CASE REPORT

Cervical vagal schwannoma. A case report

Schwannoma del tratto cervicale del nervo vago

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SUMMARY

Schwannoma originating from the cervical vagus nerve is an extremely rare neoplasm. Vagal nerve schwannoma usually occurs between the third and fifth decades of life, it does not show sex predilection both sexes being equally affected and it most often presents as a painless, slow-growing, lateral neck mass. The treatment of choice is complete surgical excision with preservation of the neural pathway, when it is possible. These tumours, in fact, are almost always benign and a conservative surgical approach is emphasized by most of the Authors. A case of a cervical vagal schwannoma, in a 33-year old male with a previous medical history of malignant lymphoma, is described. The clinical features, diagnosis, management and pathological findings of cervical vagal schwannoma are discussed.

KEY WORDS: Vagus nerve • Benign tumours • Schwannoma • Magnetic Resonance imaging

RIASSUNTO

Lo schwannoma del nervo vago è una neoplasia molto rara. Questo tumore in genere insorge tra la terza e quinta decade di vita, non mostra predilezione di genere e spesso si presenta come una tumefazione laterocervicale indolore a lenta crescita. Il trattamento di scelta è la completa escissione con preservazione dell'integrità della via nervosa quando questo sia possibile. Tali tumori, infatti sono quasi sempre benigni e la maggior parte degli Autori raccomandano un approccio chirurgico conservativo. Riportiamo un caso di neurinoma del nervo vago cervicale in un uomo di 33 anni con pregressa storia clinica di linfoma. Vengono discussi le caratteristiche cliniche, la diagnosi, il management e gli aspetti istologici dello schwannoma del nervo vago cervicale.

PAROLE CHIAVE: *Nervo vago • Tumori benigni • Schwannoma • Risonanza Magnetica Nucleare*

Acta Otorhinolaryngol Ital 2009;29:33-35

Introduction

Cervical vagal schwannomas are rare, slow-growing tumours usually reported to occur in patients between 30 and 50 years of age. There does not seem to be a sex-related predisposition. They are usually asymptomatic benign lesions and complete surgical resection is the treatment of choice. Imaging plays a central role in diagnosing vagal nerve neoplasm and in particular Magnetic Resonance imaging (MRI) has become the routine imaging study for these tumours. MRI provides, in fact, important pre-operative information useful in planning optimal surgical treatment.

Case report

A 33-year-old male was referred to our Department for a palpable lump in the right side of the neck. Past medical history included a malignant lymphoma which occurred 20 years earlier and which was treated with chemotherapy and radiotherapy.

About one year before admission, he started to complain of mild hoarseness. Physical examination revealed a soft, smooth-surfaced mass in the right lower cervical region, measuring 3x3 cm. Upon palpating the mass, a paroxysmal cough was elicited.

Ultrasound (US) of the neck showed a hypoechoic nodule in the right side of the neck, 3 cm in diameter. A duplex doppler scan showed a flow signal in the peripheral position. MRI scan of the neck demonstrated a well-circumscribed mass, with high and dishomogeneous signal intensity, on the right side of the neck, between the internal jugular vein and the carotid artery (Fig. 1A, B).

The results of an ultrasonography (US)-fine needle aspiration biopsy (FNAB) of the mass, performed before the admission, were inconclusive. The patient, therefore, underwent surgery. Under general anaesthesia, a cervical incision along the anterior border of the sternocleidomastoid muscle was made and the dissection proceeded beneath the muscle. A yellowish-white, ovoid-shaped mass was observed, measuring ~3x3 cm lying between the carotid

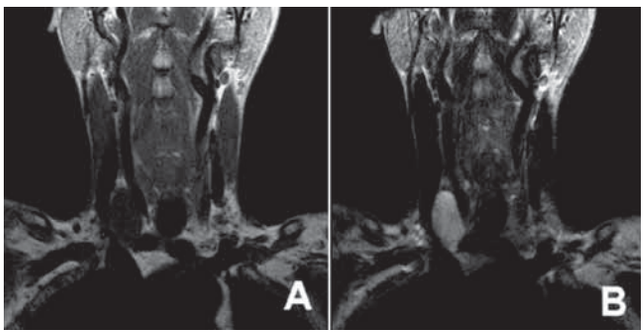

Fig. 1. MR imaging: Coronal T1-weighted (500/15) image (A) and coronal T2-weighted (5000/131) image (B) show an ovoid mass in right carotid sheath, between internal jugular vein and carotid artery. NB dishomogeneous high signal intensity in T2-weighted sequence.

