CASE REPORT

Two synchronous glomus tumors simulating a single mass: glomus vagale and glomus caroticum

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ABSTRACT
Glomus tumors, especially with some definitive MR imaging features, are diagnosed easily. However, when two glomus tumors are suspected in one single mass, careful evaluation is necessary. We report one patient with two synchronous glomus tumors, ipsilateral glomus tumor of the carotid body and glomus vagale on the right neck, simulating a single mass.

Key words: • glomus tumor • magnetic resonance imaging

Glomus tumors are rare, benign neoplasms arising from paraganglionic tissue, derived from the neural crest origin distributed widely throughout the autonomic nervous system (1). The 4 main locations of glomus tissue within the head and neck are: (a) the carotid bifurcation (carotid body tumor), (b) the inferior ganglion region (ganglion nodosum) and cervical portion of the vagus nerve (glomus vagale or vagal paraganglioma), (c) the jugular bulb region (glomus jugulare), (d) the middle ear cavity (glomus tympanicum) (2).

We report a patient with 2 synchronous glomus tumors, an ipsilateral glomus tumor of the carotid body and a glomus vagale on the right neck simulating a single mass, with magnetic resonance (MR) imaging features.

Case report

A 56-year-old man presented with a mass on the right side of the neck, which had grown in size over the previous 12 months. He had no other symptoms like neck pain, dysphagia, hoarseness, nasal obstruction, and epistaxis. No family history of glomus tumors was reported. On examination, a painless, semi-mobile mass was palpated in the right jugulodigastric region. His blood pressure was within normal limits and the systemic physical examination findings were normal, except for the mass.

MR imaging examinations were performed with a 1.5 T unit MR imaging scanner (GE Healthcare, Signa Excite, Milwaukee, WI, USA). Fast spin-echo T1- and T2-weighted axial and coronal images were obtained. MR imaging parameters for T1-weighted images were; TR, 400–540 ms and TE, 13–22 ms, and for T2-weighted images; TR, 3700–5320 ms, TE, 74–80 ms. Section thickness was 6 mm with 1 mm interslice gap and an acquisition matrix of 256×192.

Pre-contrast T1-weighted MR images demonstrated a large solid mass on the right side, beginning at the level of the carotid artery bifurcation and extending superiorly, causing carotid artery branch separation and filling up the retrostyloid compartment of the parapharyngeal space. The mass was of approximately equal signal intensity to adjacent muscle, with slight heterogeneity.

On T2-weighted MR imaging (Fig. 1), 2 different hyperintense signals reflecting 2 different tumor matrixes within a single mass were noted. On post-contrast MR imaging (Fig. 2), 2 different contrast enhancement patterns were also noted. The smaller and less hyperintense anterior component of the mass was at the carotid bifurcation and was causing marked separation of the internal and external branches. Prominent anteromedial displacement of the internal and external carotid arteries, and posterolateral displacement of the internal jugular vein were present. The larger and more hyperintense posterior component of the mass began at the carotid bifurcation and extended up to the level of

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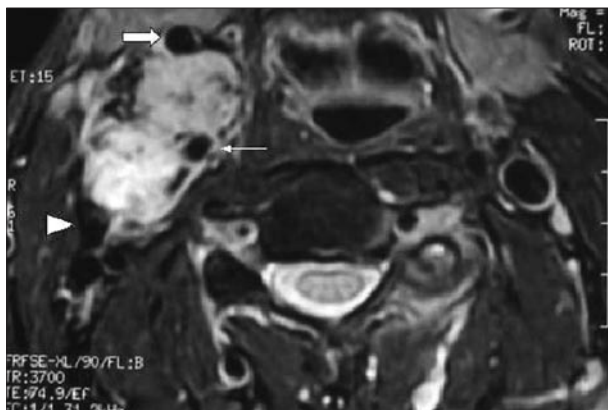
Figure 1. On axial T2-weighted MR image, 2 different hyperintense signals reflecting 2 different tumor matrixes within a single mass are noted. Signal void areas of vascularity are also seen (*thick arrow:* external carotid artery; *thin arrow:* internal carotid artery; *arrowhead:* internal jugular vein).
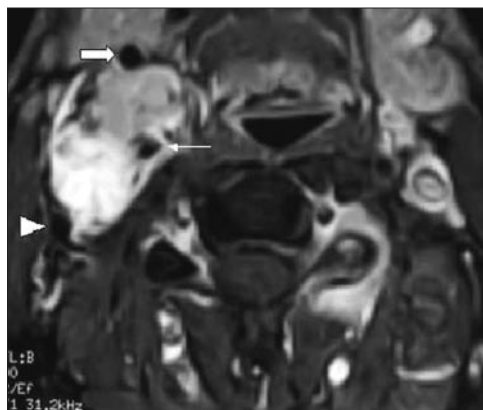

