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RESEARCH PAPER

Extracranial Trigeminal Schwannomas: A Retrospective Analysis

Aviral Agrawal[1] · Virendra Singh[2] · Amrish Bhagol[2] · Pradeep Kumar[2] · Anjali Narwal[3]

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Abstract
Objective To analyse the clinic-radiological features and treatment outcome of extracranial trigeminal schwannomas.
Method Medical records and radiographs of patients treated, from January 2011 to December 2013, for neurogenic tumors were retrospectively reviewed. Extracranial schwannomas other than those of trigeminal nerve were also excluded. A number of parameters, including the patient's age, gender, site, clinical features, radiographic features, histologic variants and treatment provided as well as any associated complications were recorded and analysed.
Result A total of 5 patients met the inclusion criteria. The patients were males, aged 16–56 years. All the schwannoms appear to originate from the terminal branches of trigeminal nerve.
Conclusion Long standing asymptomatic swelling of cheek should include trigeminal schwannomas as the differential diagnosis. The diagnosis though confirmed by the histologic examination but can also be made on the basis of MRI finding. Neural function can be preserved by meticulous surgery.

Keywords Extracranial Schwannoma · Trigeminal nerve · Access osteotomy

✉ Aviral Agrawal
aviral2011@yahoo.co.in

Virendra Singh
drvirendrasingh1@yahoo.co.in

Amrish Bhagol
bhagol.amrish@gmail.com

Pradeep Kumar
pradeepjaglan143@gmail.com

Anjali Narwal
anjalinarwal@yahoo.com

[1] Department of Oral and Maxillofacial Surgery, Kalpana Chawala Government Medical College, Karnal, Haryana, India

[2] Department of Oral and Maxillofacial Surgery, Post Graduate Institute of Dental Sciences (PGIDS), Rohtak, Haryana, India

[3] Department of Oral Pathology, PGIDS, Rohtak, India

Introduction

Schwannoma, also known as neurilemmoma, is a benign tumor of the nerve sheath arising from the perineural schwann cells. Approximately 25–45 % of all schwannoma variants occur in the head and neck, usually presenting in the fourth decade of age [1]. Schwannomas can occur along the pathway of any somatic or sympathetic nerve. Typically, these lesions are freely mobile, with a single attachment at the nerve of origin. The lesions are solitary, slow growing, and not commonly associated with pain or neurologic symptoms, except in the setting of the third form of neurofibromatosis termed "schwannomatosis" [2].

If pain or neurologic symptoms are present, they are usually associated with a mass effect.

The nerves most commonly involved in schwannomas of the head and neck are the vagus and the cervical sympathetic chain [3]. Though the trigeminal nerve constitutes the second most frequent site for intracranial schwannoma occurrence, after the vestibular nerve [4, 5], literature on extracranial trigeminal schwannoma is very scanty and thus forming the purpose of the present study which is to analyse the clinic-radiological features and treatment outcome.

Materials and Methods

The medical records and radiographs of patients treated for neurogenic tumors at our centre, over a 3 year period (from January 2011 to December 2013) were retrospectively reviewed. Patients who were diagnosed other than schwannoma on the basis of histopathological examination and intra-cranial schwannomas were excluded from the study. Extracranial schwannomas other than those of trigeminal nerve were also excluded. A number of parameters, including the patient's age, gender, site, clinical features, radiographic features, histologic variants and treatment provided as well as any associated complications were recorded and analysed.

Results (Table 1; Figs. 1, 2, 3, 4, 5, 6)

A total of 5 patients met the inclusion criteria. All the patients were males, aged 16–56 years treated for extracranial schwannoma over a period of 3 years. All the schwannomas appear to originate from the terminal branches of trigeminal nerve except one which originated from main trunk as it was seen exiting from the foramen ovale. The most common site of the tumor was buccal space i.e. 4 cases while in one the tumor mass was present in parapharyngeal space. The clinical features varied from one patient to another but their chief complaint remained same i.e. asymptomatic swelling on cheek and facial asymmetry. Their radiographic investigation done was MRI which showed signal patterns i.e., isointense T1 signal relative to skeletal muscle; increased and slightly heterogeneous T2 signal.

Enucleation of the tumor mass was provided as the treatment modality to all patients. To expose the tumor mass access osteotomy was required in 2 patients. All the patients were kept on follow up to check recurrence. All the excised specimens were sent for histopathological examination which confirmed it to be schwannoma. Histologically, all trigeminal schwannomas were of classical type except one which was of ancient type.

On the basis of clinical location of swelling and radiographic location of tumor mass, the diagnosis of schwannoma of terminal branches of trigeminal nerve was made. The follow up period of all the patients were uneventful.