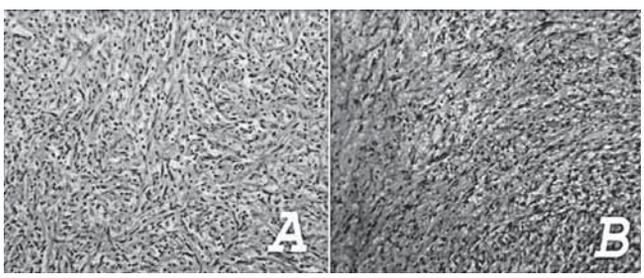

Fig. 2. Histological findings: A) Typical benign-appearing, spindle shaped cells (H&E, original. magn. x 400). B) S100 protein immunohistochemical positivity of the schwannoma.

artery and the internal jugular vein. Both the superior and inferior ends of the mass appeared in continuity with the vagus nerve. Since an adequate dissecting plane could not be reached, it was impossible to dissect the splayed nerve trunk off the tumour. The tumour was completely resected "en bloc" and end-to-end anastomosis of the vagus nerve was performed, after mobilization from the internal jugular vein, using microsurgical techniques.

The pathological examination confirmed the diagnosis of benign schwannoma of the vagus nerve. Microscopically, the neoplasia was composed of spindle cells organized in small fascicles, mainly in an oedematous background. Verocay bodies with spindle cells, organized in a palisading fashion, were rarely observed (Fig. 2A, B).

Post-operatively the hoarseness became more severe and examination of the larynx showed paralysis of the right vocal cord.

At follow-up, one year after surgery, the patient was well, without evidence of disease. Vocal cord palsy was still present.

Discussion

Schwannomas are rare peripheral nerve tumours; about one third occur in the head and neck region [1]. Clinically, they present as asymptomatic slow-growing lateral neck masses that can be palpated along the medial border of the sternocleidomastoid muscle.

Pre-operative diagnosis of schwannoma is difficult because many vagal schwannomas do not present with neurological deficits and several differential diagnoses for tumour of the neck may be considered, including paraganglioma, branchial cleft cyst, malignant lymphoma, metastatic cervical lymphadenopathy [2]. Furthermore, due to their rarity, these tumours are often not even taken into consideration in the differential diagnosis.

When symptoms are present, hoarseness is the most common. Occasionally, a paroxysmal cough may be produced on palpating the mass. This is a clinical sign, unique to vagal schwannoma. Presence of this sign, associated with a mass located along the medial border of the sternocleidomastoid muscle, should make clinicians suspicious of vagal nerve sheath tumours [1 3-5].

The usefulness of FNAB is still controversial; the majority of Authors do not recommend open or needle biopsy for these masses [2]; in our case, a FNAB was performed before admission, by the physicians that first evaluated the patient, but it was inconclusive.

There is general agreement concerning the great value of MRI in the pre-operative work-up as it is helpful in defining diagnosis and in evaluating the extent and the relationship of the tumour with the jugular vein and the carotid artery. The MRI appearance is considered quite typical and may lead to suspicion of the diagnosis pre-operatively as the cervical vagal neurinoma frequently appears as a well-circumscribed mass lying between the internal jugular vein and the carotid artery. As reported by Furukawa et al., MRI findings are also useful in providing a pre-operative estimation of the nerve of origin of the schwannomas and to differentiate pre-operatively between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain. The vagal schwannomas, in fact, displace the internal jugular vein laterally and the carotid artery medially, whereas schwannomas from the cervical sympathetic chain displace both the carotid artery and jugular vein without separating them [6-10]. In our case, the criteria of Furukawa et al. [6] were present (Fig. 1 A-B).

Treatment of vagal nerve tumours is complete surgical excision. At surgery, these tumours appear as yellowish-white, well-circumscribed masses. Dissection of the tumour from the vagus with preservation of the neural pathway should be the primary aim of surgical treatment for these tumours. Incomplete treatment, such as open biopsy, should be avoided, since it makes definitive excision of the tumour much more difficult.

If it is impossible to find an adequate plane and is technically difficult to preserve the integrity of the nerve trunk, the involved segment may be resected and an end-to-end anastomosis performed using microsurgical techniques [3]. This type of procedure often results in definitive vocal cord paralysis.

The reported incidence of pre-operative vocal cord paralysis is about 12%, but hoarseness is almost always present

following surgery. Therefore, pre-operative assessment of vocal cord mobility should be strongly recommended. Although it is very rare, clinicians should bear in mind the possibility of a nerve sheath tumour in the presence of a neck mass. Pre-operative suspicion is very important, because the patient, and the patient's family, should be informed about the possible post-operative neurological complications [7]; as far as concerns post-operative vocal cord palsy, an incidence of 85%, has, in fact, been reported [3 11]. Furthermore, since vagal schwannomas are almost invariably benign in nature, a conservative approach should always be considered in first instance [2-4]. In the presence of post-operative vocal cord palsy, aggressive voice therapy, for vocal cord compensation, should be started soon after surgery.

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Received: February 18, 2008 - Accepted: September 27, 2008

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