Giant Trigeminal Nerve Schwannoma Operated in Two Steps: case report

Schwannoma Gigante de Nervo Trigêmeo Operado em Duas Etapas: relato de caso

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ABSTRACT

Trigeminal schwannomas are very rare, representing 0.36% of all intracranial tumors. The symptoms associated with these tumors are usually related to a reduction in the sensitivity of the innervated territory, as well as facial pain and a reduction in the corneal-eyelid reflex. We describe the case of a 21-year-old male patient who presented with hypoacusis and hypoesthesia in the left face, left parietal headache, as well as difficulty chewing and anxiety. On investigation, magnetic resonance imaging showed extensive formation in the posterior fossa with extension to Meckel’s cave, middle fossa and pterygopalatine. Due to the extent of the tumor, the patient underwent two surgical resections. Tumor pathology was compatible with trigeminal nerve schwannoma.

Keywords: Neoplasm; Neuroma; Schwann cells; Trigeminal nerve; Neurosurgery

RESUMO

Os schwannomas do nervo trigêmeo são muito raros, representando 0.36% de todos os tumores intracranianos. A sintomatologia associada a esses tumores geralmente está relacionada à redução de sensibilidade do território inervado, assim como à dor facial e à redução do reflexo córneo-palpebral. Descreve-se o caso de um paciente do sexo masculino, 21 anos, o qual apresentava hipoacusia e hipoestesia em face esquerda, cefaleia parietal esquerda, assim como dificuldade para mastigar e ansiedade. Na investigação, imagens de ressonância magnética mostraram formação extensa na fossa posterior com extensão para caverna de Meckel, fossa média e pterigopalatina. Devido à extensão do tumor, o paciente foi submetido a duas ressecções cirúrgicas. O exame anatomopatológico tumoral foi compatível com schwannoma de nervo trigêmeo.

Palavras-Chave: Neoplasia; Neuroma; Células de Schwann; Nervo trigêmeo; Neurocirurgia

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INTRODUCTION

Trigeminal schwannomas are very rare, representing 0.36% of all intracranial tumors and 8% of all intracranial schwannomas1-3. These tumors can develop in any segment of the trigeminal nerve, being more common in the trigeminal ganglion1. Samii et al.2 classified the extent of these tumors into 4 types: type A, intracranial tumors predominant in the middle fossa; type B, intracranial tumors predominant in the posterior fossa; type C, intracranial tumors in the form of a dumbbell with components of the middle and posterior fossa; and type D, extracranial tumor with intracranial extensions.

In most studies, the classification of schwannomas according to size considers as small a neuroma of up to 2 centimeters, an average of up to 3 centimeters, a large up to 4 centimeters and a giant being larger than 4 centimeters2,4. Large brain tumors are usually a challenge for treatment, as they often cause distortion of the brainstem and cerebellum5. In this sense, they are associated with higher rates of surgical mortality6, as well as morbidity, because in surgeries usually occurs damage to arteries and cranial nerves5. Due to such difficulties, the staged removal of large brain tumors has been analyzed in several researches. Although not absolutely indicated, this technique should be recommended for large volume tumors, especially when they are deeply installed in the brainstem and cerebellum5.

CASE REPORT

Male patient, 21 years old, presented with symptoms of hypoacusis and hypoesthesia on the left cheek, left parietal headache, difficulty of chewing and anxiety. In the investigation, magnetic resonance imaging (MRI) showed extensive left formation in the posterior fossa with extension to the Meckel’s cave, middle fossa and pterygopalatine (Figure 1). Due to the extension of the tumor, the patient underwent two surgical resections. In the first surgery, subtotal resection of the tumor was performed in the region of the cerebellopontine angle. In the postoperative period, an MRI showed a large left heterogeneous expansive formation, extending to the left middle and pterygopalatine fossa (Figure 1). Due to the extension of the tumor, the patient underwent two surgical resections. In the first surgery, subtotal resection of the tumor was performed in the region of the cerebellopontine angle. In the postoperative period, an MRI showed a large left heterogeneous expansive formation, extending to the left middle and pterygopalatine fossa, which caused enlargement of the foramen ovale and bulging over the temporal pole. After 45 days, the second surgery was performed, which resulted in resection of the tumor in the left temporal fossa. After both surgeries, the patient presented with abducent nerve palsy, diplopia and left keratitis. Anatomopathological results were compatible with trigeminal schwannoma.

