CASE REPORT

Glomus vagale tumour, can it be diagnosed only on sonography?

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Abstract Vagal paragangliomas are rare benign tumors accounting for only 3% of head and neck paragangliomas and arise from vagal ganglions, predominately the nodose ganglion in the carotid sheath. Imaging plays an important role in their diagnosis, differentiation from more common lesions presenting in the carotid space, and management. MRI is the method of choice for investigation when paragangliomas are clinically suspected. The typical salt-and-pepper appearance in T1-weighted images is well known but not specific to paragangliomas. Demonstration of the continuity of the lesion with the vagus nerve on magnetic resonance imaging (MRI) further asserts the diagnosis of glomus vagale. Although this tumor can be diagnosed on a contrast-enhanced computed tomography (CECT) examination, it is limited by the non-demonstration of its neural origin. Sonography is considered only a preliminary investigation in patients, and a CECT and/or MRI study of the neck is conducted before treatment planning. This case is unique as a confident diagnosis of glomus vagale was made on high-resolution sonography of the neck. A middle-aged male patient presenting with a neck swelling was diagnosed with a glomus vagale tumor on USG by the direct demonstration of its continuity with the vagus nerve, and Doppler examination confirmed its vascularity. The diagnosis was confirmed on histopathology after surgical

excision. The sonographic appearance, Doppler characteristics, and imaging differential diagnosis of glomus vagale are discussed in detail.

Keywords Glomus tumor · Diagnosis · Differential · Paraganglioma · Extra-adrenal · Tomography · X-ray computed

Sommario I paragangliomi vagali sono rari tumori benigni e rappresentano solo il 3% dei paragangliomi del distretto testa-collo; originano dai gangli vagali e più frequentemente dal ganglio nodoso localizzato all'interno della guaina carotidea. L'imaging svolge un ruolo importante nella sua diagnosi, nella differenziazione dalle lesioni più comuni che possono trovarsi nello spazio carotideo e, nella sua gestione, una volta identificato. La RM è tuttavia l'indagine di scelta; tuttavia il tipico aspetto "sale e pepe" in T1 pesate pur essendo ben noto non è specifico per i paragangliomi. Pertanto la dimostrazione della continuità della lesione con il nervo vago con la RM è fondamentale per la diagnosi di glomo carotideo. La TC con mdc, può suggerire la diagnosi, ma è limitata nel dimostrarne la sua origine neurale. Pertanto l'ecografia è considerata solo come un'indagine preliminare in questi pazienti e TC con mdc e/o RM del collo vengono effettuate prima di pianificare il trattamento. In questo caso viene mostrato come la diagnosi di Tumore del glomo carotideo sia stata effettuata con esame ecografico. Un pz maschio di mezza età presentatosi con tumefazione del collo, grazie all'esame ecografico che dimostrava la presenza di una massa riccamente vascolarizzata al color-Doppler e in continuità con il nervo vago veniva inviato col sospetto di tumore glomivo, a esame di II livello e quindi a intervento chirurgico che ne confermavano, grazie all'esame istologico, la diagnosi. L'aspetto ecografico, le caratteristiche Doppler e di imaging del tumore del glomo vagale è di seguito discusso nei dettagli.

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Introduction

Glomus vagale tumor is a glomus tumor of the vagus nerve that arises from extra-adrenal nonchromaffin cells. It presents as a mass in the cervical region in close proximity to the carotid vessels or rarely presents as vocal cord palsy [1–3]. It must be differentiated from other lesions presenting as a neck mass in the same location as that of carotid body tumor, carotid aneurysm, or nerve sheath tumor. Imaging modalities such as CT or MRI are required to characterize and diagnose such neck lesions and map their relationship to the carotid vessels.

MRI is the method of choice for investigation and shows a typical salt-and-pepper appearance in T1-weighted images characteristic of paragangliomas. Demonstration of the continuity of the lesion with the vagus nerve on MRI is an advantage over a CECT study and confirms a lesion arising from the nerve, thus asserting the diagnosis of glomus vagale. Sonography is considered only a preliminary investigation in patients, and a CECT and/or MRI study of the neck is conducted before treatment planning.

Imaging findings are routinely described on CT/MRI, and gray-scale sonography is used only to rule out a vascular lesion before invasive biopsy is performed. This case report highlights how sonography can be used make a confident diagnosis of glomus vagale tumor sufficient to take a management decision without requiring any sophisticated, expensive, less accessible, and more time-consuming investigations like MRI. Moreover, radiation associated with CT/MDCT can be avoided, in addition to the risk involved in intravenous contrast. Here, we discuss the sonographic appearance of the glomus vagale and its differential diagnosis to help radiologists diagnose this condition based on ultrasound only.