Discussion

Schwannomas are slow-growing, benign neoplasms derived from the sheath cells that encompass myelinated nerve fibers. They may be encapsulated and are most frequently located in the head and neck [6].

Table 1 Retrospective data of extracranial trigeminal schwannoma

S No.	Age (years)	Sex (M/F)	Site	Clinical picture	Radiographic finding	Treatment done	Histological type	Follow up (months)	Recurrence
1	55	Male	Right cheek	Painful swelling of 3 months duration with reduced mouth opening	MRI with contrast showed well defined enhancing soft tissue mass	Enucleation	Classical	15	Nil
2	16	Male	Left para-pharyngeal	Facial asymmetry with displacement of soft palate	MRI with contrast showed well defined enhancing soft tissue mass	Enucleation through mandibular access osteotomy	Classical	18	Nil
3	25	Male	Left cheek	Asymptomatic, nonfluctuant swelling, firm in consistency, free overlying skin	MRI with contrast showed well defined enhancing soft tissue mass	Enucleation	Classical	18	Nil
4	55	Male	Right cheek	Asymptomatic swelling of 4 months duration	MRI with contrast showed well defined enhancing soft tissue mass	Enucleation through mandibular access osteotomy	Ancient	5	Nil
5	56	Male	Left cheek	Asymptomatic swelling of 6 months duration	MRI with contrast showed well defined enhancing soft tissue mass	Enucleation	Classical	20	Nil

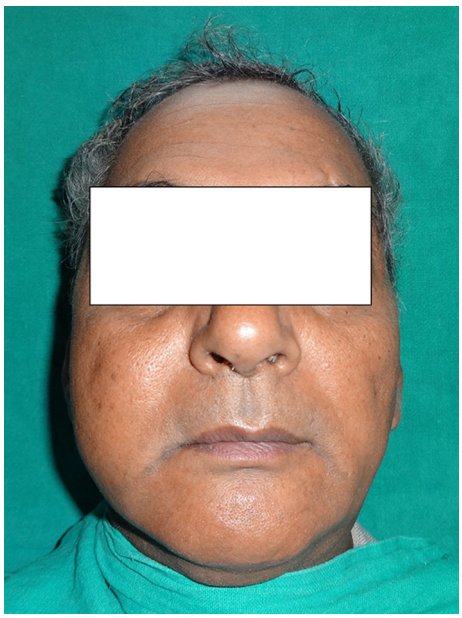

Fig. 1 Preoperative picture showing swelling on cheek

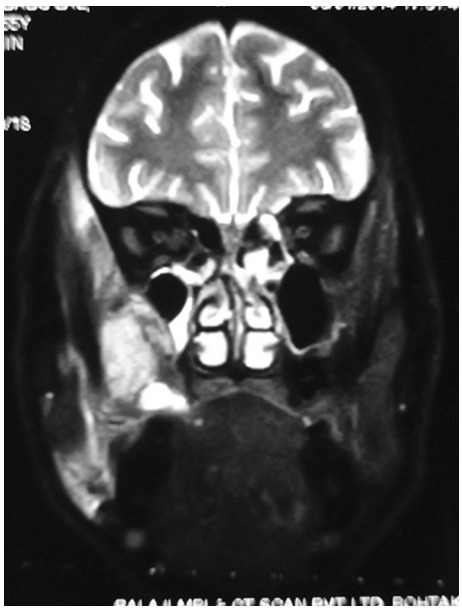

Fig. 2 Preoperative MRI Coronal image showing a well defined mass on right side between medial and lateral pterygoid muscles (along the course of mandibular nerve) which is hyperintense

Both sexes are probably affected by schwannomas, a study by Torossian et al. [7] observed female predominance in extracranial cephalic schwannomas. All the patients in our study group were males which co-relates with that of Leu and Chang's series [8] noting male predilection. Schwannomas may occur at any age. However they are more often seen in the second and third decades [9]. In cases of extra-cranial trigeminal

