

Radiological Findings in a Case of Internal Jugular Vein Duplication Associated with Thyroid Hemiagenesis

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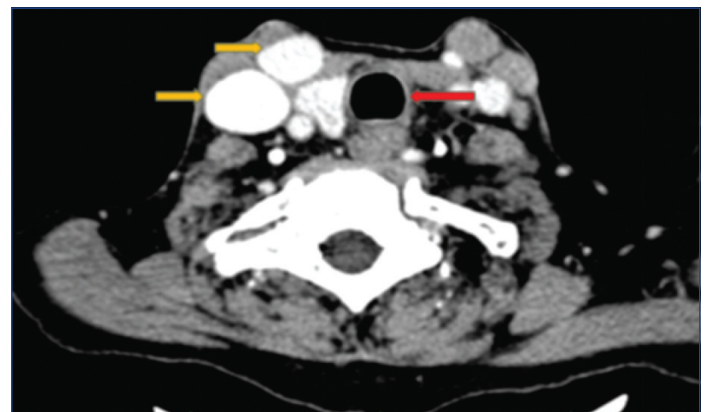
ABSTRACT

The Internal Jugular Vein (IJV) is the principal vein draining blood from either side of the head and neck region towards the heart. The thyroid gland, one of the endocrine glands in the body, lies in close proximity to the IJV and is drained by its tributaries. One variation of the IJV, in the form of duplication, can be traced embryologically during the development of the cardinal veins. The present case discusses the findings from Contrast Enhanced Computed Tomography (CECT), which show the duplication of the IJV accompanied by contralateral hemiagenesis of the thyroid gland. The embryological basis for these variations needs to be highlighted separately. Duplication of the IJV is hypothesised based on several theories, including venous, neuronal and bony factors. The agenesis of the thyroid gland is noted during the lobulisation phase of its development. Identifying such variations is crucial to differentiate them from other surgical and radiological pathologies. During invasive therapeutic and diagnostic procedures in the head and neck region, these superficial structures are frequently encountered. Therefore, knowledge of the various variations and anomalies of these vital structures plays a pivotal role in planning for both emergency and elective procedures in the head and neck region. In modern medicine, this case contributes to the rare instances that can be encountered during routine practice.

Keywords: Agenesis, Cardinal veins, Contralateral, Duplicate, Hypothesis

CASE REPORT

A 62-year-old lady with no prior known co-morbidities presented with a history of hoarseness of voice lasting four months. Based on this history, the patient was evaluated through indirect laryngoscopy, which revealed a cystic lesion on the left-side of the epiglottis. To better understand the nature of the lesion, the patient was referred for a Contrast Enhanced Computed Tomography (CECT) scan of the neck. According to the radiological findings, there was duplication of the right Internal Jugular Vein (IJV) at the level of the C3 vertebra [Table/Fig-1], and it remained separate throughout its entire course. Both veins were noted to drain separately into the right subclavian vein, classified as Type-B IJV duplication according to Nayak SK et al., [1]. These findings depicted on CECT were incidental during the evaluation. The left lobe of the thyroid was not visualised [Table/Fig-2]. An exophytic cystic lesion was observed arising from the lingual surface of the epiglottis. Histopathological analysis of the lesion showed normal mitotic activity, and it was subsequently excised. Thyroid function tests were within normal limits. The patient had not undergone any thyroid-related procedures in the past. The



[Table/Fig-2]: CECT neck axial images showing agenesi of the left lobe of thyroid (red arrow) and duplication of right IJV (yellow arrows).

presence of any ectopic thyroid tissue was ruled out by conducting a thyroid uptake study.

DISCUSSION

The IJV is one of the principal vascular structures draining blood from the superficial parts of the head and neck region [2]. The course of the IJV begins at the jugular foramen (posterior compartment) at the base of the skull. It is a continuation of the sigmoid sinus, and during its commencement, it has a dilated part called the superior bulb, which is a valveless structure. The superior bulb is related to the floor of the tympanic cavity. The IJV traverses through the neck, where it is related to the Internal Carotid Artery (ICA) and the vagus nerve medially; the latter lies in the posterior plane between both vascular structures. The IJV in its upper course is related to the ICA, whereas towards its termination, it approximates the Common Carotid Artery (CCA). The IJV, along with the carotid arteries and vagus nerve, is enclosed within the carotid sheath. Before terminating, the IJV has a dilated part known as the inferior bulb, which contains a pair of valves. The IJV drains into the subclavian vein behind the sternal end of the clavicle. Numerous veins drain into the IJV during its course



[Table/Fig-1]: CECT neck axial images showing duplication of right Internal Jugular Vein (IJV) (yellow arrows) at C3 level.

