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B-mode and color Doppler imaging of carotid paragangliomas in different neck regions

Antigone Delantoni[1], Apostolos Sarafopoulos[2], Aspasia Polanagnostaki[2], Kaan Orhan[3]

[1] *Department of Oral Surgery, Implant Surgery and Radiology, Faculty of Dentistry, Aristotle University of Thessaloniki, Greece*
[2] *Department of Radiology, AHEPA General Hospital, Thessaloniki, Greece*
[3] *Faculty of Dentistry, Department of Dentomaxillofacial Radiology, Ankara, Turkey*

Correspondence: Dr. Kaan Orhan, Ankara University, Faculty of Dentistry, Department of Dentomaxillofacial Radiology, Besevler, Ankara Turkey 06500; tel.: 00 90 (535) 6765010, fax: 00 90 (312) 2123954, e-mail: call53@yahoo.com

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glomus tumor, paraganglioma, Doppler ultrasonography, common carotid artery

Abstract

Glomus tumors or paragangliomas are rare benign neoplasms which are very uncommon in the head and neck region. They grow slowly from paraganglion cells, which serve various regulatory tasks in the body and form the basis of the extra-adrenal neuroendocrine system. They have benign properties, but tend to cause local invasion and expansion to adjacent anatomic structures. The majority of glomus tumor cases are incidental findings, since the clinical symptoms are mostly absent. The purpose of this paper is to distinguish the ultrasonographic features of different glomus tumors located within the neck region, based on their varying anatomical location and space occupying behavior.

Introduction

Glomus tumors (GT) or paragangliomas (PG) are rare benign neoplasms which usually occur in the upper and lower extremities. They are rare in the head and neck region, and mostly located in the jugular fossa[(1)]. They grow slowly and have benign properties, but they tend to cause local invasion and expansion to adjacent anatomic structures[(2–4)]. Since the clinical symptoms that should accompany the lesion are mostly absent, the majority of the GT cases are incidental. The aim of this paper was to present an ultrasonographic description of paragangliomas of the neck region, based on their anatomical location, imaging characteristics and space-occupying behavior. Since upon their occupancy they present in typical locations and with characteristic clinical behavior, the understanding of their symptomatology together with their imaging appearance can lead to accurate and prompt diagnosis. In most cases, their imaging evaluation utilizes multiple imaging modalities, both to evaluate the extent of the disease as well as to help guide their surgical excision.

GTs are initially diagnosed as incidental findings in many asymptotic cases. During neck US, these pathologies can be diagnosed in a routine exam. The purpose of this paper is to distinguish the ultrasonographic features of different glomus tumors located within the neck region based on their varying anatomical location and space occupying behavior.

Case reports

Two cases (Tab. 1), which will be presented, are at different locations in the neck area. The diagnosis was made with ultrasonography (US), and particularly with the use of color Doppler.

Case 1

Patient #1 was a 49-year-old female who was referred to our hospital due to thyroid hormone disorder. Color Ultrasonograph GE LOGIC 5 with a linear 12 MHz probe was used.

Tab. 1. *Demographics, medical history and symptoms of the patients*

	Case 1	Case 2
Gender	Female	Female
Age	49	55
Medical history	Hyperthyroidism, Hypertension	Hyperlipidemia, GERD
Symptoms	None	Submandibular swelling, pulsatile mass, mild pain

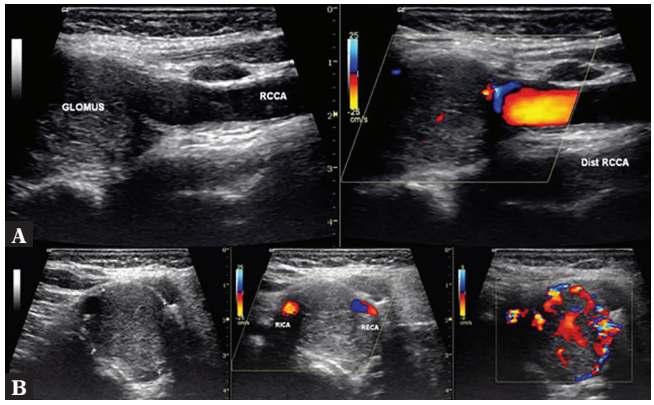

Fig. 1. A. *Horizontal sections: A hypoechoic well-delineated borders nodule of compound structure at the peripheral section of the common carotid artery.* **B.** *Vertical sections: A distinct lesion measuring 28 × 24 mm, causing dilatation of the internal and external carotid arteries in their course. Increased vascularity with the use of color Doppler from external carotid branches in their course*

