A Large Extra Cranial Cystic Trigeminal Schwannoma of the Parapharyngeal Space – Exploring the Right Approach

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Abstract Tumors of the parapharyngeal space (PPS) are uncommon, comprising less than 1 % of all head and neck neoplasms. Neurogenic lesions are the most common tumors of the poststyloid PPS accounting for nearly 25–30 % of all the PPS lesions. Schwannomas are uncommon nerve sheath tumors that may originate from any peripheral, cranial or autonomic nerve of the body, about one-third of them are known to arise in the head and neck region. Trigeminal schwannomas are known to arise from the root, ganglion, or intracranial portion of the three peripheral divisions of the nerve – ophthalmic (CN V1), maxillary (CN V2) and mandibular (CN V3). Schwannomas involving the mandibular division of the trigeminal nerve and localizing exclusively in the parapharyngeal space (PPS) is extremely rare and a surgical approach to such tumors has not been well established. The management of a PPS schwannoma is thus a clinical challenge, an adequate preoperative imaging with the identification of the possible nerve of origin allows for making informed decisions regarding the various approaches of management.

Keywords Schwannoma · Trigeminal nerve · Parapharyngeal space

Introduction

Schwannomas are uncommon nerve sheath tumors that may originate from any peripheral, cranial or autonomic nerve of the body with the exception of the olfactory and optic nerve, about one-third of them are known to arise in the head and neck region. Trigeminal schwannomas are known to arise from the

root, ganglion, or intracranial portion of the three peripheral divisions of the nerve – ophthalmic (CN V1), maxillary (CN V2) and mandibular (CN V3). Schwannomas involving the mandibular division of the trigeminal nerve and localizing exclusively in the parapharyngeal space (PPS) is extremely rare and a surgical approach to such tumors has not been well established. We recently got to manage a rare case of an extra cranial cystic schwannoma in the PPS arising from the CN V3 and discuss the various surgical approaches to its management.

Case Report

A 41-year-old gentleman was referred to us for evaluation of a slowly growing left parotid gland tumor for a year. Clinical evaluation revealed a well circumscribed mass in the left parotid region, which was also appreciated intra-orally as evidenced by a bulge in the left tonsillar fossa. His mouth opening was not restricted, although he reported a vague left sided facial pain. Evaluation of the facial and all other cranial nerves was unremarkable except for mild hyperesthesia along the distribution of CN V3. MRI scan revealed a 6.4×6×4.9 cm well defined, multi-lobulated mass in the left PPS with heterogeneous contrast enhancement and a cystic component in the medial aspect. The mass was seen extending into the parotid space, compressing and pushing the lobes on the parotid gland laterally. Medially the mass was seen pushing the pharyngeal mucosal space and was seen pushing the great vessels in the neck posteriorly. Superiorly the mass was seen widening the foramen ovale and indenting the inferior aspect of the temporal lobe. The radiological impression was suggestive of a cystic schwannoma arising from CN V3 (Fig. 1a and b). The patient was taken up for surgery using a combined trans-cervical and a trans-parotid approach. The entire mass was gently delivered in the neck and was enucleated, preserving the integrity of the V and VII cranial nerves. Figure 2a-d the patient had an uneventful post

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Fig. 1 **a** Axial **b** Coronal: MRI scan showing a 6.4×6×4.9 cm well defined, multi-lobulated mass in the left PPS with heterogeneous contrast enhancement and a cystic component in the medial aspect

operative period; the final histopathology confirmed the diagnosis of a cystic schwannoma (Fig. 3a and b).

Discussion

Tumors of the parapharyngeal space (PPS) are uncommon, comprising less than 1 % of all head and neck neoplasms. Of the PPS tumors, 70–80 % are supposedly benign and 20–30 % are malignant [1]. Neurogenic lesions are the most common tumors of the poststyloid PPS and account for nearly 25–30 % of all the PPS lesions. Schwannomas are the most common neurogenic tumors and are known to arise from any nerve surrounded by schwann cells. They are slow growing, encapsulated tumors, histologically distinct from the nerve of origin and hence rarely cause palsy of the nerve. In the PPS, the most common nerves of origin of schwannomas are the vagus nerve

and the sympathetic chain. The mandibular nerve runs laterally along the skull base then exits the cranium by descending through the foramen ovale into the masticator space and is rarely involved by a schwannoma.

