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Case Report

Cystic trigeminal schwannomas

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ABSTRACT

Trigeminal Schwannomas are less than 1% of intracranial tumors, of which only 7% have a cystic component. We documented 2 cases of males with cystic trigeminal Schwannomas, their symptoms, the diagnosis process and the imaging characteristics. In addition, a review of the literature is performed, with emphasis on the radiological classification of this rare entity, that constitutes a diagnostic challenge for the radiologist, who has an essential role in the approach to the disease and therefore in its management.

Introduction

Schwannomas are benign tumors of the myelin sheath, originating from Schwann cells. Approximately 25%-40% occur in the soft tissues of the head and neck [1], usually from the vestibulocochlear nerve. Trigeminal nerve is the second most common intracranial location [2,3], however these is a rare entity, responsible only for 0.07%-0.36% of intracranial tumors [4,5]. According to their radiological characteristics, schwannomas are classified as solid or cystic, with cystic presentation being the most unusual, with a reported incidence of less than 7%, of all trigeminal schwannomas [4,6]. Two cases of trigeminal cystic schwannoma are documented, and a review of the literature is performed emphasizing its clinical presentation, magnetic resonance imaging (MRI) imaging features and its classification.

Case reports

Case 1

A 31 years old man with 5 months of a left fronto-temporal cephalea, associated with blurred vision, paresthesia in the left side of the face and pain in the ipsilateral molar region. No relevant findings were found at the physical examination. In the MRI, an expansive lesion was observed in relation to the greater wing of the left sphenoid, with well-defined margins, diploic remodeling and extension through the foramen ovale, with a dumbbell shape. The lesion had dimensions of 16 × 14.7 × 21 mm (L × AP × T). It presented predominantly cystic features, been hyperintense in T2 and FLAIR sequences, and hypointense in T1, with intralesional foci suggestive of cellular proteinaceous detritus. Post-gadolinium

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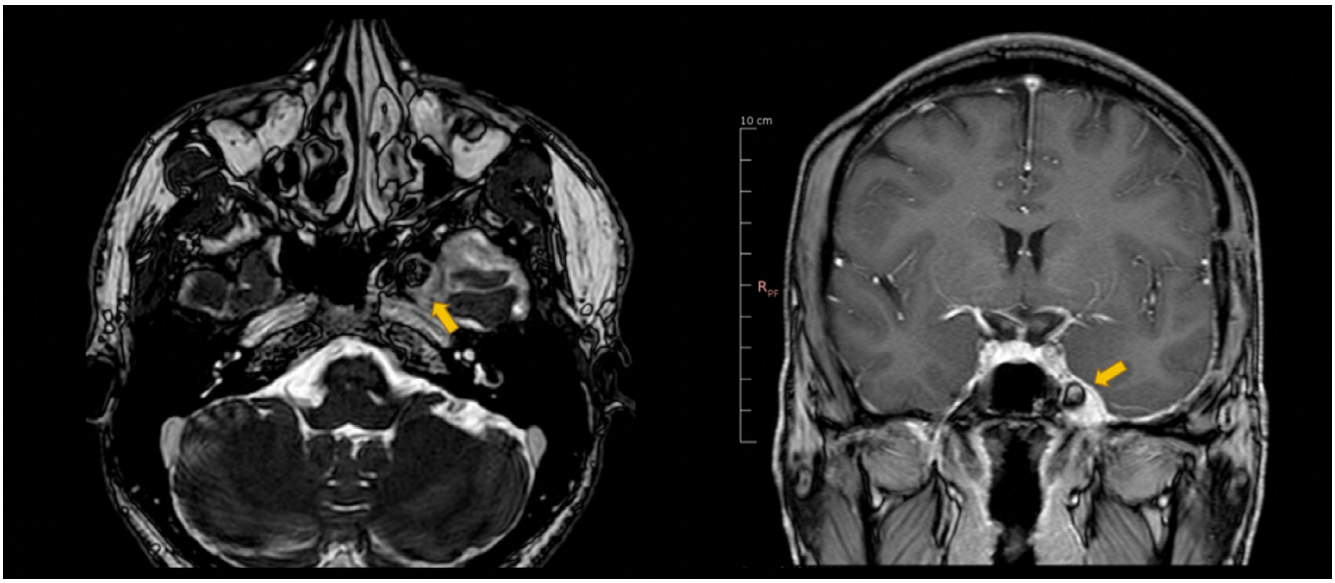

Fig. 1 – Left axial FIESTA (Fast Imaging Employing Steady-state Acquisition). Right coronal T1-weighted post-gadolinium magnetic resonance imaging. Arrows showing CTS.

