Trigeminal schwannoma

The trigeminal schwannoma most commonly arise from the gasserian ganglion, but may also originate more proximally from the trigeminal root or more distally from one of the three post-ganglionic divisions of the trigeminal nerve; the ophthalmic nerve branch is the site of origin more frequently than either the maxillary nerve or mandibular nerve branches.

Epidemiology

Trigeminal schwannomas (TSs) comprise 0.07-0.36% of all intracranial tumors and are the second most common type of intracranial schwannoma following the vestibular schwannoma, which account for 0.8-8% of intracranial schwannomas, and the dumbbell-shaped subtype is the most challenging.

Classification

The first classification system was proposed by Jefferson in 1955 who categorized TSs into three different types:

Type A, which described tumors originating from the gasserian ganglion in the middle cranial fossa;

Type B, which comprised of tumors originating from the roots of the trigeminal nerve in the posterior fossa; and finally, Type C, or the so-called “hour-glass” tumors, which occupy both the middle and posterior fossae.

Some authors have added a fourth classification, Type D, tumors with extracranial extension.

In 1986, Lesoin et al. classified TSs into three categories: Type I schwannomas, which originate from the roots of the posterior fossa

Type II schwannomas, which originate from the gasserian ganglion

Type III schwannomas, which originate from the trigeminal branches.

Yoshida and Kawase proposed a classification that categorized TSs into six types:

Type P, which comprise posterior fossa tumors originating from the root of the trigeminal nerve

Type M, which comprise middle fossa tumors originating from the gasserian ganglion or the peripheral branch at the lateral wall of the cavernous sinus

Type E, which include tumors arising from the extracranial peripheral branches of the trigeminal nerve

Type MP, ME, and MPE, which indicate a combination of P, M, and E tumors.

Jeong et al. modified Kawase's classification to offer information about the locational predominance, shape, and extension of the tumor into the adjacent compartment by representing them with capital (primary location) and lowercase letters (extension).

Clinical features

Trigeminal nerve dysfunction is a presenting complaint in 70-95% of patients with TS, and includes

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numbness and/or paresthesia in one or more of the three trigeminal branches, facial pain, and atrophy of the masseter muscle. One study reported facial hypesthesia in 11 of 16 patients (68.8%) by the time of diagnosis, while another study reported 13 of 27 patients (48.1%) had facial hypesthesia preoperatively.

In a study, facial hypesthesia was observed in 76.2% of the patients. The reported incidence of facial pain at presentation varies from 10% to 45% [8,13,14,15], and can be either typical trigeminal neuralgia or atypical facial pain. The facial pain seems to be associated more with tumors originating from the gasserian ganglion than those from the trigeminal nerve roots, although there are controversies.

In the case series, facial pain was more frequent in tumors originating from the middle fossa (40.0%) than from the posterior fossa (15.0%). It has also been suggested that constant facial pain is more associated with middle fossa schwannomas, which may be attributed to the relatively fixed position of the gasserian ganglion in the trigeminal impression of the petrous bone, while intermittent facial pain is closely related to posterior fossa schwannomas compressing more mobile trigeminal roots. Weakness of the temporalis and pterygoid muscles was reported in up to 60% of cases in one large series, however, most series have reported an incidence between 25% and 35%.

Trigeminal nerve-related symptoms, such as numbness, facial pain, and wasting of the masseter muscle are useful in diagnosis TSs. Although trigeminal nerve dysfunction is a common presentation in TSs, some patients can be asymptomatic. Given that TSs grow slowly with vague initial clinical features, diagnosis of TS is often delayed and most of the tumors are found to be large in size at the time of operation.

**Diagnosis**

An observational study of the diffusion tensor imaging tractography (DTT) results and intraoperative findings was performed. Wei et al. preoperatively completed tractography from images of patients with TSs who received surgical therapy. The result was later validated during tumorectomy.

A total of three consecutive patients were involved in this study. The locations of CNs V-VIII in relation to the tumor was clearly revealed in all cases, except for CN VI in case 3. The predicted fiber tracts were in agreement with intraoperative observations.