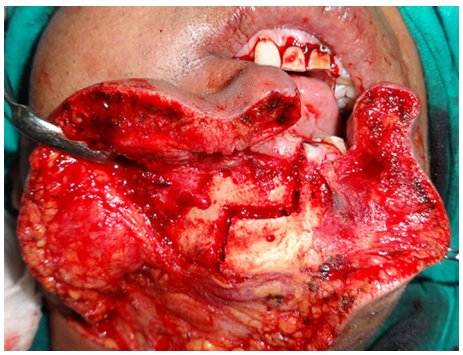

Fig. 3 Access osteotomy to approach tumor mass

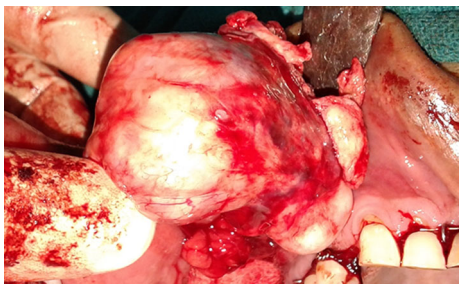

Fig. 4 Exposed tumor mass

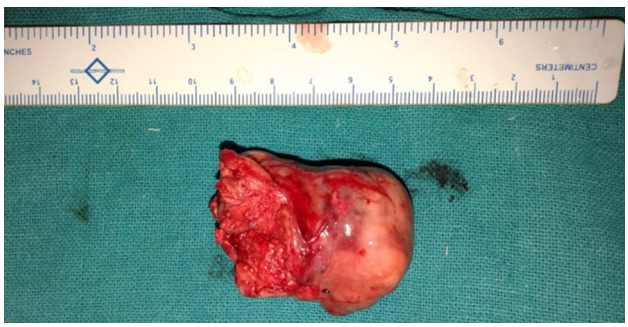

Fig. 5 Excised tumor mass

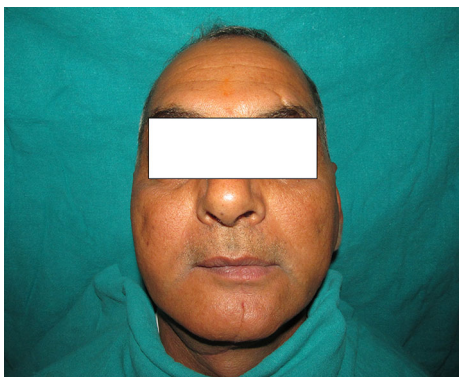

Fig. 6 Follow up

schwannoma, these were consistent with our two cases though three patients were in the fifth decade of life.

Clinical symptoms are of partial value in the pre-operative diagnosis of trigeminal schwannoma. Symptoms such as facial pain and numbness are usually absent. Neurological symptoms such as cramps, paralysis, numbness, are lacking. Mouth opening difficulty is generally not seen, the patient may be asymptomatic, and the lesion may be found incidentally [10]. They slowly enlarge until they cause functional or esthetic limitations. Similar findings were found in all patients in present study except one. In that case, patient also reported some limitation in mouth opening and was associated with pain. It may be due to the size of the lesion which was exerting pressure on jaw opening and closing muscles.

The neural origin has been of particular interest to clinicians. The nerves most commonly involved in schwannomas of the head and neck are the vagus and the cervical sympathetic chain [3]. Trigeminal schwannomas are rare tumours and extension into parapharyngeal space is extremely uncommon, reported only a few times in the literature [11]. In present study, all the lesions were diagnosed as schwannoma pre-operatively (based on radiographic features). In our study most of the lesions were present on the face i.e. in the cheek region. In this region, anatomically two nerves are present; these are the branches of facial nerve and trigeminal nerve. It was intra-operative finding which suggested that the lesion was originating from the terminal branch of trigeminal nerve and lack of postoperative neurological deficit confirmed it. In our one case, the tumor was in parapharyngeal space. In the parapharyngeal space, MRI plays a vital role to differentiate between schwannomas of trigeminal and vagus. In MRI, if the epicentre of the tumor mass is from the vagus then the carotid sheath will be dilated and internal carotid will be shifted away from the nerve, while those occurring in the trigeminal one will cause deviation of the carotid sheath along with its content as was seen in the parapharyngeal case of the present study.

Though the diagnosis of schwannoma is confirmed by histopathological examination, neuroimaging can play a major role in establishing the diagnosis and in planning the surgical approach. Asaumi et al. [12] reported the characteristics of advanced imaging findings associated with a common schwannoma of the upper lip. They indicated that computed tomography will show a soft tissue mass, with clear margins and areas of cystic degeneration, depending on the variant. MRI is the gold standard for evaluating trigeminal schwannomas. MRI characteristics [13] of schwannomas include specific signs (split fat sign, fascicular sign, target sign) and signal patterns i.e. tumours appear isointense or slightly hyperintense on T1-weighted images, and have high signal intensity on T2-weighted images, with significant enhancement after contrast injection [11]. In our study, MRI showed masses which were iso-intense or slightly hyperintense on T1-weighted images and had high signal intensity on T2-weighted images with significant enhancement after contrast injection. Based on tumour location and MRI signal characteristics, the radiological diagnosis of schwannoma was suggested in all the cases preoperatively.

