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High duplication of the internal jugular vein: clinical incidence in the adult and surgical consequences, a report of three clinical cases

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Abstract Duplication of the internal jugular vein (IJV) is a rare malformation. Three intraoperative cases are reported. In our personal experience, the clinical incidence of the anomaly is approximately 4 per 1,000 unilateral neck dissections. The venous duplication is at a variable height, affecting the superior part of the IJV. The lateral branch of the accessory nerve (XI) always passes medially to the anterior vein and laterally to the posterior vein, between the venous duplication. This is most often unilateral but sometimes bilateral. The IJV may be normal, dilated or ectatic. The discovery of this anatomical variation has practical implications during cervical lymph node clearance, either functional or radical, during oncological surgery necessitating viewing the IJV and its affluents and the lateral branch of the accessory nerve. The embryological explanation suggests a topographical “conflict” between the development of the IJV and the lateral branch of the accessory nerve. The French version of this article is available in the form of electronic supplementary material and can be obtained by using the Springer LINK server located at <http://dx.doi.org/10.1007/s00276-002-0020-y>

Duplication haute de la veine jugulaire interne: prévalence, clinique chez l'adulte et conséquences chirurgicales, à propos des 3 cas cliniques

Résumé La duplication de la veine jugulaire interne (VJI) est une malformation rare. Trois observations opératoires sont rapportées. Dans notre expérience personnelle, l'incidence clinique de cette anomalie est approximativement de 4 pour 1000 dissections cervicales unilatérales. La duplication veineuse est de hauteur variable, touchant la partie supérieure de la VJI. La branche latérale du nerf accessoire (XI) passe toujours médialement à la veine antérieure, latéralement à la veine postérieure, à travers la duplication veineuse. Celle-ci est le plus souvent unilatérale, parfois bilatérale. La VJI peut être normale, dilatée ou phlébectasique. La découverte de cette variation anatomique a des implications pratiques lors des curages celluloganglionnaires cervicaux, fonctionnels ou radicaux à visée carcinologique, imposant le repérage de la VJI, de ses affluents et de la branche latérale du nerf accessoire. L'explication embryologique privilégie un «conflit» topographique de développement de la VJI et de la branche latérale du XI.

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Keywords Internal jugular vein · Accessory nerve · Cervical lymph node clearance

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The jugular vv. (from the Latin: *jugulum*, throat) are the collecting vessels of the cranium, the face and the anterior region of the neck. The internal jugular v. (IJV) or the carotid v. of Sébileau is the principal deep vein of the neck and the return route for intracranial blood. It rises in the jugular foramen where it is the continuation of the sigmoid sinus running through the posterior subparotid space then the carotid gutter, describing a spiral around the arterial axis. It ends at the base of the neck in front of the sternoclavicular a. by uniting with the subclavian v. [2, 12, 18]. There are two dilatations in its course: the superior bulb of the IJV or the jugular bulb of Haller lying in the jugular fossa of the cranium, and the oval inferior bulb of the IJV just before its end.

The IJV and its affluents, notably the thyro-linguo-facial trunk (of Farabeuf), constitute an essential landmark during oncological cervical lymph node clearance [1, 13]. The demonstration intraoperatively or by imaging of duplication of the IJV is rare, reported episodically in the literature (Table 1). This malformation raises both clinical and embryological problems [4].

We report the observations on three patients operated on during the last 5 years for cervical lymph node clearance in whom we discovered duplication of the IJV.

Case reports

Case 1

A man born 21 October 1928 presented with an endolaryngeal epidermoid carcinoma affecting the anterior commissure and the anterior third of the two vocal cords without fixation. Infra-centimetric bilateral subdigastric satellite lymph nodes were seen (T₂, N₂b). Reconstructive laryngectomy with bilateral cervical lymph node clearance was performed. The cervical dissection discovered duplication of the IJV on the left, 5 cm in length, between the exocranial aspect of the jugular foramen down to the anastomosis of the thyro-facial venous trunk of Farabeuf. The lateral branch of the accessory n. passed medially to the more voluminous anterior vein and laterally to the posterior vein. We also noted a relative dilatation of the jugular v. with respect to the contralateral IJV, which was normal. The procedure preserved the two left-sided venous trunks and the accessory n. (Fig. 1).