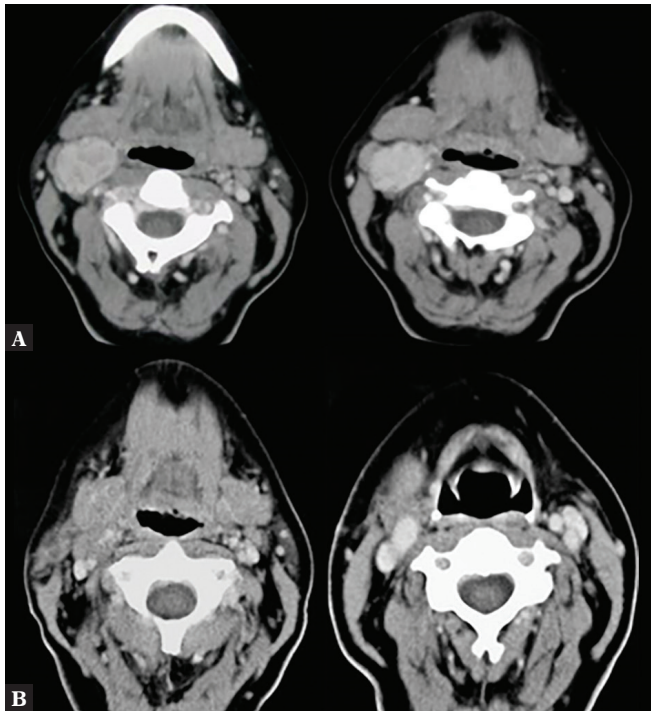

Fig. 2. A. *CT axial sections after iv enhancement: Nodule with clear distinct borders and homogeneous intake of contrast media in the arterial phase.* **B.** *Postoperative image with the lack of residual tissue as well as the lack of pathological enhancement in the area*

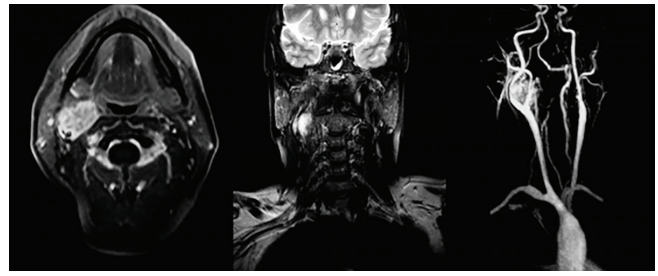

Fig. 3. *MRI axial, coronal and MRA images: High signal tumor with characteristic salt and pepper image from the dilatation of intermediate signal vessels in the lesion content*

B-mode US showed a compound, nodular lesion at the bifurcation of the right common carotid artery The lesion measured 28 mm × 24 mm, with clear border, low echogenicity and homogeneous structure. The dilatation between the bifurcation of the carotids, presented an image of a smooth repulsion with a widened space between the internal and external carotids. Triplex US imaging showed high vascularization from collateral branches rising mostly from the external carotid (Fig. 1). After the US checkup, the initial diagnosis of GT of the carotid body was set.

The patient underwent surgical excision of the lesion with excellent results that were confirmed with computed tomography (CT) at postoperative follow up (Fig. 2). Moreover, a more detailed exam including magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) was planned, and confirmed the initial findings from the ultrasound (Fig. 3).

Carotid body paragangliomas are the most common paragangliomas of the neck region accounting for about 60% of all cases[5,6]. They usually occur in older patients and arise from the carotid body, at the bifurcation of the common carotid artery. The majority of cases are unilateral, and they classically present as slow growing painless swellings of the lateral neck, often laterally mobile, but fixed vertically[7,8].

Case 2

Patient #2 was a 55-year-old female who was referred for a US examination of the neck due to a palpable swelling, pain and tension in the right submandibular area. For the US examination, a GE LOGIC S8 ultrasound machine was used with frequencies Linear 6–15 and 9 MHz.