[2,3]. The left IJV is usually smaller than the right IJV [4]. Anatomical variations or anomalies can create difficulties during interventional procedures such as IJV catheterisation and neck dissections, thus increasing the rate of complications [5].

Embryologically, the thyroid is the first organ to mature developmentally [6]. The thyroid assumes its dual lobe architecture once it has reached its final destination in the neck. After its descent, the thyroglossal duct detaches from the diverticula of the thyroid once the latter reaches its ultimate location [7]. The first case of thyroid hemiagenesis was documented in India in 1962 [8]. In females, thyroid hemiagenesis, characterised by an absent left lobe, is more common [9].

IJV variants, such as duplications and fenestrations, are infrequent congenital anomalies and are usually incidental findings. The following hypothesis have been proposed to explain the embryological development of IJV duplication [1]. Firstly, the venous hypothesis suggests that the development of the neck veins occurs during the 3rd to 8th week of foetal life. This is marked by the condensation of embryonic venous capillary plexuses during the development of venous vessels. IJV duplication results from the persistence of two venous channels [10].

In the second hypothesis, the accessory nerve becomes entrapped in the embryonic venous capillary plexuses during development [11]. This results in the formation of the posterior ventral pharyngeal veins (anterior and posterior), leading to an increased length of the duplicated vessel caused by the downward migration of the accessory nerve with its corresponding myotome [3]. Lastly, the bony hypothesis suggests that the duplication of the jugular foramen during its ossification leads to IJV duplication [11]. Among the vascular, neural and bony hypothesis, the vascular hypothesis is the most preferred [12]. However, a study conducted in 1986 on the foetal skull failed to support this hypothesis for IJV duplication. The study concluded that there was only a single IJV traversing through the corresponding duplicated or triplicated jugular foramen [13].

IJV duplication can be broadly classified into three different varieties as described below. The Type-A variety involves a duplicated state of the IJV that, in its cranial part, extends from the inferior border of the posterior belly of the digastric muscle and merges into a single vascular structure at the level of the central tendon of the omohyoid until its termination. In Type-B, the rostral end is similar to that of Type-A, but the difference lies in its caudal part, where it continues in a duplicated state throughout its course. Thus, from its origin to its termination, it remains in a duplicated form. The Type-C variation is not much different from Type-A; however, its formation is the opposite, with duplication occurring at its caudal end while its cranial part remains a single vascular structure. Type-B and Type-C variations have high clinical significance as they can pose difficulties during interventional and surgical procedures [14].

Looking at the embryological aspect, the fate of the cardinal veins (anterior, common and posterior) varies with their evolutionary phase. Ultimately, the formation of IJV takes place from the distal part of the anterior cardinal vein [15]. Fagman H et al., mentioned the development of third arch vessels during the lobulation of the thyroid gland. The final localisation, along with the formation of the carotid arteries, marks the termination of this process [16]. Hemiagenesis of the thyroid can be explained by thyroid transcription factors. Factors such as TTF1, TTF2, and PAX8 are involved in the formation of the thyroid gland and its cell differentiation [17]. Transient embryonic vessels play a role in guiding and inducing the descending diverticulum of the thyroid gland. Williams syndrome and DiGeorge syndrome are both associated with anomalies related to vascular rearrangement [16]. Some authors discuss the

duplication of the IJV in relation to phlebectasia and aneurysm. In phlebectasia, there is thinning of the vessel wall, whereas in the case of an aneurysm, there is degeneration of the vessel wall following its dilation. The incomplete formation of the tunica media [18] and turbulent flow at the bifurcation [19] are descriptive explanations that point towards the justifications for the duplication of the IJV. According to the literature available to date, there is no direct association between the hemiagenesis or agenesis of the thyroid gland and the duplication of the IJV. However, the explanation for both findings can be justified independently. As of now, there are no documented past studies highlighting the duplication of the IJV alongside agenesis of the thyroid gland, making this case report the first documented study on the subject.

