Three Cases of Vagal Nerve Schwannoma and Review of Literature

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Received: 3 February 2009 / Accepted: 18 October 2009 / Published online: 8 April 2011
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Abstract Schwannoma arising from the vagus is an uncommon (2–5%) benign nerve tumour. This tumour most often presents as a slow growing asymptomatic solitary neck mass which rarely undergoes malignant transformation. Definitive pre-operative diagnosis may be difficult and investigations such as FNAC have low specificity. The carotid artery and internal jugular vein may be displaced antero-laterally. Diagnosis is based on clinical suspicion and confirmation is obtained by means of surgical pathology. Surgical excision is the treatment of choice for this tumour, with recurrence being rare. We describe three cases of schwannoma involving the vagus who presented differently to our unit during past 5 years.

Keywords Schwannoma · Vagus · Neuromas · Parapharyngeal tumour

Introduction

Schwannoma arising from the vagus is an uncommon (2–5%) benign nerve tumour. This tumour most often presents as a slow growing asymptomatic solitary neck mass, which rarely undergoes malignant transformation. Investigations such as FNAC have low specificity. The carotid artery and internal jugular vein may be displaced antero-laterally. Diagnosis is based on clinical suspicion and confirmation obtained by means of surgical pathology. Schwannomas of the vagus nerve must be differentiated from the carotid body and glomus vagale tumors because

the distinction may influence treatment planning. Surgical excision is the treatment of choice for vagal schwannoma, with recurrence being rare. We describe three cases of schwannoma involving the vagus who presented to our unit during past 5 years.

Case 1

A 58 year old female patient presented with a gradually progressive swelling in right side of the neck since 1 yr, with no history of pain or weakness of upper limb. Examination showed 5 × 4 cm oval, smooth surfaced, non-tender firm swelling under the upper part of right sternocleidomastoid muscle, which was mobile in the horizontal but not in vertical direction. Carotid artery was felt in an anterior displaced position. Fine needle aspiration cytology showed benign spindle cell lesion with possibility of peripheral nerve sheath tumor or parotid tumor. Computerized tomography showed a well-defined mass with anterior displacement of the common and internal carotid artery (Fig. 1).

Using a skin crease incision a well encapsulated tumor arising from the vagus nerve was noted. Tumor was enucleated with sparing of the nerve. Post-operative period was uneventful with no evidence of IX cranial nerve paresis. Histopathology showed Antoni A and B bodies, palisading nucleus and verruca bodies suggestive of schwannoma (Fig. 2).

Case 2

A 22 year old female presented with an asymptomatic swelling in the left side of the neck since 2.5 years.

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Fig. 1 CT showing well-defined mass with anterior displacement of the common and internal carotid artery

Fig. 3 Carotid angiogram showed tumor blush and the carotids pushed anteriorly with splaying of bifurcation of carotid arteries

Fig. 2 Intraoperative photograph showing well encapsulated tumor arising from the vagus nerve

Fig. 4 Tumour arising from vagus nerve with both the carotids displaced anteriorly

Examination showed 6×4 cm swelling in upper 1/3 of the sternocleidomastoid muscle which was non-tender, firm, and mobile in the transverse direction. Carotid artery pulsation was displaced anteriorly.

Carotid angiogram showed tumor blush and the carotids pushed anteriorly with splaying of bifurcation of carotid arteries (Fig. 3).

Preoperatively an encapsulated tumor in the parapharyngeal fossa with the hypoglossal nerve traversing across it was noted. The carotids splayed over the capsule was also noted along with the vagus immediately deep and stuck to capsule. Histopathology showed schwannoma arising from vagus. Post operatively patient had features of vagal/glossopharyngeal paresis which recovered in 6 months.

Case 3

A 13 year old female presented with an asymptomatic swelling in the right posterior triangle 7×3 cm in size, vertically oval in shape with features similar as in case 1. Computerized tomography showed an encapsulated parapharyngeal tumour. On exploration it was seen arising from vagus nerve with both the carotids displaced anterior and deeply. Enucleation of the tumour was done. Histopathology was suggestive of schwannoma. Patient recovered uneventfully (Fig. 4).

Discussion

Tumors of the parapharyngeal space are rare, with neurogenic tumors being the most common. Neurilemmomas (also

known as schwannomas or neuromas) account for 55% of these tumors [1]. Approximately half of the reported parapharyngeal schwannomas arise from the vagus nerve [1]. Neoplasms of the vagus nerve include paragangliomas (50%), schwannomas (31%), neurofibromas (14%), and neurofibrosarcomas (6%) [2].