Case 2

A man born 26 June 1949 presented with an epidermoid carcinoma of the glosso-epiglottic vallecula and of the epiglottis on the left side with centimetric subdigastric lymphadenopathy (T₂N₁). A horizontal supraglottic partial laryngectomy was done together with bilateral lymph node clearance. The cervical dissection on the left demonstrated duplication of the IJV over the 3 cm above the thyrofacial trunk (of Farabeuf). There was no venous dilatation. Between the exocranial aspect of the jugular foramen, the lateral branch of the accessory n. passed between the two veins, medially with respect to the larger more anterior vein and laterally to the



Fig. 1. Case 1. Duplication of the left internal jugular vein (IJV) (*open arrows*) from the base of the cranium above to the superior border of the linguo-facial venous trunk (*blocked arrow*). Intravenous passage of the accessory n. (*XI*)

more posterior vein. The lymph node dissection was done with preservation of the neurovascular structures. On the right, the IJV was normal. MR angiography done after the surgical procedure

Table 1. Duplication of the internal jugular vein (IJV): literature data

1 case Total duplication of the IJV on the right and ipsilateral inferior ectasia	Zukschwerdt, 1929 [19]	Fortuitous intraoperative discovery
1 case Duplication of the superior IJV on the right linked to ipsilateral inferior ectasia	Som et al., 1985 [15]	Basi-cervical swelling linked to the ectasia – imaging (CT)
1 case Isolated superior duplication of the IJV on the right.	P Sylaidis et al. (1997)	Fortuitous intra-operative discovery during node clearance
1 case Isolated total duplication of the IJV on the right	Munoz-Guerra et al., 2000 [11]	Fortuitous intraoperative discovery during nodal clearance
1 bilateral case Duplication of the superior IJV on the left and complete duplication of the IJV on the right with inferior ectasia	Rossi et al., 2001 [14]	Basi-cervical swelling on the right linked to the ectasia – imaging (MRI)

demonstrated the functional character of the venous duplication and showed a single sigmoid sinus (Fig. 2).

Case 3

A man born 12 April 1939 presented with an epidermoid carcinoma of the right piriform sinus fixed to the glottic plane on the same side with a voluminous 4 cm diameter subgastric lymph node mass (T₃N₃). A radical right lymph node clearance was done including the IJV, the lateral branch of the accessory n. and the sternocleidomastoid m. A dissection of the accessory n. showed duplication of the superior part of the IJV over 2 cm at the exocranial inferior aspect of the jugular foramen. The lateral branch of the accessory n. passed medially to the anterior vein and laterally to the posterior vein. There was no ectasia of the jugular. The two veins were ligated at the exocranial aspect of the jugular foramen with division of the accessory n., allowing liberation of the metastatic nodal mass. Histopathological examination did not reveal any venous parietal anomaly in the internal jugular duplication.

Discussion

Duplication of the IJV seems to be a rare anatomical variation. Several isolated cases have been reported in

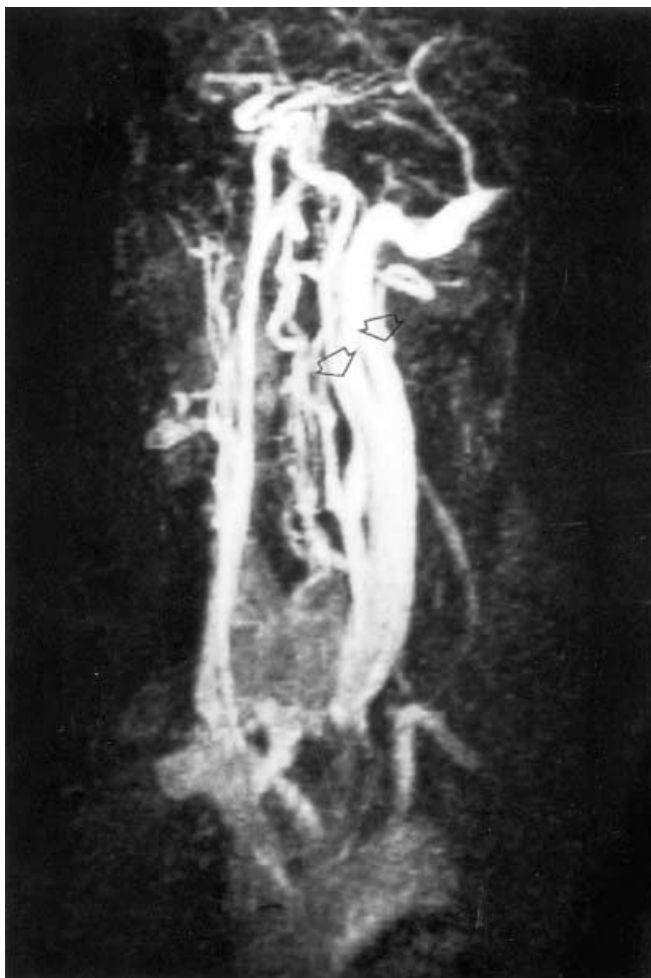


Fig. 2. Case 2. Postoperative MR angiography shows functional duplication of the IJV (*open arrows*). The sigmoid sinus is normal

the medical literature [11, 14, 15, 17]. In our personal experience of neck dissection during oncological lymph node dissection, the landmark of the IJV and its affluents, of the lateral branch of the accessory n., of the sternocleidomastoid m. and the cervical plexus is constant [1, 13]. Over the last 5 years, 150 unilateral jugulo-carotid node dissections have been done per year and three patients presented with duplication of the IJV during dissection. The estimated incidence is, therefore, approximately 4 per 1,000. It does not seem to have a preferential side. The venous anomaly may be bilateral (a single case reported), most often superior and more rarely total [14].