The diagnosis of a schwannoma though can be established by neuroimaging, it is confirmed by histologic examination [6]. Histologically, many schwannoma variants have been described, including common, glandular, plexiform, cellular, epithelioid, melanotic, and ancient schwannomas [14].

The microscopic portrait of schwannoma is distinct and can hardly ever be confused with that of other lesions. In classical variant, the core of the tumor is composed of a mixture of two cellular patterns Antoni A and Antoni B [15–17]. Antoni A areas are collection of compact spindle cells with twisted nuclei arranged in bundles or fascicles. In extremely differentiated areas there may be nuclear palisading and formation of verocay bodies, which are formed by arrangement of two rows of nuclei and cell processes which assume oval shape. Antoni B variant is less cellular and less organized, representing degenerated Antoni A areas composed of randomly arranged spindle or oval cells within myxoid, loosely textured, hypocellular matrix punctuated by microcyst, inflammatory cells and delicate collagen fibers [15–17].

Ancient schwannoma exhibits benign degenerative changes in a classic schwannoma, taking place over a period of time. This change comprises cystic, myxoid, edematous and fibrotic areas, vascular abnormalities and atypical cells with pleomorphic nuclei. Ancient schwannomas behaves much like a benign neural neoplasm [17]. In the present study, four cases were of classical schwannoma while one was of ancient variant.

Simple excision or enucleation of the tumor is the indicated treatment. As a rule, schwannomas are well encapsulated and excision is easily performed. When the tumor involves a large nerve, separation and preservation of the nerve during surgical removal deserves special attention by the surgeon. Careful stripping of the nerve bundles from the surface of the growth will permit enucleation of the tumor in majority of the cases.

The literature suggests that the nerve should be detached from these tumors and preserved when the lesion is being removed [18]. However, some reports state that the nerve of origin should be resected due to the possible risk of recurrence, even if the tumor is benign [19]. In the present study, all the patients were treated with enucleation of the tumor mass and the nerve was preserved. The postoperative

Table 2 Review of literature of non-osseous schwannoma of oral cavity

Investigator	Gender	Age (year)	Site	Duration (year)
Reported cases of schwannoma in oral cavity				
Eversole and Howell [21]	Female	58	FOM and ventral tongue	Unknown
Marks et al. [22]	Female	65	Right FOM	Unknown
McCoy et al. [23]	Female	36	Maxillary left posterior mucobuccal fold	Unknown
Dayan et al. [24]	Female	52	Upper left vestibule	Unknown
Nakayama et al. [25]	Female	40	FOM and ventral tongue	2 month
Ledesma et al. [26]	Female	21	FOM and ventral tongue	5 month
Chen et al. [27]	Male	34	Left FOM	18 year
Subhashraj et al. [28]	Male	18	Mandibular posterior vestibule	8 month
Humber et al. [29]	Female	82	Upper right lip	Long standing[a]

Treatment–in all cases surgical excision was done

FOM floor of mouth

[a] Unknown duration but longer than 2 years

period showed no sensory as well as motor functional disability in all the cases.

The involvement of nerve branch and location of these tumors decide the choice of the surgical approach. A number of surgical approaches have been proposed for the treatment of extracranial trigeminal schwannomas, including transmandibular, transmaxillary, facial translocation, transpterygoid, infratemporal fossa and orbito-zygomatic approach [11]. These approaches, provide enough exposure of the skull base and therefore of the higher part of the tumour. In the present study, two patients required access osteotomy. In these patients, as the tumor mass was extending to infratemporal region and parapharyngeal region, transmandibular approach was taken to expose and completely excise the tumor mass. In two of the patients transoral approach was taken while in one patient transfacial approach was taken to expose and excise the tumor.

A schwannoma does not recur if totally excised and the prognosis is usually outstanding. No recurrence was seen in any cases till date in the present study. Malignant transformation is rare [20]. These tumors are highly radioresistant and radiotherapy has no place in the therapeutic management [18]. A small review (Table 2) also shows the same i.e. schwannoma if totally excised, the prognosis is usually outstanding.

To summarize, any long standing asymptomatic swelling of cheek should include trigeminal schwannomas as the differential diagnosis. The diagnosis though confirmed by the histologic examination but can also be made on the basis of MRI finding. As the tumor mass is well encapsulated, irrespective of the size; simple excision of the tumor can be done. Neural function can be preserved by simple excision though it may require complex access osteotomy.

Compliance with Ethical Standards

Conflict of interest None.

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