Figure 2. On axial post-contrast T1-weighted MR image, 2 different contrast enhancement patterns are also noted. The smaller and less hyperintense anterior component of the mass is at the carotid bifurcation, causing branch separation. The larger and more hyperintense posterior component of the mass is within the retrostyloid compartment of parapharyngeal space. Prominent anteromedial displacement of the carotid branches and posterolateral displacement of the internal jugular vein are present. Signal void areas of vascularity are also seen (*thick arrow:* external carotid artery; *thin arrow:* internal carotid artery; *arrowhead:* internal jugular vein).
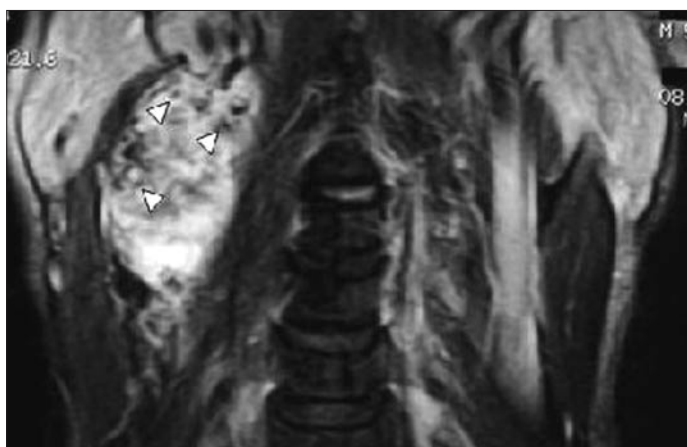

Figure 3. On coronal T2-weighted MR image, the heterogeneously hyperintense posterior component of the mass extends up to the level of the C1 vertebra. Multiple flow voids (*arrowheads*) indicating its hypervascular nature are scattered through the mass (salt-and-pepper appearance).
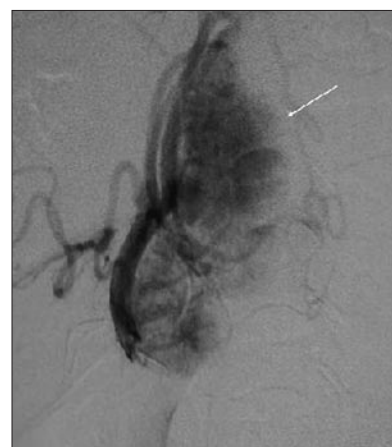

Figure 4. Lateral angiographic view obtained with a right external carotid artery injection reveals an intense blush of the hypervascular mass (*arrow*).

the C1 vertebra within the retrostyloid compartment of parapharyngeal space (Fig. 3).

Black areas of signal void that represented high vascularity were scattered throughout the mass on both T1- and T2-weighted images.

Digital subtraction angiography imaging revealed a tumor blush that was characteristic of the intense vascularity of the mass (Fig. 4). The feeding vessel of the mass at the carotid bifurcation was embolized. There was no feeding vessel of the mass located more posteriorly.