Case report

A 51-year-old male patient complained of a painless mass on the left side of his neck. The swelling had grown to its current size in 7–8 months and has been stable since then. The patient showed no generalized symptoms. No pain associated with the swelling or decrease in the size of the mass was reported. No neurological complaint was presented. No similar swelling was present elsewhere in the body. The patient denied any significant past or family medical history. A firm, nontender, noncompressible swelling of approximately the size of a pear was present in the posterolateral aspect of the neck on the left side. Both the superior and inferior borders of the swelling were palpable with no thrill/bruit over the swelling. The skin over the swelling was freely mobile and appeared normal.

Inspection of the oral cavity revealed some bulge on the left oropharyngeal wall with a normal-looking intact mucosa over it. The size of the swelling did not change in terms of posture. Indirect laryngoscopy revealed medial bulging on the left pharyngeal wall. The hypopharynx and vocal cords appeared normal. Routine laboratory blood investigations showed unremarkable results, and the patient was sent to the radiology department for a preliminary sonographic examination, followed by a contrast CT of the neck.

High-resolution sonography of the neck with a 5–7.5 Hz probe revealed an isoechoic to hypoechoic, ovoid, well-defined, solid mass measuring 3.5×4.2 cm in close proximity to the carotid vessels and the internal jugular vein (Fig. 1a, b). The lesion was homogeneous in echotexture with no calcification or necrosis. The carotid vessels were displaced anteriorly, while the vein was displaced posteriorly and outward with evidence of compression but revealed normal flow on the Doppler examination. A meticulous sonographic examination of the caudal end of the lesion revealed its relationship with a linear isoechoic structure with a fascicular architecture corresponding to a nerve. This was seen lying posterolaterally to the common carotid artery corresponding to the vagus nerve (Fig. 2a). Tracing the nerve cranially showed an expansion of the nerve, which continued imperceptibly into the lesion palpable as neck swelling (Fig. 2b). A sonographic diagnosis of nerve tumor arising from the vagus nerve was made. Doppler showed marked internal vascularity in the lesion with an arterial waveform on spectral tracing (Fig. 3a, b). Sonography also showed anterior displacement of the carotid vessels and posterior displacement of the internal jugular vein. Therefore, because of the normally seen vessels, vascular lesion was ruled out. With the demonstration of a solid lesion of a nerve origin in this location with a characteristic displacement pattern of the carotid sheath vessels, the sonographic appearance of a nerve tumor with vascularity shown on Doppler helped in making a confident diagnosis of glomus vagale.

We proceeded with the contrast-enhanced CT examination of the neck to confirm the sonographic diagnosis and to look for the vascular supply of the tumor to plan further management. Computed tomography was performed on a 64-slice MDCT by injecting 70 ml of iohexol intravenously at a rate of 3.5 ml/s, and the contrast scan was acquired after a delay of 30 s. The source axial images were processed into sagittal and coronal multiplanar reconstructions. CECT revealed a strongly enhancing heterogeneous mass in the left carotid space, displacing the internal jugular vein posteriorly and the internal and external carotid artery anteriorly (Fig. 4a, b). The CECT findings were consistent with the diagnosis of glomus

Fig. 1 **a** Well-defined ovoid to round mass iso to hypoechoic mass, **b** lying in close proximity to internal jugular vein and carotid bulb

vagale, but its direct relationship with the vagus nerve was not demonstrated.

In due course, the patient was operated, and he developed hoarse voice and mild facial nerve palsy in the follow-up period. Post-operatively, the patient was counseled and reassured regarding the development of the neurological deficit. The patient recovered from facial palsy in 4 months, but he continues to have vocal cord palsy even on 15 months' follow-up. Histopathological examination confirmed the diagnosis of glomus vagale and showed a tumor adjacent to a nerve bundle composed of monomorphic cells with vesicular salt-and-pepper chromatin arranged in a typical Zellballen pattern (Fig. 5a–d). Typical findings of paraganglioma were revealed on

Fig. 2 **a** A round echogenic structure with fascicular pattern lying posterolaterally between the internal jugular vein and the common carotid artery and **b** on cranial tracing, nerve is seen to expand and continue into the fusiform mass seen to displace and compress the IJV in the cervical region

immunohistochemistry with tumor cells being chromogranin positive, and sustentacular cells were immunopositive for S100.

