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Cervical vagal schwannoma

Single case report

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Cervical vagal schwannoma. Single case report

Schwannomas of the cervical vagal nerve are rare neoplasms, usually occurring between the third and the sixth decade of life. They don't demonstrate any sex predilection, and they often present as slow-growing, palpable neck masses, left-right sided, without early neurological symptoms, and they are often confused with enlarged lymphnodes or lipomas. Several differential diagnosis should be considered. Imaging techniques are largely used to define their etiology. If they are considered primary to evaluate their relationship with surrounding structures (i.e. omolateral neck vessels and nerves, oesophagus, trachea), they don't always result decisive for a correct differential diagnosis. Surgical excision is the treatment of choice: when technically possible, nerve sparing technique has to be preferred to en-bloc resection, because of the possibility of neurological outcomes such as dysphonia, dysphagia, hoarseness, vocal cord paralysis after surgical therapy. We report our case about a 34 years-old male, evaluating differential diagnosis course, choosing the correct therapy in relation with literature cases, and including new techniques for post-operative outcomes, such as injectable soft-tissue bulking agent performed in the last years to ameliorate dysphonia after nerve trunk injury.

KEY WORDS: Enucleation; Neurinoma; Schwannoma

Introduction

Cervical vagal schwannoma is a rare neoplasm, occurring between 20 and 50 years[1], with no sex predilection. Pre-operative diagnosis is difficult because it most often presents as palpable neck mass, without neurological symptoms and several differential diagnosis for tumor of the neck may be considered. When technically possible, the treatment of choice is the complete surgical excision with vagal nerve sparing[1-5]. We report our case evaluating diagnosis and therapy in relation with literature.

Case report

In July 2011, a 34-years-old male was referred to our Department for a palpable lump in the right side of the neck, appeared two months before, in association with hacking cough. Physical examination revealed a movable painless mass, measuring 3 x 3 cm.
The patient underwent ultrasonographic (US) examination with Color Doppler of the mass, showing a round nodule near the right inferior thyroid lobe, measuring

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3,5 x 2,3 x 2,6 cm, well-defined, with small colliquative areas, between the internal carotid artery and the internal jugular vein (Fig. 1).
A preoperative fine needle aspiration biopsy (FNAB) showed neurogenic mesenchymal tumor characteristics[2]; the neck CT-scan showed a compact mass with late and dishomogenous contrast pattern, adjacent to the thyroid right lobe side, to the internal jugular vein and internal carotid artery (Fig. 2).
One month later the patient underwent surgery under general anesthesia. A cervical incision along the anterior border of the sternocleidomastoid muscle was made, and a capsulated mass was observed, measuring 5 x 2 cm, lying between the carotid artery and the jugular vein (Fig. 3). It was impossible to dissect the tumor off the vagal nerve trunk, so it was completely resected "en bloc" from the proximal and distal stumps.
The pathological examination showed a classical beningn Schwannoma of the vagus nerve, with "Antoni A" and "Antoni B" areas. Post operatively the patient showed right signs of mild disphony; in 3rd post-operatively day he underwent video-laryngoscopy that showed right vocal cord paralysis, and he was discharged with cortisone therapy.

