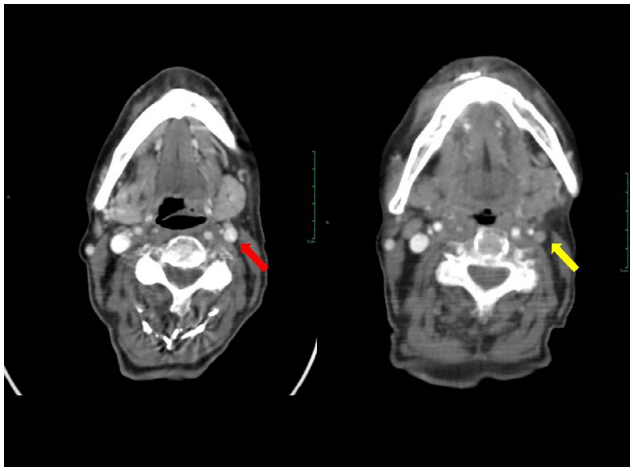

Fig. 4 – The figures shows the internal jugular vein before a thrombosis (red arrow), with regular intravascular enhance, and after a thrombosis (yellow arrow), where is showed an enhance deficit. (Color version available online.)

that the physician should be aware of the possible presence of this rare anomaly to reduce errors during surgical or vascular procedure (for example, venous cannulation).

The incidence of developmental vascular anomalies in general population are seen in about 0.05%-0.25%, however agenesis of the IJV is rarely encountered and confirmed [2]. Different IJV abnormalities have been reported in scientific literature: partial or complete duplication [8–12], stenosis, complete occlusion, distortions, and intraluminal structures, such as membranes, webs, and inverted valves [13,14], congenital external carotid artery-IJV fistula [15], congenital aneurysm [16], and fenestration [17].

Cerebral venous abnormalities in patients with cervical or facial venous malformation tend to coexist in rates up to 20% [4]. These are often diagnosed accidentally or during imaging investigations of the neck due to a suspected mass, occasionally result of compensatory contralateral IJV enlargement [2].

Contralateral IJV enlargement is usually a consequence of IJV agenesis.

There is no knowledge of clinical implication, but both IJV are used for essential medical procedures, especially in oncologic patients: cannulation, intravenous infusion, and central venous pressure monitoring.

Awareness of these venous variations is significant for clinician, anesthesiologist, and surgeons to avoid any mistakes in long-term management of patients or intraoperative surgical complications.

For example, central venous cannulation in a patient with absent IJV is associated with a higher risk of arterial puncture and pneumothorax [6]. Above all, knowledge about the IJV anomaly assumes relevance if it is bilateral because it leads to severe intracranial venous hypertension and may result in potentially cerebral edema [5].To prevent these complications, Venous Doppler assumes importance to localize IJV and it is a precautionary measure to underline vascular anomalies. A simple, focused ultrasound examination of the neck during preoperative assessment has been recommended by National Institute for Health and Care Excellence in the United Kingdom since 2002 [18].

Thrombosis or altered hemodynamic state should be considered as part of the differential diagnosis of not visualizing IJV. The incidence of not visualizing IJV with the use of ultrasound can reach up to 2.5% probably due to thrombosis (Fig. 4) [19].

The IJV agenesis must not, however, be mistaken for the above-mentioned pathological conditions. Thus, when in presence of the clinical or ultrasound suspect of IJV anomalies, we suggest the use of CT scan.

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