Most schwannomas of the vagus nerve are benign tumors. Most cases of schwannomas manifest between the third and sixth decades of the patient's life as a slow growing firm, painless mass in the lateral neck. Hoarseness, pain, or cough may be the presenting complaints. They displace the carotid arteries anteriorly and medially, jugular vein laterally and posteriorly. These swellings are mobile transversely but not vertically. These typical features were found in all our cases.

Schwannomas are usually confined to the retrostyloid parapharyngeal space, although patients with schwannomas that extend into the posterior cranial fossa through the jugular foramen have been reported [1]. Schwannomas are well-encapsulated tumors with distinctive cylindrical structures (Antoni type A tissue), which are set into indistinctive, loose stroma of fibers and cells (Antoni type B tissue). Typical features include necrosis, hemorrhage, and cystic degeneration.

On imaging, anterior displacement of the common or internal carotid artery is a characteristic finding of parapharyngeal neurogenic tumors [3]. On CT images, vagal schwannomas appear as well-defined masses, usually of higher attenuation than muscle on contrast-enhanced images. MR evaluation typically shows masses of intermediate signal on T1-weighted images and increased signal intensity on T2-weighted images with smooth, well-delineated margins and a homogeneous overall appearance [4]. Occasionally, as in our case 2, necrosis and cystic degeneration are seen. Furukawa et al. [5] found that vagal schwannomas separate the common or internal carotid artery from the jugular vein, whereas schwannomas of the cervical sympathetic chain do not.

Schwannomas of the vagus nerve must be differentiated from the carotid body and glomus vagale tumors because the distinction may influence treatment planning. Carotid body tumors arise at the carotid bifurcation, splaying the external and internal carotid arteries, whereas glomus vagale tumors usually displace the internal carotid artery anteriorly or medially or both. Both tumors enhance intensely on both CT and MR images and reveal a characteristic "salt-and-pepper" appearance on enhanced T1-weighted MR images because of flow voids frequently noted within the mass. This salt-and-pepper appearance is not a feature of schwannomas.

Surgical excision is the treatment of choice for both schwannomas and glomus tumors of the carotid space.

Because of the necessity to preserve important nerve functions, the surgical planning is often influenced by lesion size, location, vascularity, and relation to adjacent structures such as the vagus nerve, sympathetic chain, carotid artery, and jugular vein. Imaging can be very useful in differentiating vagal schwannomas from other lesions in this area, allowing the surgeon to plan the operative procedure to remove these tumors.

The literature six [6] hitherto appears to be in favour of the concept of subtotal or near-total tumour resection for non-vestibular head and neck schwannomas in situations where tumour is extensive and complete tumour resection cannot be achieved without compromising neural integrity and causing significant morbidity from paralysis and sensory loss. This approach is particularly suited to elderly patients or unfit patients who cannot tolerate prolonged surgery.

Histologically, it exhibits two main patterns, Antoni A and Antoni B. Antoni A tissue is represented by a tendency towards palisading of the nuclei about a central mass of cytoplasm (Verocay bodies). In contrast, Antoni B tissue is a loosely arranged stroma in which the fibers and cells form no distinctive pattern. A mixed picture of both types can exist. Other typical features include necrosis, hemorrhage, and cystic degeneration.

Malignant change in nerve sheath tumors in the head and neck is very rare. Malignant transformation is exceptional in solitary schwannomas and evidence suggests that subtotal resection may provide adequate disease control of non-vestibular head and neck schwannoma, although continued follow-up is mandatory.

References

1. Yumoto E, Nakamura K, Mori T, Yanaghiara N (1996) Parapharyngeal vagal neurilemmomas extending to the jugular foramen. *J Laryngol Otol* 110:485–489
2. Green JD, Olsen KD, DeSanto LW, Scheithauer BW (1988) Neoplasms of the vagus nerve. *Laryngoscope* 98:648–654
3. Som P, Sacher M, Stollman A, Biller H, Lawson W (1988) Common tumors of the parapharyngeal space: refined imaging diagnosis. *Radiology* 169:81–85
4. Som PM, Braun IF, Shapiro MD, Reede DL, Curtin HD, Zimmerman RA (1987) Tumors of the parapharyngeal space and upper neck: MR imaging characteristics. *Radiology* 164:823–829
5. Furukawa M, Furukawa MK, Katoh K, Tsukuda M (1996) Differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain by imaging diagnosis. *Laryngoscope* 106:1548–1552
6. Liu R, Fagan P (2001) Facial nerve schwannoma: surgical excision versus conservative management. *Ann Otol Rhinol Laryngol* 110:1025–1029