Discussion

Vagal paragangliomas, commonly known as glomus vagale, are unusual tumors arising in the perineurium in the paraganglionic tissue of the vagal nerve, most frequently in the ganglia region. They account for only 3% of

Fig. 3 **a** Color Doppler shows marked internal vascularity in lesion and **b** with arterial waveform

paragangliomas in the head and neck region [4] and are the third most prevalent tumor after carotid body and glomus jugulare tumors [1, 2].

In 1935, Stout identified the first paraganglioma of the vagus nerve [5], and Birell termed it a vagal body tumor [6]. As it arises from the vagus nerve, which is in the carotid sheath, it has a close relationship with the carotid vessels and the internal jugular vein (IJV). Therefore, it needs to be differentiated from other vascular lesions in this location before any biopsy or surgical intervention is planned. Routine imaging modalities such as CECT and MRI are preferred to characterize these lesions, define their anatomical extent, study their vascularity, and trace the

Fig. 4 **a** CECT axial images show the presence of a strongly enhancing mass causing anteromedial displacement of carotid vessels and posterolateral displacement of internal jugular vein. **b** coronal image showing the *left sided* strong but heterogeneously enhancing mass in the carotid space

vessels to map the arterial supply of the lesions. Sonographic assessment in the neck is limited to differentiating cystic from solid lesions, ruling out a vascular lesion on Doppler, or guiding FNAC/biopsies of superficial lesions. However, a smart and targeted examination can provide more than the routine information from a simple imaging investigation like sonography, which may prove to be adequate for taking management decisions.

This case study highlights the unexplored role of sonography in diagnosing a lesion arising from the vagus in the neck region. The vagus nerve is the 10th cranial nerve

◀**Fig. 5** Hematoxylin and Eosin—tumor adjacent to a nerve bundle (**a**, 4×) composed of monomorphic cells with vesicular salt and pepper chromatin (**c**, 40×) arranged in typical zellballen pattern (**b**, 20×). **d** Immunohistochemistry A: tumor cells were chromogranin positive (40×) B-Sustentacular cells were immunopositive for S-100 (20×). These features are typical of paraganglioma

and carries sensory, motor, and parasympathetic fibers. The cervical part of the nerve runs within the carotid sheath with the internal carotid artery anteromedially and the IJV anterolaterally [7–9]. Within the jugular foramen, the vagus bears a ganglionic enlargement (jugular or superior ganglion), and just below the skull base, a 2.5-cm-long larger inferior ganglion (nodose ganglion) is present [10]. Looking for the vagus nerve on imaging is not a routine practice, but any lesion suspected to be arising from it should prompt the radiologist to look for the nerve in the vicinity of the lesion.

Usually, the nerve is best seen on MRI examination along the plane of the nerve. On MR, the normal nerve appears as a round or oval structure with a fascicular pattern on the cross section. It is slightly hyperintense to the muscle in T2W images and isointense to the muscle with intervening areas of relatively high signal in T1W images. On MR examination, glomus vagale appears isointense in T1W images and heterogeneously hyperintense in T2W images with multiple flow voids, giving a salt-and-pepper appearance. Only direct demonstration of the lesion arising from the vagus nerve can make a definite diagnosis, which is possible on MR.

MDCT is not a modality that directly visualizes the normal nerves or lesions arising from them. It can only suggest that the lesion is of neural origin based on its imaging appearance, location, and relationship with the carotid sheath vessels. On the contrary, sonography can show even the normal nerves, which are accessible on high-resolution sonography. The radiologist only has to be aware of the normal appearance to be able to recognize the nerve (Fig. 2a, normal vagus nerve). The nerve appears as a round structure with a fascicular architecture with small hypoechoic areas (representing the nerve fascicles) separated by the hyperechoic septa of the perineurium [11]. The typical honeycomb appearance in axial sections is appreciated as a bundle of straws in longitudinal sections [11]. Sometimes a nerve is confused with a tendon close by on sonography. But close examination reveals that tendons are more hypoechoic than nerves and have a characteristic fibrillar pattern. This appears as multiple fine echogenic lines and multiple echogenic dots on the longitudinal and transverse sections, respectively [12]. As sonography is a real-time modality, tracing the nerve up to the neural lesion is possible, and their interrelationship can be examined.

Little experience is required to appreciate a nerve on sonography, and a focused examination can render success, as it was possible in our patient.

