CASE REPORT

Duplication of the Internal Jugular Vein: A Rare Case Report with a Review of Literature

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Abstract

Anatomical variations of the Internal Jugular Vein (IJV), such as duplication, are rare but clinically significant findings. A lack of awareness of these anomalies can lead to iatrogenic vascular or nerve injury during head and neck procedures. We report a case of IJV duplication discovered during a neck dissection for thyroid carcinoma.

A 27-year-old female with papillary thyroid carcinoma and cervical lymph node metastases underwent a total thyroidectomy with a right-sided lateral compartment neck dissection. Intraoperatively, a duplication of the right IJV was discovered. The duplication originated at the level of the omohyoid muscle, with two trunks of equal calibre draining separately into the subclavian vein. This anatomical variation presented a challenge during the dissection of Level II-V Lymph nodes.

The neck dissection was completed successfully with the preservation of both IJV trunks, and the patient's postoperative course was uneventful. This case highlights the surgical importance of IJV duplication. Preoperative identification of such anomalies through careful review of imaging is crucial for surgical planning to prevent complications.

Keywords: internal jugular vein duplication; anatomical variations

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Introduction

The Internal Jugular Vein (IJV) collects blood from the cranium, face, and neck. It originates from the jugular foramen and joins with the subclavian vein to form the brachiocephalic vein. As the largest vessel in the neck, it is a major landmark for adjacent structures such as the carotid artery, the vagus trunk, cervical lymph nodes, and the spinal accessory nerve during head and neck surgeries. A thorough understanding of IJV anomalies is crucial for preventing iatrogenic damage and unexpected surgical complications during procedures like neck dissections and central venous catheterization. Anomalies of the IJV, such as fenestrations and duplications, are rare, with a reported clinical prevalence of approximately 1% (1). Therefore, we present a case of IJV duplication to highlight its clinical value.

Case Report

A 27-year-old female patient with a history of type 2 diabetes mellitus and a recent diagnosis of papillary thyroid carcinoma with cervical lymph nodes metastases underwent a total thyroidectomy with a right-sided lateral compartment neck dissection. During the neck dissection collar incision extended right laterally and superior and inferior flap raised. Upper jugular nodes (Level II) from skull base to hyoid was dissected by retracting Sternocleidomastoid muscle laterally, identifying IJV and fibrofatty package sweeping laterally and inferiorly (2). Spinal accessory nerve identified early as it crosses obliquely through this level and preserved. Dissection was continued inferiorly along the IJV to dissect Level III - Mid jugular nodes (from hyoid to cricoid). Omohyoid muscle divided to improve access and marks the Level III/IV boundary. At this point a right internal jugular vein duplication was identified. The duplication originated at the level of the omohyoid muscle, and both trunks drained into the subclavian vein posterior to the clavicle. Both trunks were of equal calibre. Level III-V lymph nodes dissection was difficult due to IJV duplication, but neck dissection was continued inferiorly along the duplicated IJV preserving both IJV trunks. Level IV lymph nodes were extracted lateral and in between divided IJV trunks. Phrenic nerve on scalenus anterior preserved by keeping prevertebral fascia as the deep limit at all levels. Posterior triangle (Level V) dissection continued behind SCM to trapezius. SCM retracted anteriorly and dissection carried over prevertebral fascia. Spinal accessory nerve traced across the posterior triangle to trapezius. Cervical plexus branches divided at Erb's point. Closed suction drain applied at closure (Figure 1).

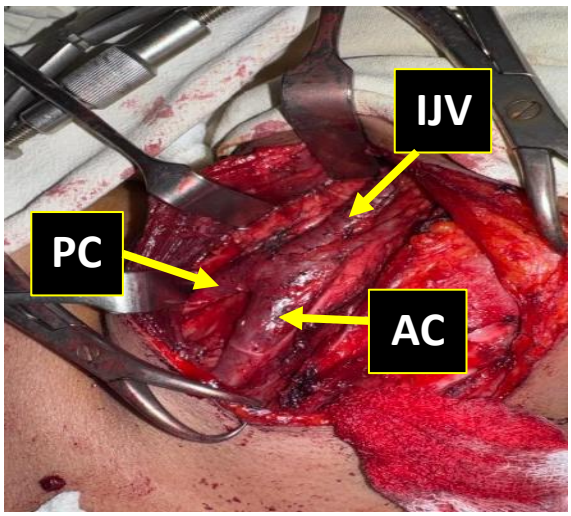


Figure 1: Intraoperative photograph of Internal jugular vein duplication right side. AC = Anterior Component; PC = Posterior Component; IJV = Internal Jugular Vein. The top-right corner of the image represents the superior aspect (cephalad), and the bottom-left represents the inferior aspect (caudad).

