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Case Report

Multicompartmental cystic trigeminal schwannoma as an uncommon differential diagnosis of cerebellopontine angle tumors ☆

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ABSTRACT

Trigeminal schwannoma is the second most common intracranial schwannoma yet accounts for less than 0.5% intracranial tumors [1]. Cystic degeneration is uncommon. We would like to report a pathologically proven multicompartmental cystic trigeminal schwannoma in a young adult presenting with chronic headache. A literature review on the imaging features of trigeminal schwannoma is performed to assist radiologists in accurate disease localization and prioritizing differential diagnosis in challenging cases. Confident preoperative radiological diagnosis would directly affect management strategies.

Introduction

Schwannomas are benign, slow-growing neurogenic tumors arising from Schwann cells of the myelin sheath of the peripheral nervous system [2]. While smaller lesions may be asymptomatic, voluminous tumors may lead to radiating pain due to peripheral nerve compression or neurologic deficits specific to innervating tissues [3], providing clues to clinical and/or radiological diagnoses.

Intracranial schwannomas are predominantly vestibular nerve in origin, (accounting for more than 90% [4]), followed by trigeminal nerve. The latter constitutes only 0.8%-8% of intracranial schwannomas, corresponding to 0.07%-0.36% of intracranial neoplasms [5]. Cystic degeneration is rare, accounting for 7% of these lesions [6]. There is a predilection for middle-aged (40 to 60 years of age) women [7]. We have documented 1 case of a young male with multicompartmental cystic trigeminal schwannoma presenting with occipital headache. A literature review aimed at summarizing features of trigeminal schwannoma, providing a guide to radiologists to overcome the diagnostic challenge in differentiating these multicompartmental schwannoma with atypical features from other tumors at cerebellopontine angle and petrous apex.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Case report

A 33-year-old male with otherwise good pass health presented to the general practitioner for worsening occipital headache for 3 months, which is exacerbated with the Valsalva maneuver. Clinical examination revealed bilateral past-pointing and intention tremor.

Magnetic resonance imaging (MRI) shows a large 6.9 × 5.5 × 6.1 cm (long dimension x short dimension x craniocaudal) lobulated mass centered at the left cerebellopontine angle, extending from the left ambient cistern to the left premedullary space craniocaudally. The Meckel's cave is involved and expanded with a small extension into the left infratemporal fossa via foramen ovale. The mass is heterogeneous in appearance, comprising multiloculated T1 hypointense, T2 fluid attenuated inversion recovery (FLAIR) partially suppressible, nonenhancing cystic components with interspersed enhancing septa, as well as T1/2 isointense enhancing solid components (Fig. 1). Hemorrhagic foci, as evident by curvilinear signal losses in susceptibility weights imaging (SWI), with corresponding T1 hyperintense signals, are seen (Fig. 2). No evidence of diffusion restriction in diffusion-weighted imaging (DWI) was apparent.

Brainstem and left cerebellar compression, third and fourth ventricle, and associated lateral ventricular dilatation are evident. The cavernous sinus is displaced anteriorly. The left petrous internal carotid artery (ICA) is encased and mildly narrowed. There is no evidence of masticator muscle atrophy.

Upon reviewing the noncontrast computed tomography (CT) brain study immediately prior to admission, perilesional smooth bone remodeling of the left skull base (Fig. 3) and 2 punctate calcifications are evident.

The preoperative radiological diagnosis of cystic trigeminal schwannoma is suggested.

Gross total resection of tumor achieved by neurosurgical colleagues via combined fronto-temporal and retro-sigmoid craniotomy with intraoperative confirmation of tumor of trigeminal nerve in origin with V3 extension. Histopathology demonstrates alternating densely cellular and myxoid areas with nuclear palisading and Verocay bodies. Immunohistochemistry shows the expression of S100 and SOX10 proteins.