MRI showed a peripheral ring enhancement, associated with hyperintense focal thickening of the adjacent dura mater. In the diffusion-weighted imaging (DWI) sequence, areas with restriction to diffusion were not identified (Fig. 1). Based on the imaging findings, a diagnosis of trigeminal cystic schwannoma was suggested, which was confirmed histopathologically after complete surgical resection. 18 months follow-up showed no recurrence of the tumor and the patient presented partial improvement of the initial symptomatology, especially facial paresthesia.

Case 2

Male patient of 68 years, with 4 months global headache episodes, which changes its characteristics increasing its intensity and localizing in the right parietoccipital region. MRI images were obtained, which showed the presence of a nodular mass in the Meckel cave, which widened the inferior contour of the cavernous sinus. Inside the nodulations it presented a granular appearance with partitions and liquid content in a seemingly capsule. The lesion showed no signs of infiltration of adjacent structures and had dimensions of $13 \times 15 \times 3$ mm (L $\times$ AP $\times$ T); findings suggest a mass originating from meningeal envelopes (Fig. 2). A partial tumor resection was performed, and schwannoma histopathological confirmation was subsequently obtained. The patient had no symptoms ameliorations and refuse to a new surgical intervention.

Discussion

Trigeminal schwannomas are a rare entity, difficult to diagnose clinically, because it can develop in any part of the trigeminal nerve, from the cistern to its peripheral divisions, giving a varied symptomatology. Given its low incidence, there are few case series where the clinical presentation has been evaluated. Guthikonda et al. analyzed 23 patients with trigeminal schwannoma, describing the initial symptoms being headache (74%), diplopia (22%), and auditory alterations (17%). In the physical examination, the most common finding was trigeminal sensory deficit in 87% of patients, while motor weakness was found only in 39% [7]. Yoshida and Kawase, conducted a review of 241 patients, finding that more than 70% of the patients presented trigeminal nerve dysfunction, mainly trigeminal hypoesthesia [8]. Additionally, the presence of extratrigeminal symptomatology is common, specially the paralysis of the abducens nerve (26%) [9–12].

Despite being a solid neoplasm, schwannomas can present cystic degeneration. Different causal mechanisms have been proposed, such as intratumoral bleeding, central ischemic necrosis and degenerative changes. No relation has been established with cystic degeneration and location of the schwannoma or size of the lesion. There are reports of other rare causes such as the formation of an associated arachnoid cyst and the presence of glandular or pseudoglandular elements in the tumor.

Radiological findings

Intracranial schwannoma presents in CT as an isodense to hyperdense mass that generates an increase in foramen ovale size, with erosion and remodeling. When IV contras medium is injected, it has heterogeneous enhancement. In the trigeminal cystic schwannoma, the cystic portion is identified as a hypodense area, without enhancement.

MRI shows an isointense mass when compared to the brain in T1-weighted images, with homogenous enhancement after the administration of contrast medium, while in T2-weighted images the lesion is hyperintense. Additionally, in the trigem-