In this study, preoperative DTT accurately predicted the location of the majority of the nerves of interest. This technique can be applied by surgeons to preoperatively visualize nerve arrangements.

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.

**Differential diagnosis**

Chordomas located in the Meckel's cave can be radiologically confused with trigeminal schwannomas.

Syphilis mimicking trigeminal schwannoma.
Treatment

Trigeminal schwannoma treatment.

Surgical outcomes

Advances in surgical techniques over the last 50 years have been mirrored by better resection rate with reduced mortality and morbidity. For example, Jefferson described six operative cases that included two deaths and one recurrence in 1955. In 1960, Schisano and Olivecrona reviewed nine surgical cases with a mortality rate of 40%. A study by Konovalov et al. of 111 cases, of which 28 were performed between 1962 and 1977, reported a total resection rate of 68% and a 7% mortality rate. Following the introduction of the microscope, the total resection rate improved to 78% with a mortality rate of 1% between 1978 and 1989. The application of skull base techniques between 1990 and 1994 further improved the total resection rate to 87% with zero mortality. Overall, there has been an improvement in total resection rates from as low as 33%, with conventional approaches, to 84-100% with skull base techniques. This can advance be easily visualized in the total resection rate reported by Yoshida and Kawase, which was 33% with conventional approaches and 94% for skull base approaches. Moreover, Sarma et al. reported 26 cases that were all completely resected using skull base techniques. The skull base technique offers more success as it provides a wide exposure of the mass and facilitates more complete resection of the tumor. In the current series, the total resection rate was 95.2% with no mortalities. The most frequent reasons for residual tumors were that they had infiltrated into the cavernous sinus or that they adhered to the brainstem. It should be noted that, even when complete microscopic resection is thought to have been achieved, small pockets of tissue can still be left in the cavernous sinus, and this is often the most common site of recurrence. In the current study, gross total resection was achieved in 47 patients (95.9%) with no mortality. Of the two cases of subtotal resection, one was a large-sized MP-type tumor, which was firmly adhered to the dura of the posterior fossa, while the other case was a Pm-type tumor where the residual tumor was found in Meckel's cave by MRI after surgery. In the latter case, the residual portion of the tumor was not visible with a microscope, and adjuvant cyberknife radiosurgery was applied 28 months after the initial operation.

Facial pain has been reported to be relieved after surgery in 73% to 100% of cases. However, only 19-44% of patients show an improvement in facial hypesthesia. Furthermore, trigeminal motor dysfunction, which includes wasting of the pterygoid and temporalis muscles, show poor recovery rates. In this series, 81.8% of patients showed an improvement in facial pain, whereas only 11.4% recovered from facial hypesthesia. In terms of masseter weakness, 60% of patients recovered from the symptom, but this result might be attributed to recall bias or limitations of the retrospective study. Meanwhile, tumors that were predominantly located in the middle fossa showed better postoperative trigeminal functional outcomes than of those in the posterior fossa, suggesting that preganglionic nerve injury was more critical in trigeminal dysfunction than postganglionic damage.

Recurrence

The recurrence rate of TSs in older surgical series before 1990 ranged from 0% to 25%, while more recent series have reported recurrence rates of 0-17%. Across all series, recurrence rate was higher in larger schwannomas. According to the literature, the most common site of recurrence is the cavernous sinus, followed by Meckel's cave. Time taken to recur is highly variable, ranging from 1 to 9 years. The most important factor predicting recurrence has been identified as the completeness of surgical resection.

In conclusion, comprehensive knowledge of the anatomical features of TSs and the selection of the
most appropriate surgical approaches are essential to achieve complete resection. TSs are well to be classified with our modified classification and able to be removed effectively and safely by selecting appropriate surgical approaches \(^\text{19}\).

## Case series

### Trigeminal schwannoma case series.


11) http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4231622/ 14


14) Akay A, Göde S, Çağlı MS. Neuronavigation-guided endoscopic endonasal excision of schwannoma-like