At the cervical level of IIa triangle of the neck, a compound nodule with maximum dimensions of 38 mm × 26 mm was found adjacent to the submandibular gland. The lesion had a homogeneous echostructure and clear borders in relation to adjacent tissues and anatomical structures. Above the carotid bifurcation, a mild transposition of the internal and external carotid arteries was observed with convergence of the two arteries and a reduced space between the arteries (Fig. 4). This is different from the first case, which

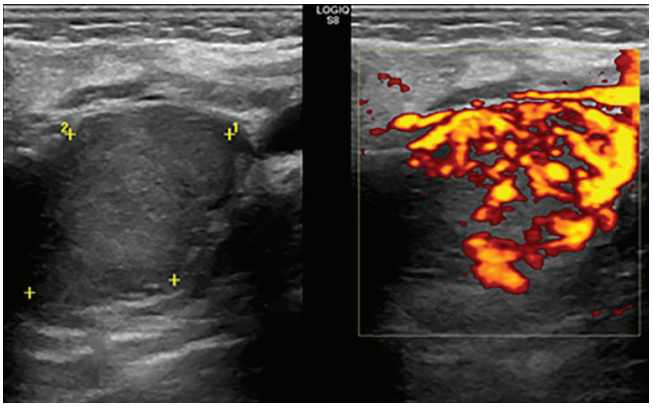

Fig. 4. *Frontal sections: Compound 38 × 26 mm tumor in the submandibular area with low echogenicity, homogeneous texture and high vascularity in Power Doppler*

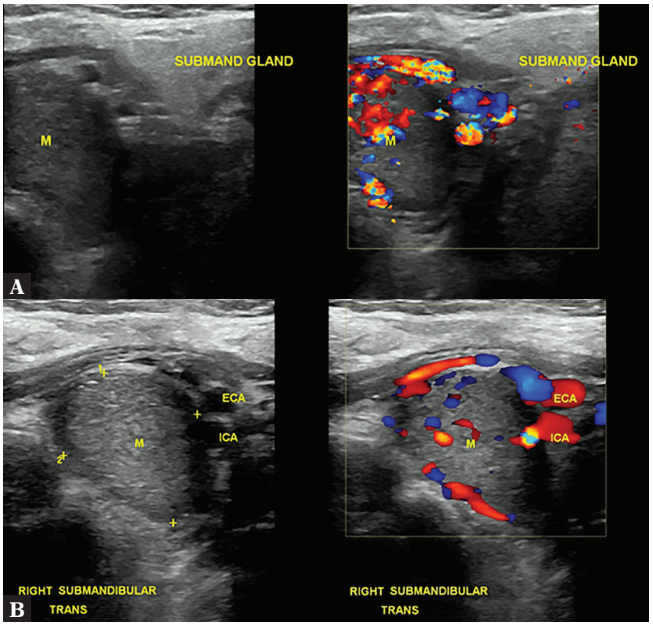

Fig. 5. A. *Angulated sections: At the level of the submandibular gland the lesion causes shift of the carotid space vessels towards the inner structures.* **B.** *Frontal sections: The GT of the vagus nerve causes alterations in the course of the internal and external carotid artery decreasing their in between distance*

demonstrated increment in the distance between the two carotids. The expansion pattern of the lesion and the higher cervical position was important in the diagnosis of different cervical locations of GTs (Fig. 5). The second patient was sent for further diagnostic imaging with MRI with diagnosis of GT of the vagal nerve (Fig. 6). The diagnosis was verified with MRI and surgical excision was performed.

Vagal paragangliomas occur within one of the ganglia of the vagus nerve (CN X). They are the least common head and neck location, accounting for approximately 5% of all cases[6,9]. They are typically seen in middle-aged patients. They typically present as a slow growing painless neck mass. However, their symptomatology differs in that both vagal nerve palsy, and voice hoarseness due to vocal cord paralysis may appear[6,10].

Differential diagnosis

Ultrasonography is a modality typically used early in the diagnosis of the majority of palpable neck masses. The differential diagnosis of paragangliomas of the carotid body from schwannomas and lymph node swellings involves the proper definition of ultrasonographic features[8,9,11]. Schwannomas arise from Schwann cells of the vagus nerves and, less commonly, the ganglia of the sympathetic chain. They are mostly benign low-echoic tumors with mainly smooth borders and with abundant vascularity in Doppler imaging. Enlarged schwannomas may show areas with cystic degeneration. The location and the way in which the adjacent vascular structures are extended or repelled, can be depicted as the main criteria for their ultrasound differentiation from the corresponding cervical paragangliomas.