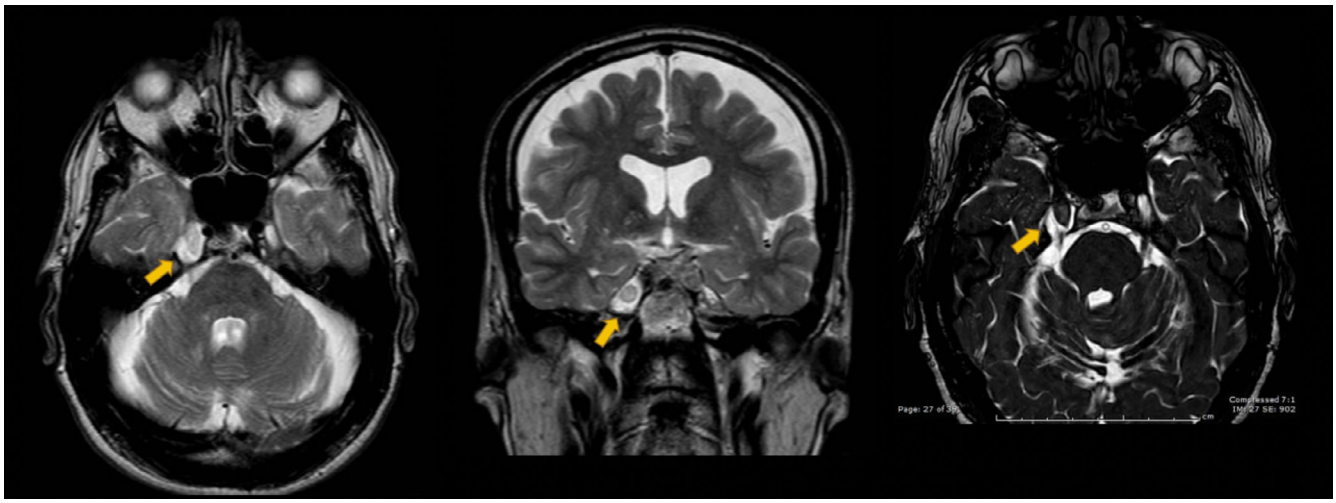

Fig. 2 – Axial and coronal T2-weighted magnetic resonance imaging. Right; axial FIESTA (Fast Imaging Employing Steady-state Acquisition). Arrows showing CTS.

inal cystic schwannoma the cystic portion is identified as a hypointense area in T1, without enhancement after administration of gadolinium and isointense when compared to the solid portion in T2 sequences. When evaluating the mass in FLAIR, the nodular and cystic portion has high signal intensity [1–5].

The fluid-fluid levels are exceptionally rare in intracranial schwannomas. Its formation occurs when two liquid substances of different composition and density are within the cyst. The hypointense signals that are usually located in the dependent layer in T2-weighted images, and are related to the selective shortening in T2 caused by intracellular paramagnetic substances such as deoxyhemoglobin or methemoglobin; while serum and plasma, which are free of methemoglobin, assume a nondependent position and show high signal intensity in T1 and T2 enhanced images [13].

Classification

Trigeminal schwannomas can be classified in multiple ways. The first, proposed by Wanibuchi et al., [4] trigeminal schwannomas are divided depending on the origin of the lesion; he classifies them in peripheral, Meckel cave, posterior fossa root or dumbbell. Peripheral tumors are those originating in the extracranial portions of the trigeminal nerve, compromising its divisions V1, V2 or V3. Those classified as Meckel cave, have their origin in the Gasser ganglion. The tumors of the trigeminal cistern have their origin in the pons and are part of the tumors of the posterior fossa root type. Finally, dumbbell tumors include both cavernous root and cavernous peripheral subtypes.

There is a second classification based on the development patterns of these tumors. There are 3 types of tumors, which limit their development to 1 compartment, type M are tumors of the middle fossa, including those originating in the Gasser ganglion and peripheral branches, located on the lateral wall of the cavernous sinus; type P, where posterior fossa tumors are classified, originating from the trigeminal root and type E, which refers to extracranial tumors. The other types of tumors in this classification are those that extend in more than 1 compartment and are classified as MP, ME, and MPE, depending on the extent of the mass [8].

The last classification and currently the most used is the one created by Jefferson in 1995 [7,13,14], who describes 4 types of tumors, type A, where the tumor is located mainly in the middle fossa, type B, which is predominantly posterior. The dumbbell tumors that occupy the middle and posterior fossa are classified as type C and type D that have extracranial extension (Fig. 3).

Management and prognosis

The management of this entity has represented a therapeutic challenge due to the difficult surgical exposure of the lesions and the high possibility of recurrence. In recent years, thanks to new surgical techniques, there has been a decrease in morbidity and mortality [15]; while in 1960 a mortality of 40% was reported in a series of 9 cases, in more recent series the mortality oscillates between 0%-3% [7], this decrease is mainly due to the introduction of the microscope and the neurosurgical cranial base techniques.