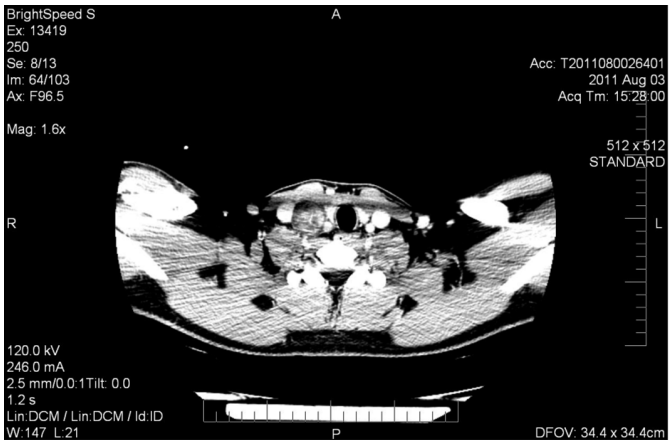

Fig. 1

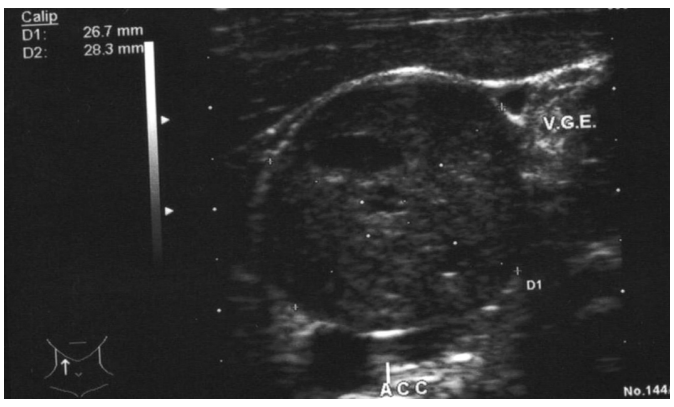

Fig. 2

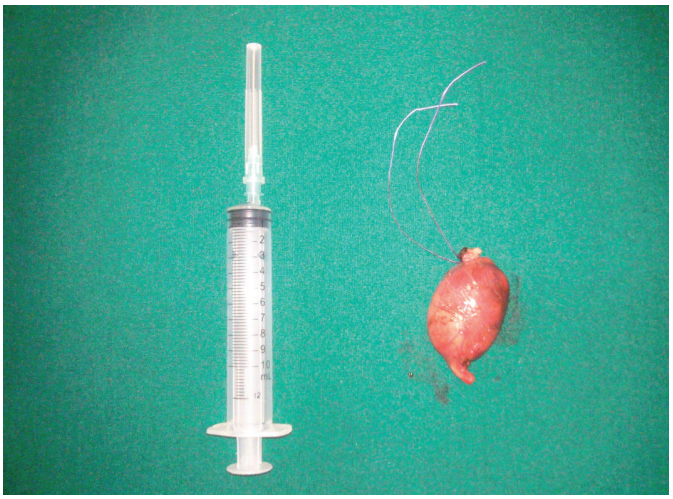

Fig. 3

In September 2011 he underwent new video-laryngoscopy, showing vocal cord paralysis in intermediate position with early left cord balance. He started logopedic therapy and at two months follow-up, a new video-laryngoscopy showed clear left cord balance with right vocal cord paralysis in paramedian position.
The patient continued logopedic therapy until March 2012, with small results. He still suffered for disphagy, tachycardia, hoarseness and mild dysphonia. In April 2012 he underwent surgery for implantation of an injectable soft-tissue bulking agent used to treat vocal cord paralysis (VOX® - Uroplasty) and he partially recovered vocal cord functions[6].
At 1 year follow up, he still presents vagal nerve resection symptoms, but he recovered his voice, except for high tones.

Discussion

Schwannomas are rare peripheral nerve neoplasms, with difficult diagnosis. Clinically, a neck region schwannoma appears like a palpable mass along the medial border of the sternocleidomastoid muscle. Several differential diagnosis may be considered, like neurogenic tumors (paraganglioma, neurosarcoma, neurofibroma), enlarged lymph nodes, branchial cleft cysts, lipomas and aneurysms [1,2].
Preoperative US seems to be an useful imaging examination, evaluating vagal nerve morphology, the contiguity with the mass, the absence of internal vascularity [7].
As reported by Le Corrolier et al. [8], sonograms of Schwannoma typically show a well-defined, round, hypoechoic, homogeneous solid mass, with moderate to marked posterior acoustic enhancement. In our case, the mass was clearly identified between internal carotid artery and internal jugular vein, with typical sonographic pattern. However, unlike classical schwannoma, during US we found colliquative areas, which don't represent clas-

TABLE I

Author	Technique	Deficit	Symptoms
De Araujo et al[1]	Enucleation	30%	Transient Paralysis
De Araujo et al[1]	En-bloc resection	100%	Cord Paralysis
Gilmer-Hill et al[4]	Enucleation	25%	Mild dysphonia, hoarseness
Valentino et al[5]	Enucleation	29%	Transient cord paralysis
Valentino et al[5]	En-bloc resection	64%	Cord Paralysis, hoarseness

sical scwannomas patterns. For this reasons, we performed a FNAB of the mass as reported by Chiofalo et al.[2]
CT scan shows topographic details about the lump and adjacent organs, with typical anteromedial displacement of the internal carotid artery [9].
The treatment of choice is enucleation with dissection of the tumor off the vagus nerve and preservation of the nerve trunk [1-5]. Otherwise, some Authors refer to shell out the tumor off the nerve [1,5]. When those techniques are not achievable, it's possible to realize an "en bloc" resection of the mass, without nerve sparing [1].
As reported by de Araujo et al. [1], function preservation rates change depending of the surgical technique (Table I). Two cases of nerve reconstruction with an end-to-end anastomosis are described; the nerve functional results are controversial, with vocal paralysis (Chiofalo et al. [2]) or nerve function sparing (Gilmer-Hill et al. [4]).
According to the literature, we didn't perform anastomosis because of the technical difficulties and uncertain results of the procedure. Post-operatively we found vocal cord paralysis and other classical symptoms such as disphagy, tachycardia, hoarseness and mild dysphonia.
An interesting treatment for vocal cord paralysis is the implantation of a an injetable bulking agent (VOX® - Uroplasty), made up of two parts: a water soluble gel (polyvinylpyrrolidone) and a silicone elastomer implant material (cross-linked polydimethylsiloxane). As reported by Turner et al. [6], 32 of 39 patients treated with this procedure were satisfied with the post-operative vocal results. According to the literature, also in our case we found a quick recovery of the voice quality.
An appropriate diagnostic course remains essential in the pre-operative diagnosis for the treatment of palpable neck masses, that could deceive also experienced general surgeons.