After 6 months of the last surgery, an MRI showed an area of contrast enhancement in the region of the left internal auditory canal of 5x3 mm, in addition to left temporal gliosis (Figure 2). After one year of the last appointment, the patient returned for medical follow-up, reporting persistence of difficulty in chewing due to masseter atrophy and anxiety. On physical examination were found isophotoreagent pupils (RFIP), preserved extrinsic ocular musculature, click on the left temporomandibular joint and hypoesthesia in the left hemiface. MRI showed no alterations in relation to the examination of the previous consultation. The patient was referred to an otorhinolaryngologist and instructed to return after one year.

DISCUSSION

Trigeminal nerve schwannoma usually appears in young adults, and the mean age of onset of symptoms is 40 years2,7,8. In up
to 95% of patients with this tumor, symptoms suggestive of trigeminal dysfunction are present, and may present numbness and/or paraesthesia in at least one of the three branches of the trigeminal nerve. Malignancy of the tumor should be suspected when there is complete anesthesia of the three branches. In the case presented, the patient reported hypoesthesia in left hemiface, indicating involvement of the three branches of the trigeminal nerve.

Another frequent feature of trigeminal nerve schwannoma is the reduction or abolition of the corneal-eyelid reflex, occurring in up to 72% of patients. In addition, almost half of patients with trigeminal schwannoma (45%) present pain in the face, and this pain may be a typical or an atypical facial pain. Aspects of the duration of this pain may help in the localization of the trigeminal schwannoma, because some studies suggest that a constant pain indicates a tumor in the middle fossa, while intermittent pain is related to schwannomas of the posterior fossa.

In cranial computerized tomography and MRI, the trigeminal nerve neuromas generally present well-defined limits and homogeneous contrast uptake. However, less often, schwannomas may present heterogeneous contrast uptake, as in the case presented.

The treatment of trigeminal schwannomas is based on microsurgical resection, radiotherapy and follow-up. The option of microsurgical resection is usually the most used for the treatment of trigeminal neuromas, being considered the choice in up to 80% of cases. However, the results of this surgical modality depend on the presence of preoperative symptoms, as well as the size and location of the tumor. Studies suggest that, in some cases, microsurgical resection increases the deterioration of the trigeminal nerve, resulting in symptoms such as abducent nerve palsy and trigeminal neuralgia.
Follow-up by imaging is usually the treatment of choice for small size tumors found accidentally, since these patients are asymptomatic. Radiotherapy is designated for patients with small to moderate size tumors who have preserved trigeminal nerve function or in cases of isolated nerve dysfunction. In this sense, the treatment option should be carefully analyzed for each case, depending on the size and location of the tumor, the risks to the patient and the expected results.1,2,3

The staged removal of tumors is usually taken into account in cases of large tumors, especially those that reach the posterior fossa. In addition, removal of tumors in more than one step prevents possible damage to intracranial structures, which can be caused by decompression and displacement.4 A possible explanation for this avascular state of the residual tumor lies in the fact that the first surgical manipulation may trigger some mechanism of tumor thrombosis, reducing the blood supply.5,6 Although it involves additional surgery, staged removal of brain tumors does not provide increased morbidity and mortality when compared to single-stage removals.7 In the case of this patient, a microsurgical section of the tumor was performed in two stages. In the first surgery was performed a microsurgical section of the tumor was performed, and in the second in the left temporal fossa.

According to Lazard et al., keratitis is a common postoperative complication of tumors in the region of the cerebellopontine angle, occurring due to the damage caused to the superficial petrous nerve, which causes damage in ocular humidification. Abducent nerve palsy, worsening trigeminal nerve deficits and headache are some of the post-surgical complications of trigeminal nerve schwannomas.8,9,10,11,12,13 We have shown in this report that our patient postoperatively presented abducent nerve palsy, diplopia and keratitis in the left side, being a drawback compatible with the literature.

CONCLUSION

Trigeminal nerve schwannomas are extremely rare and are more frequent in young adults. Symptoms of the presence of this tumor vary between paresthesia, and facial pain, however, due to the compressive effect of the tumor, dysfunction of other cranial nerves may occur. In most cases, microsurgical resection is the treatment of choice, although studies have pointed to post-surgical symptoms of abducent nerve palsy and trigeminal neuralgia. Staged surgical resection can be a considerable option in cases of large volume tumors, as it facilitates the removal of these tumors, in addition to preventing complications. In this sense, the treatment of these tumors should be carefully analyzed, considering the size and location of the tumor, as well as the risks and expected outcome.

REFERENCES

Case Report

Rossa ICM, Chaves JPG, Mattozo CA - Giant Trigeminal Nerve Schwannoma Operated in Two Steps: Case Report


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