Schwannomas of the vagus nerve increase in size, occupying the space between the medial carotid artery and the jugular vein, and widening the distance between them. The schwannomas of the sympathetic chain cause misalignment of the large vessels of the carotid space.

Reactive, deep cervical lymph nodes show an ultrasonic portal of adipose tissue and moderate vascularity from a passing central vascular branch. In contrast, pathological lymph nodes show an architectural disorder with decreased echogenicity, foci of necrosis, and abnormal vasculitis that most often affects the entire parenchyma. In many cases, they also tend to form oversized blocks that repel or surround large vessels.

Compared to paragangliomas, lymph node blocks have a multilobular morphology, a lobed margin and an inhomogeneous texture. Patients with abnormal lymph nodes usually have a history of other diseases, inflammation, or tumor.

Discussion

Paragangliomas of the sympathetic system or GTs are rare mesenchymal tumors that occur due to glomus body hyperplasia or hamartomatous development, and they appear to originate from modified smooth muscle cells.

They have a female prevalence of about 3:1 and mainly present in middle-aged people[2,12]. The initial clinical presentations are insidious even in cases where the tumors reach a large size. Accurate and comprehensive imaging evaluation is the key for the diagnosis of GT. Since GT is located in the skull base and is not readily viewed by ultrasound, it is most commonly found by other imaging techniques[13,14]. The preoperative diagnosis of GTs remains challenging. Inaccurate diagnoses are largely attributed to the rarity of this tumor and the lack of distinguishing clinico-morphological characteristics.

Their sonographic evaluation demonstrates a well-defined, heterogeneously hypoechoic mass, with characteristic internal vascularity with the use of color Doppler. A careful assessment of the displacement pattern of the carotid arteries can raise the suspicion for both carotid body and vagal paragangliomas.

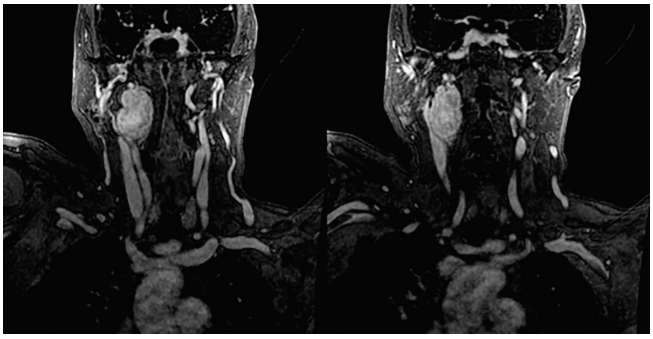

Fig. 6. *Coronal MRI T1-W sections demonstrated glomus tumor at the level of vagal nerve with high signal of the lesion with clear borders and no adjacent tissue involvement*

Furthermore, such lesions have heterogeneous appearances on radiologic images. A GT may initially be diagnosed as a salivary tumor, sebaceous cyst, neurofibromatosis, dermoid cyst, developmental tumor, vascular malformation, or another type of mesenchymal neoplasm[4]. Although vascular malformations and cystic soft tissue lesions can usually be ruled out using color Doppler ultrasonography, the differential diagnosis of solid tumors remains challenging. Recently, fluorodopa [F-DOPA] positron emission tomography was used for detecting GTs[15]; however, the validity and specificity of this technique for tumors in the head and neck region requires verification. As formal diagnostic guidelines are absent, a thorough radiographic set of examinations and studies may be needed for the diagnosis, but histological examination and immunohistochemical analysis remain the gold standards.

In the present study, the authors aimed to present the variability in locations and symptoms of the GT of the neck area. The majority of GTs of the neck are most frequently occurring at the carotid bulb and the bifurcation area, while they develop less commonly at the level of the vagal ganglion. They are most frequent in female patients, with multilocular location in 25% of the cases.

In the case of carotid bulb paraganglioma, the tumor presents as a pulsating painless swelling at the angle of the mandible, while in the cases of ganglion location, they usually present with neuralgic symptoms. A large number of tumors may produce no symptoms[16] and appear as incidental findings on US.

Conclusion

In conclusion, color Doppler is an easy readily available technique that can locate, isolate and, to a certain degree, set the diagnosis of neck paragangliomas. The vasculature level of the tumor with Triplex in combination with more detailed imaging techniques such as MRI and MRA set and compose the additional steps to a final definite diagnosis.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organizations that might adversely affect the content of this publication or claim any right to the publication.

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