In all cases, the lateral branch of the accessory n. passed between the two internal jugular vv. The variable relationships of the lateral branch of the accessory n. with the IJV are important to know for cervical lymph node clearance. The accessory n. crosses the IJV at the level of the transverse process of the atlas in an anterolateral position in 56–90% of cases, and in a posteromedial position in 10–44% of cases [1, 6, 12]. In a prospective in vivo study of 123 patients, Levy et al. [9] emphasized the rarity (0.8% of cases) of the medial position of the nerve with respect to the IJV. The possibility of a transvenous course of the accessory n. and of duplication of the IJV has been noted in the anatomical treatises [5, 12, 18] of Hovelacque (1927), Testut and Latarjet (1948) and Paturet (1958). Krmpotic-Nemanic et al. [7] did not mention it in the possible variations of the IJV. In our three cases, the lateral branch of the accessory n. always passed medially to the anterior vein and laterally to the posterior vein. The duplication always involved the upper third of the IJV from the exocranial aspect of the jugular foramen. The duplication extended inferiorly in a variable fashion.

The discovery of duplication of the IJV poses practical problems of surgical dissection during functional node clearance preserving the neurovascular structures or during radical nodal clearance requiring ligation of the two venous trunks and division of the lateral branch of the accessory n. [6, 13].

Other than ectasia of the IJV, most often inferior, no other cervical venous anomaly has been seen [14, 16]. Nevertheless, our first case showed a diffuse dilation of the internal jugular venous system and its branches without any true ectasia (Fig. 1). We could not demonstrate any obstruction but there were parietal venous anomalies involving the smooth muscle, the elastic fibers or represented by abnormal fatty deposits which suggest a congenital origin [16].

Embryology does not clearly explain the venous duplications [8, 10]. Three hypotheses – vascular, neural or bony – may nonetheless be considered. The vascular hypothesis is most likely. It cites as the cause of the duplication the formation of the internal venous jugular access. In general, the nerves appear after the vessels and venous duplication could result from inadequate condensation of the embryonic capillary plexus. Tandler, in [5], described a lateral and medial vein of the head

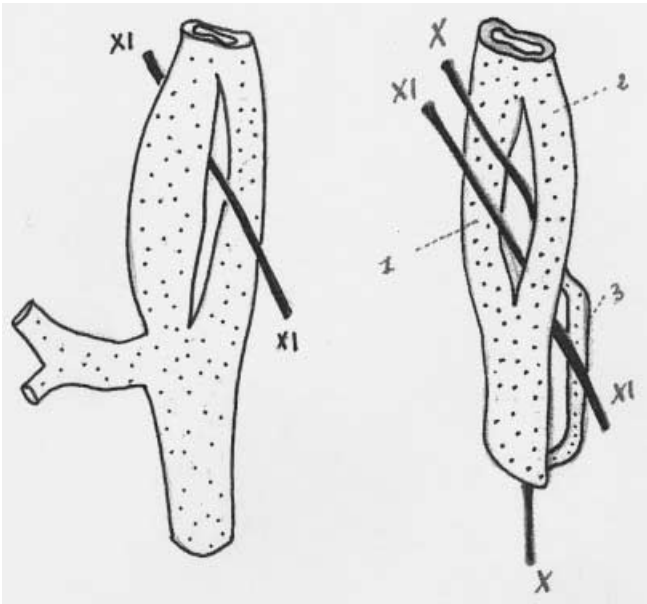


Fig. 3a, b. Duplications seen and organogenesis of the internal jugular v. (IJV) according to Tandler, in [5]. **a** High asymmetrical duplication of the IJV above the linguo-facial venous trunk. The lateral branch of the accessory n. runs behind the anterior vein. **b** The lateral branch of the accessory n. and the vagus n. (X) runs behind the anterior vein. 1, Medial vein of the head; 2, lateral vein of the head; 3, secondary venous ring which is the origin of the duplication of the asymmetrical IJV

associated with an accessory venous ring. The lateral vein or more rarely the medial vein disappears, explaining the topographical variations of the lateral branch of the accessory n. with respect to the IJV. If the secondary venous ring persists, duplication of the IJV is seen with transvenous passage of the nerve (Fig. 3A, B).

The neural hypothesis is linked to migration of the lateral branch of the accessory n. itself. There is, in effect, a vertical topographical variation of the accessory n. The nerve is, in general, at the level of the transverse process of the atlas but may lie up to 2 cm below it according to Sébilleau, in [5], and be in conflict with the IJV during its formation.

The bony hypothesis underlines the variations in ossification of the jugular foramen being the origin of the bony bridges responsible for variable partitioning [3]. These bony partitions involve the jugular foramen itself but do not explain the topographical variations of the lateral branch of the accessory n. with respect to the IJV.

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