Discussion

Anomalies of the Internal Jugular Vein (IJV) are a rare phenomenon (3). The exact prevalence is difficult to determine due to this rarity and under reported cases (4-8). These anatomical variations are most often identified intraoperatively, with fewer cases detected through radiological imaging or during cadaveric dissection (1). These may present in the form of duplication or fenestration. IJV duplication is defined as a bifurcation of IJV, where each resulting branch joins the subclavian vein separately, whereas in an IJV fenestration, the divided branches reunite before draining into the subclavian vein (8). The spinal accessory nerve (SAN) may pass through a fenestration in the IJV with only a few reported cases in the literature (10). Some ambiguity exists among the head and neck surgeons regarding the terms 'bifurcation' and 'duplication' of the IJV. Some authors suggest the term 'bifurcation' when the IJV splits above the level of the omohyoid muscle (10). Shukla and Agarwal reported 7 cases of IJV duplication in over 350 neck dissections performed over two years (11). This corresponds to a prevalence of 2% in their study. Prades JM *et al.,* estimated clinical prevalence of IJV duplication to be 0.4% and Wang *et al.,* estimated prevalence of IJV duplication as 0.9% (13, 18) (Table 1).

Classification of IJV anomalies

Several classification systems for IJV anomalies exist, but one of the most widely accepted was published by Sandeep Nayak *et al.,* (1) (Figure 2).

Type A: The duplication begins above the inferior border of the posterior belly of the digastric muscle and reunites at the level of the omohyoid muscle. This accounts for more than 75 % of all the Internal Jugular Vein Duplications that have been reported. This type is considered the least complex. This configuration, where the divided vein reunites, is also referred to as an IJV fenestration (1).

Type B: This type differs from Type A only in its caudal extension, with the duplication extending inferior to the omohyoid tendon.

Type C: The duplication starts at or below the level of the hyoid bone and is considered more intricate. The lateral component takes a course outside the carotid sheath and lateral to the omohyoid muscle, traversing the posterior triangle of neck, before curving and entering the carotid sheath at the root of the neck. A case of bilateral Type C IJVD has been reported by Downie SA *et al.,* (12), in which one side developed a large, bulbous jugulo-vertebro-subclavian venous sinus.

These Type B and C anomalies are considered the most difficult to manage clinically, because IJV below the omohyoid tendon often use for catheterization and in neck dissections presence of a distal duplication can pose difficulties both in preserving vein and ligation. Our case is classified as a Type C anomaly due to the distal extent of the duplication. However, it is noteworthy that the division in the reported case began at the level of the omohyoid tendon, not the hyoid bone as described in the classification. This highlights that anatomical variations exist within these classification types and that the landmarks may not be absolute.

Aetiology

The exact aetiology of IJV duplication remains unknown; however, four main theories have been proposed.

Vascular hypothesis

The development of the neurocranium, cranial nerves, and cervical venous system occurs

between the third and eighth weeks of gestation. The duplication is thought to arise when secondary venous channels fail to regress, leaving two distinct veins. The subsequent elongation of the fetal neck and the downward migration of the myotome and accessory nerve further extend the length of this duplicated segment (1, 13).

Neuronal hypothesis

This theory suggests that primitive embryonic venous capillary networks inadvertently entrap developing supplementary neurons. This neurovascular entrapment splits the developing vein into anterior and posterior segments. Similar to the vascular theory, the natural caudal migration of the accessory nerve and myotome as the embryonic neck elongates stretches this localized splitting into a longer duplication (13).

Bony hypothesis

This theory links the anomaly to the bony development of the skull base, specifically during the ossification of the neurocranium which initiates around the fourth gestational week and is largely complete at birth. Variations such as duplication or triplication of the jugular foramen can physically divide the venous outflow at the skull base, leading to a duplicated IJV in the neck (3).

Table 1: Prevalence of anatomical variations of the Internal jugular Vein in world literature

Author	Study population	Number of variations	Type of variation	Prevalence
Prades *et al.,* (12)	750	3	Duplication	0.4%
Wang X *et al.,* (17)	221	2	Fenestration & duplication	0.9%
Hashimoto *et al.,* (18)	123	4	Fenestration	3.3%
Contrera *et al.,* (8)	295	3	Fenestration, duplication and bifurcation	1%
Shukla I and Agarwal (10)	350	7	Duplication	2%