The diagnosis of schwannoma is suggested by clinical features (a history of slow and indolent growth of a firm mass which is more mobile along the plane perpendicular to the course of the nerve and rarely a palsy of the nerve of origin) and is supported by imaging. MRI scan with gadolinium-based contrast medium is the preferred technique of choice for imaging. It allows for an accurate radiological diagnosis of a schwannoma with the identification of the nerve of origin in a majority of the cases, which is crucial to the management [2, 3]. Trigeminal schwannomas are more likely to contain cystic components and are more varied in appearance than acoustic schwannomas, as was also seen in our patient. Most of the time, the diagnosis can only be confirmed on the histological

Fig. 2 **a** Clinical photograph at presentation. **b** Intra operative photograph demonstrating the combined transcervical transparotid approach to delineate the PPS schwannoma. **c** Intra operative photograph following superficial parotidectomy and removal of the PPS schwannoma. **d** Specimen photograph showing the left parotid gland and the resected PPS schwannoma

Fig. 3 **a** H&EX10: Shows focal cellular spindle cells with dark staining nuclei, areas with marked myxoid degeneration and hyalinization. Scattered calcifications are seen. Few blood vessels are also seen

surrounded by hyalinized stroma. **b** H&EX40: Shows tumor tissue with hypo (Antoni B) & hyper cellular areas (Antoni A) many vascular channels and cystic spaces

examination of the resected surgical specimen as a fine needle aspiration cytology studies are of limited value.

Surgery is the mainstay of treatment for schwannomas of the parapharyngeal space (PPS) [3–10]. The choice of operation is mainly determined by the relationship between the tumor and the nerve of origin. Complete intra-capsular excision preserving the nerve of origin should be attempted when feasible, [3] but for extensive schwannomas, nerve sacrifice with reconstruction and rehabilitation are important considerations.

The choice of surgical approach is dictated by the size of the tumor, its location, its relationship to the great vessels and the suspicion of malignancy. A number of middle cranial fossa skull base approaches have been described for resection of trigeminal schwannomas involving the root, ganglion and the intra cranial portion of the trigeminal nerve [4, 5]. Approaches to the PPS schwannomas include, transcervical, transzygomatic, transmaxillary, the combined transcervical transparotid approach, the combined transcervical transmandibular approach and a combination of the above approaches.

The combined transcervical-transmandibular approach is preferred in malignancies in which better exposure facilitates oncologic resection and in cases in which distal control of the carotid artery at the skull base is required [6–8]. It is also the preferred approach for large tumors with superior PPS and extension. This approach is associated with many unique complications, primarily because of incorporation of a mandibulotomy. The complications include infection, temporomandibular joint dysfunction, nonunion, malocclusion, plate extrusion, and tooth loss. Moreover a tracheostomy is usually performed in conjunction with a transmandibular approach because of the significant upper airway edema caused as a result of the surgical manipulation of the oropharynx.

A combined transcervical-transparotid approach, on the other hand is devoid of the mandibulotomy associated complications. In this approach, the transcervical approach is combined with a transparotid approach by extending the incision superiorly as for a parotidectomy. The facial nerve is identified and dissected, a parotidectomy is performed and the PPS space is delineated. Although the overall exposure is limited, well encapsulated, radiologically benign appearing

large tumors such as a schwannoma can be safely and carefully removed with acceptable cosmetic or functional outcomes [9, 10], as was done for our patient.

In conclusion, the management of a PPS schwannoma is a clinical challenge, an adequate preoperative imaging with the identification of the possible nerve of origin allows for making informed decisions regarding the various approaches of management.

Conflict of Interest None.

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