During the operation, 2 separate masses were encountered as were seen on MR imaging. The smaller one was easily dissected from the internal and external branches of the carotid artery. The larger and more posterior one was extending to the jugular foramen. The latter could not be dissected from the vagus nerve and resected with the uppermost portion of it. The result of the histopathological examination of the surgical specimens was paragangliomas (glomus caroticum and glomus vagale).

Discussion

Paragangliomas comprise 0.6% of all neoplasms in the head and neck. Most paragangliomas are carotid body or glomus jugulare tumors. Glomus vagale and tympanicum tumors are less prevalent. Patients are typically middle-aged and present with a painless, slow-growing neck mass or neurootological symptoms (3).

The role of MR imaging in the diagnosis and preoperative assessment of paragangliomas is well established. MR imaging characteristics of all paragangliomas are similar. A well-defined hypointense mass with approximately equal signal intensity to adjacent muscle, with areas of signal void is typically seen on T1-weighted MR imaging. T2-weighted images also demonstrate a well-defined mass, which is heterogeneously hyperintense, with greater signal intensity than muscle. Contrast-enhanced imaging shows intense tumor

enhancement, which is a key finding in the diagnosis. Punctate flow voids are seen and are believed to represent the hypervascular nature of this tumor. In larger lesions, a salt-and-pepper appearance on T1- and T2- weighted images is characteristic of paragangliomas (3, 4). In their study of 10 patients with a total of 15 paragangliomas, Olsen et al. a salt-and-pepper pattern in all lesions larger than 2 cm in maximal dimension (4). The "pepper" component represents the multiple areas of signal void interspersed with the "salt" component seen as hyperintense foci (due to tumor matrix, hemorrhage, or slow flow) on both short TR and long TR images (4).

The relationship of a paraganglioma to the adjacent carotid artery and/or jugular vein is well demonstrated by MR imaging. This demonstration may help differentiate carotid body tumors from vagal paragangliomas. Carotid body tumors tend to splay the internal carotid artery and external carotid artery, and thus widen the carotid bifurcation. Because of the anatomic site of origin of vagal paragangliomas in the post-styloid compartment of the parapharyngeal space, these lesions tend to displace the internal and external carotid artery anteriorly and medially, and the jugular vein posterolaterally, yet do not widen the carotid bifurcation (5, 6).

Multiple paragangliomas arise in at least 10% to 50% of cases (7). The co-existence of carotid body and glomus vagale tumors ipsilaterally seems to be quite a rare phenomenon. Recently, Magliulo et al. presented a patient affected ipsilaterally by a carotid body tumor and vagal paraganglioma, with findings similar to our case. They emphasized the importance of multiplanar imaging when there is suspicion of synchronous glomus tumors (8).

Paragangliomas are highly vascular in nature and are surrounded by vital neurovascular structures. During surgery, injuries to these structures are not uncommon. To avoid injury, a careful preoperative evaluation of the extent of the tumor, including its relationship to neurovascular structures and the possibility of synchronous glomus tumors, is important for planning the appropriate surgical procedure (9). Morbidity associated with vagal paraganglioma surgery is significantly higher than that of carotid body tumor surgery. In addition to the risk of injury to the carotid arteries and jugular vein seen with resection of the glomus caroticum, vocal cord paralysis due to the resection of the most superior portion of the vagus nerve is an almost unavoidable consequence of glomus vagale surgery. Surgery for vagal paragangliomas situated below the skull base might also place cranial nerves IX, XI, and XII at risk, besides the vagus nerve itself (10); therefore, the diagnosis of a tumor originating from the vagus nerve, or distinguishing a glomus vagale from a glomus caroticum is critical. Consequently, surgeons should inform patients about the probable hoarseness and aspiration problems caused by both superior and inferior laryngeal nerve paralysis.

In conclusion, when 2 glomus tumors are suspected in a single mass, careful evaluation is necessary. Post-contrast T1- and T2-weighted MR imaging can be considered to define the intensity differences within the mass with different cellular matrixes suggesting 2 tumors. The relationship of the mass to vascular structures may also offer a clue about the nature of the mass.

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