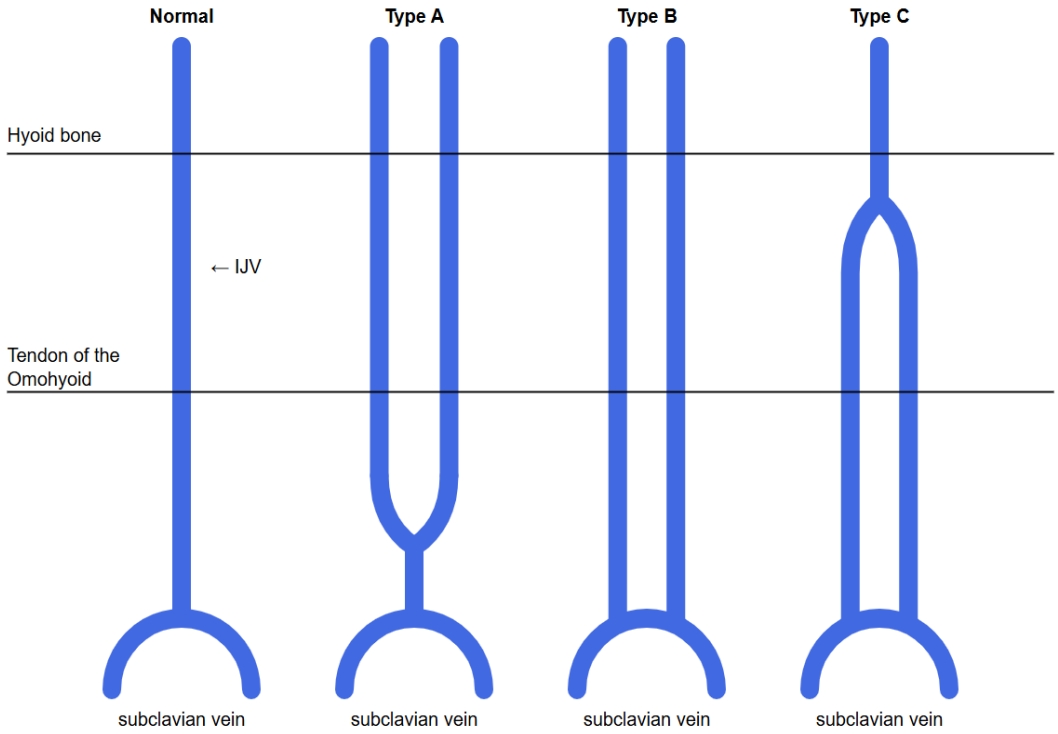


Figure 2: Pictorial representation of the classification of the internal jugular vein (IJV) duplication

Muscular hypothesis

This hypothesis proposes that IJV duplication is caused by the posterior belly of the omohyoid muscle (14).

Clinical Implication

IJV anatomical anomalies (fenestration and duplication) can be identified preoperatively by computed tomographic angiography (CTA) or during contrast enhancement CT imaging as part of clinical and radiological staging of patients. Towbin and Kanal reported two cases of IJV fenestration identified through CTA (15). However, despite the ability to identify these anomalies through contrast-enhanced CT imaging, it is seldom identified, which may reflect a lack of attention paid to the venous system in the radiological staging of patients (10).

Though IJV Duplications unlikely to cause physiological implications it can lead to difficulties during clinical interventions. Very rare cases of symptomatic IJV duplications reported in the literature, including neck swelling and dysphagia or dyspnoea. Therefore, this subset of patients might be misdiagnosed with laryngoceles or branchial cleft cysts (4).

The internal jugular vein is a common site for percutaneous cannulation. The vein is accessed between the two heads on sternocleidomastoid muscle or at the cricoid cartilage level (16). In an undiagnosed patient, this variation (especially Types B and C) can be problematic. An image guided procedure would be ideal for all patients needing percutaneous IJV

catheterization. Secondary vascular abnormalities have been reported associated with IJV duplications (17). For example, a case of a bovine aortic arch associated with a Type C IJV duplication has been reported. Therefore, consideration of contrast enhanced CT in a patient with IJV duplication is warranted if the patient undergoing any procedure involving major vessels.

An internal jugular vein has consistent anatomical course in neck. Knowledge of IJV anomalies is critical for preventing iatrogenic injury during neck dissections. In neck dissections depending on the indication lymph nodes around the IJV are removed with or without vein itself, spinal accessory nerve, sternocleidomastoid muscle in order to reduce post-operative morbidity (1). During neck dissections where ligation of IJV is essential identifying and isolating both components of the duplicated IJV is necessary to control bleeding, because copious bleeding can occur due to multiple feeding vessels from duplicated IJV (10).

Preservation of the spinal accessory nerve is important for reducing functional complications. The risk of injury to this nerve is high, as it often passes through an IJV fenestration.

Conclusion

Internal jugular vein anomalies (duplication and fenestration) are rare. These anomalies are most often identified intraoperatively, with only a few cases detected radiologically. Due to the risk of iatrogenic vascular and spinal accessory nerve injury, it is crucial to identify these anomalies preoperatively. Furthermore, because other vascular anomalies can be associated with IJV duplication, correct documentation following its identification is essential to prevent future complications.

Consent: Informed consent was obtained from the patient for the publication of this case report and any accompanying images.

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