The most common tumors affecting the vagus nerve are nerve sheath tumors (schwannomas and neurofibromas). Schwannomas more commonly show evidence of calcification and necrosis and are located eccentrically along the long axis of the nerve, with nerve fascicles seen separately. They can be differentiated from the glomus tumor by their heterogeneous sonographic appearance, less vascularity, and eccentric relationship with the nerve.

Neurofibromas are a spindle-shaped enlargement of the nerve and cause a loss of the normal fascicular architecture [13]. They are generally hypovascular and an important distinguishing point from glomus vagale, which is a very vascular lesion. In the head and neck region, neurofibromas are much rarer than schwannomas [9, 14] but are more likely to undergo a malignant change. Clinically, a history of sudden increase in size or pain in the swelling warrants exclusion of a malignant transformation. A malignant nerve tumor has ill-defined and indistinct margins, has a more heterogeneous appearance, and may show infiltration of surrounding structures on imaging. Other important differentials of glomus vagale are carotid body tumor and lesions of the cervical sympathetic chain.

Carotid body tumor is the most common paraganglioma in the head and neck region and is a solid hypervascular lesion. This is the only lesion known to cause splaying of internal and external carotid arteries. Its characteristic relationship with the carotid artery and its branches is adequate for its diagnosis. The characteristic appearance of a carotid body tumor is a well-defined round to oval heterogeneously hypoechoic solid mass showing marked vascularity in the lateral neck and causing splaying of common carotid bifurcation. Sonography combined with color Doppler imaging showed a sensitivity of 92% and a specificity of 100% in detecting these lesions in a large series of 40 patients with 60 carotid paragangliomas and detected 86% of the lesions measuring as small as 5–10 mm in diameter [15, 16].

Paraganglioma and nerve sheath tumors of the cervical sympathetic chain present as a mass in the posterior cervical space and do not cause any separation between the artery and the vein. A large tumor [16–18] can still displace the vessels but does so anterolaterally for both the carotid arteries [19] and the IJV [20]. These lesions clinically present as ipsilateral Horner's syndrome and oropharyngeal fullness besides neck swelling.

A lymph nodal mass always remains a differential of a neck mass and presents as a lobulated, heterogeneously hypoechoic lesion with a known primary, thus suggesting the diagnosis [10]. However, in the absence of a primary,

distinguishing a solitary metastatic node from glomus vagale or schwannoma may be difficult. However, the hypervascular nature, involvement of/close relationship with the vagus, and pattern of separation of carotid sheath vessels give a clue toward the diagnosis of a lesion arising from the vagus nerve.

The branchial cleft cyst, especially the one lying between the external and the internal carotid arteries, can rarely be considered in the differential diagnosis of vagus nerve paragangliomas. However, sonographically they are round to oval hypo- to anechoic masses with well-defined margins and thin walls, showing features of compressibility and posterior wall enhancement, suggesting a cystic lesion [21]. In patients where confusion persists, FNAC can be used to establish a preoperative diagnosis, which reveals a yellow fluid with keratinized anuclear cells, squamous epithelium, and a matrix of amorphous debris [21]. Being less invasive and safe, core needle biopsy of the lesions is preferred to diagnostic surgery when FNAC fails to reach a definite diagnosis in suspected neck schwannomas or any other indeterminate neck mass [22]. Management of paraganglioma involves surgical excision with preoperative embolization as an important adjunct to surgery [23–25].

Our patient developed hoarse voice after the excision of the glomus vagale tumor. This complication has been reported before and occurs because of the involvement of fibers of the recurrent laryngeal nerve during surgery [26]. According to Thabet et al., postoperative vagal paralysis was observed in all patients with vagale paraganglioma because sacrificing the vagus nerve was essential for its excision [26]. Our patient also had transient facial nerve palsy, a complication known after surgery for carotid sheath lesions [27].

A confident diagnosis of glomus vagale is possible on sonography with an attempt to trace the nerve in the carotid sheath in relation to the lesion. Demonstration of the lesion in continuity with the nerve, sonographic appearance, pattern of displacement of the carotid sheath vessels, and detection of high vascularity on Doppler are adequate to establish the diagnosis of glomus vagale.

Compliance with ethical standards

Conflicts of interest There is no conflict of interest (financial or non-financial) declared.

Ethical approval This publication complies with all the ethical standards. This case report is not under consideration for publication anywhere else. The text for publication has been approved by all co-authors.

Informed consent An informed consent was obtained from the patient regarding the imaging work-up of the patient and the use of clinical data and images for publication purpose without revealing the identity of the patient.

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