Riassunto

INTRODUZIONE: Gli Schwannomi del nervo vago sono tumori rari che si manifestano tra i 20 e i 50 anni, senza predilezione di sesso. La loro diagnosi preoperatoria risulta difficile in quanto essi si presentano clinicamente come masse palpabili nella regione del collo, in assenza di segni neurologici specifici ed entrano in diagnosi differenziale con numerose altre patologie. Il trattamento di scelta per questi tumori è l'escissione chirurgica cercando di preservare, qualora sia tecnicamente ed anatomicamente possibile, l'integrità del nervo vago.
CASE REPORT: Paziente di 34 anni, maschio, si recava presso la nostra struttura in seguito al riscontro da circa due mesi di una massa palpabile in regione laterocervicale destra e alla contemporanea comparsa di tosse stizzosa. L'esame obiettivo evidenziava una massa di circa 3 cm di diametro, mobile e non dolente.
Un'ecografia mostrava un nodulo circoscritto in prossimità del lobo tiroideo inferiore,tra l'arteria carotide interna e la vena giugulare interna. Un FNAC preoperatorio orientava per un tumore mesenchimale neurogenico e l'esame tac mostrava una tumefazione solida con pattern contrastografico disomogeneo e tardivo, che entrava in contatto con il margine esterno del lobo destro della tiroide, con la vena giugulare e con l'arteria carotide interna. Il paziente veniva sottoposto ad intervento chirurgico in anestesia generale con incisione presternoicleidomastoidea destra che evidenziava una massa capsulata ci circa 5 x 2 cm, grigio-gialla, laterale rispetto alla carotide e mediale rispetto alla giugulare. La massa, che risultava essere di pertinenza del nervo vago, veniva completamente asportata "en bloc" in quanto i peduncoli prossimale e distale sembravano contenere una struttura di tipo nervoso che non era possibile preservare. L'esame istologico evidenziava uno Schwannoma convenzionale benigno con aree di "Antoni A" e "Antoni B". Nel post operatorio il paziente evidenziò una lieve disfonia con evidenza alla laringoscopia della paralisi della corda vocale destra. Il paziente veniva dimesso con terapia cortisonica in terza giornata postoperatoria. Eseguiva dopo un mese una laringoscopia di controllo che confermava la paralisi della corda vocale destra,con parziale compenso della corda vocale sinistra. Dopo sette mesi di terapia logopedica, il paziente veniva sottoposto ad intervento chirurgico di impianto di un agente tissutale iniettabile (VOX® - Uroplasty), con recupero parziale della funzione della corda vocale destra. Ad un anno dall'intervento ha recuperato quasi totalmente la funzione vocale.
DISCUSSIONE: Gli Schwannomi sono rari tumori dei nervi periferici di difficile diagnosi che clinicamente si presentano nella loro localizzazione cervicale come masse palpabili lungo il margine mediale del muscolo sterno-

cleidomastoideo ed entrano in diagnosi differenziale con altri tumori neurogenici (neurofibromi, paragangliomi, neurosarcomi), linfonodi, cisti brachiali, lipomi ed aneurismi. L'esame preoperatorio di scelta risulta essere l'ecografia con color doppler, che permette di valutare la morfologia del nervo vago e la diretta continuità della massa con lo stesso e l'assenza di vascolarizzazione al suo interno. La TC offre dettagli topografici sul tumore e le strutture adiacenti. L'utilità del FNAB preoperatorio è controversa in quanto per alcuni autori esso è sempre indicato, per altri è controindicato e per altri ancora risulta indicato soprattutto nei casi dubbi di difficile diagnosi differenziale. Il trattamento dello Schwannoma del nervo vago è la completa escissione chirurgica con dissezione della massa dal nervo e conservazione della sua integrità ma spesso, come nel nostro caso, il tumore risulta indissociabile dal nervo e non è possibile preservarne l'integrità e ciò purtroppo comporta la paralisi della corda vocale. Un trattamento molto interessante ed innovativo per la paralisi della corda vocale è l'impianto di un agente tissutale fatto di un gel idrosolubile e di un elastomero di silicone (VOX® - Uroplasty), grazie al quale sono stati ottenuti risultati molto confortanti nel recupero della funzione della corda vocale. Un corretto iter diagnostico preoperatorio risulta fondamentale per la corretta diagnosi e trattamento delle lesioni palpabili del collo che possono trarre in inganno anche il chirurgo generale più esperto.

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