The surgical approach to trigeminal schwannomas depends on their anatomic location. For intra and interdural tumors, cranial base approaches are preferred, since they allow a better trigeminal exposure, minimizing cerebral retraction [16,17]. On the other hand, in those tumors of V1, the temporopolar approaches are the ones of choice, while those originating in V2 and V3 infratemporal resection is preferred, allowing the exposure of the foramen ovale, the pterygopalatine ganglion and the infratemporal fossa [4,18]. For the lesions located in the Meckel cave, the pericavernosal lateral approach is preferred, with which the lesion of the trochlear and abducens nerves is diminished [4,19,20]. Finally, for pons tumors, suboccipital, retrosigmoid or latero-suboccipital approaches are used.

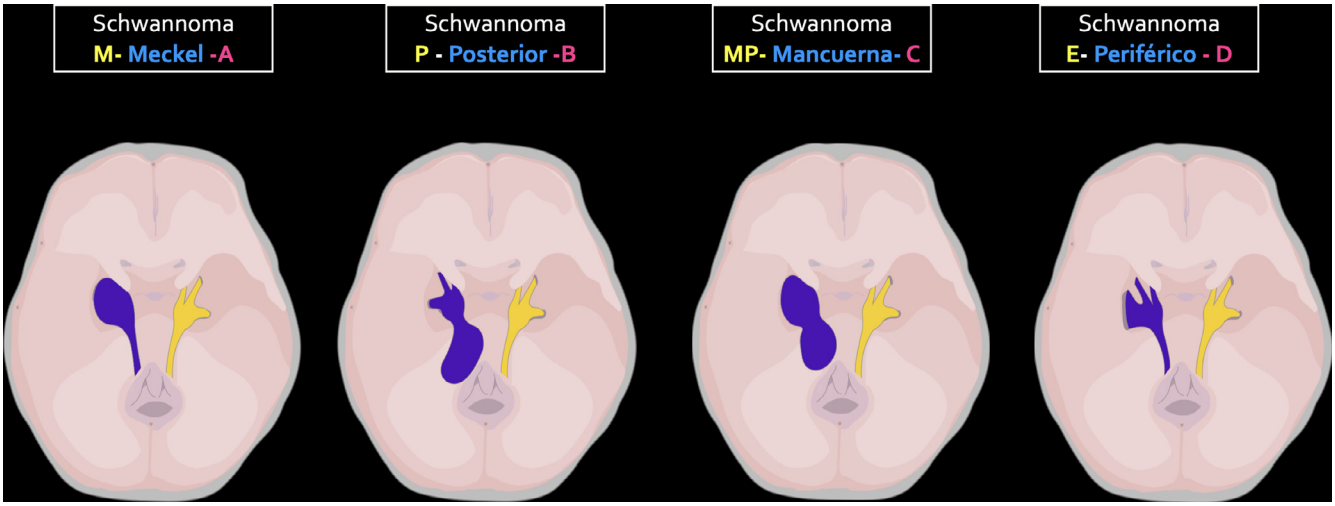

Fig. 3 – Classification of the CTS. Images show in purple the affected trigeminal nerve; in yellow normal trigeminal nerve. Nomenclature of each lesion by Jefferson (yellow), Wanibuchi (blue) and Yoshida (pink). (Color version of figure is available online.)

The recurrence of trigeminal schwannomas occurs mainly due to incomplete resection of the lesion. In the latest series of cases is a recurrence rate ranging from 0%-17%, in a period of 13 months to 8 years [4].

Another possibility for management is stereotactic radiosurgery, which in the last 20 years has been used as a primary treatment for tumors <3 cm, growth control and management of recurrences of trigeminal nerve schwannomas. stereotactic radiosurgeryis a technique that uses multiple beams of radiation converging to a volume of treatment, delivering a single high dose of radiation. This can be achieved with the Gamma knife or with a linear accelerator equipped for radiosurgery [21].

The treatment of schwannomas of intracranial localization with stereotactic radiosurgery has been increasing in recent years, because it offers advantages over conventional surgical management; a marginal dose of 12Gy to 13 Gy to treat tumors up to 3 cm in diameter have reported local control rates of 91% to 100% at 10 years and rates of trigeminal or facial nerve complications of less than 5%, with a hearing preservation from 60% to 70% of cases [22,23].

Conclusion

The trigeminal schwannomas are a benign pathology that has a low incidence in their cystic presentation. It constitutes a diagnostic challenge for the radiologist, who has an essential role in the approach to the disease and therefore in its management. It is important to take this into account as a differential diagnosis of intracranial cystic masses.

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