CONCLUSION(S)

Though IJV duplication is unlikely to cause direct mortality, understanding its anatomical variants is of paramount significance in clinical settings. Unexpected encounters with agenesis or hemiagenesis of the thyroid gland should be investigated in relation to associated vascular variations. Knowledge about the variations of the IJV and the thyroid gland will help avoid any misinterpretation in radiological evaluations. Since the IJV is often used for percutaneous catheterisation of the central line to reach the right atrium, it is important for radiologists, critical care specialists and other physicians to consider the variations and pathologies of this vascular structure. In cases of thyroid carcinoma, metastasis is often due to involvement of the IJV vessel wall. Therefore, having a sound knowledge of the different variations encountered in daily practice is essential.

REFERENCES

- [1] Nayak SK, Ashraf M, Dam A, Biswas J. Internal jugular vein duplication: Review and classification. *Indian J Surg Oncol.* 2015;8(2):222-26.
- [2] Datta AK. *Essentials of Human Anatomy (Head & Neck).* Vol-2 (6th ed) Current Books International; 2017.
- [3] Birch R, Collins P, Gray H, Standring S. *Gray's Anatomy: The anatomical basis of clinical practice.* 41st ed. Philadelphia: Elsevier Limited; 2016.
- [4] Deepak CA, Sarvadnya JJ, Sabitha KS. Variant anatomy of internal jugular vein branching. *Ann Maxillofac Surg.* 2015;5:284-86.
- [5] Hedayat F, Lauder JJ, Kyzas P, Vassiliou LV. Internal jugular vein duplication: Clinical significance for head and neck cancer ablative and reconstructive surgery. *Journal of Surgical Case Reports.* 2021;2021(7):rjab300.
- [6] Trueba SS, Aug'e J, Mattei G, Etchevers H, Martinovic J, Czernichow P, et al. PAX8, TTF1, and FOXE1 gene expression patterns during human development: new insights into human thyroid development and thyroid dysgenesis-associated malformations. *J Clin Endocrinol Metab.* 2005;90(1):455-62.
- [7] Van Vliet G. Development of the thyroid gland: lessons from congenitally hypothyroid mice and men. *Clin Genet.* 2003;63(6):445-55.
- [8] Das P. Congenital absence of one thyroid lobe. *J Ind Med Assoc.* 1962;39:302-04.
- [9] Mikosch P, Gallowitsch HJ, Kresnik E, Molnar M, Gomez I, Lind P. Thyroid hemiagenesis in an endemic goiter area diagnosed by ultrasonography: Report of sixteen patients. *Thyroid.* 1999;9(11):1075-84.
- [10] Sadler TW, Langman J. *Langman's medical embryology.* Philadelphia, Pa.: Lippincott Williams & Wilkins; 2006.
- [11] Prades J-M, Timoshenko A, Dumollard JM, Durand M, Merzougui N, Martin C. High duplication of the internal jugular vein: Clinical incidence in the adult and surgical consequences, a report of three clinical cases. *Surg Radiol Anat.* 2002;24(2):129-32.
- [12] Hashimoto Y, Otsuki N, Morimoto K, Saito M, Nibu K. Four cases of spinal accessory nerve passing through the fenestrated internal jugular vein. *Surgical and Radiologic Anatomy.* 2011;34(4):373-75.
- [13] Dodo Y. Observations on the bony bridging of the jugular foramen in man. *J Anat.* 1986;144:153-65.
- [14] Wang X, Peng L, Guo H, Hernesniemi J, Xiong X, Andrade-Barazarte H, et al. Internal jugular vein fenestration and duplication: anatomical findings, prevalence, and literature review. *Front Surg.* 2020;7:593367.
- [15] Beattie J. The importance of anomalies of the superior vena cava in man. *Can Med Assoc J.* 1931;25(3):281-84.
- [16] Fagman H, Andersson L, Nilsson M. The developing mouse thyroid: embryonic vessel contacts and parenchymal growth pattern during specification, budding, migration, and lobulation. *Dev Dyn.* 2006;235(2):444-55.
- [17] Fernández LP, López-Márquez A, Santisteban P. Thyroid transcription factors in development, differentiation and disease. *Nat Rev Endocrinol.* 2015;11(1):29-42.

- [18] Cvetko E. Unilateral fenestration of the internal jugular vein: A case report. *Surg Radiol Anat.* 2015;37(7):875-77. Doi: 10.1007/s00276-015-1431-x, indexed in Pubmed: 25875636.
- [19] Towbin AJ, Kanal E. A review of two cases of fenestrated internal jugular veins as seen by CT angiography. *AJNR Am J Neuroradiol.* 2004;25(8):1433-34. Indexed in Pubmed: 15466347.

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