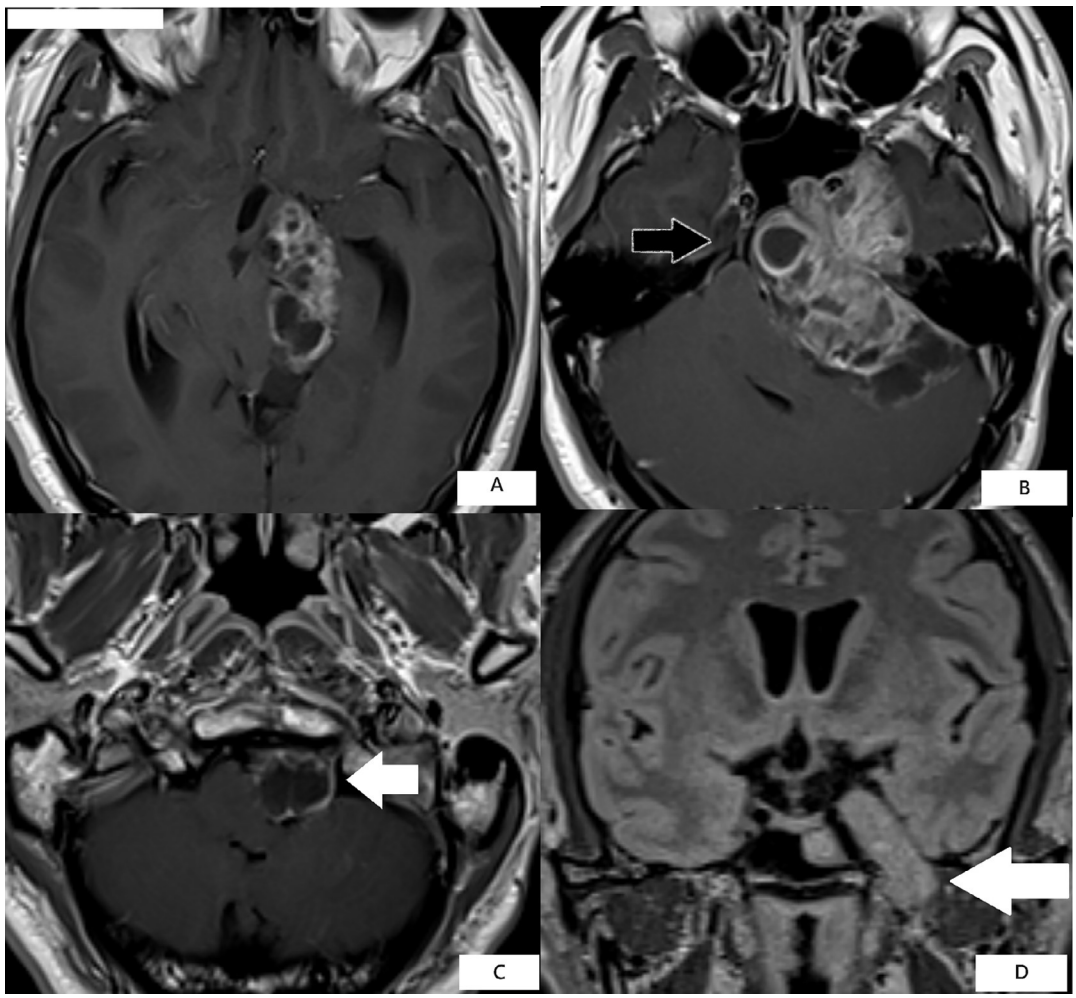

Fig. 1 – (A-C) Axial T1W with contrast imaging showing the extent of a mixed cystic solid heterogeneously enhancing lesion. Black arrow denotes the contralateral normal Meckel's cave in image B. White arrow in image C demonstrates the caudal cystic tumor component. (D) Coronal fluid attenuated inversion recovery (FLAIR) image demonstrates the left infratemporal tumor extension across the left foramen ovale (white arrow).

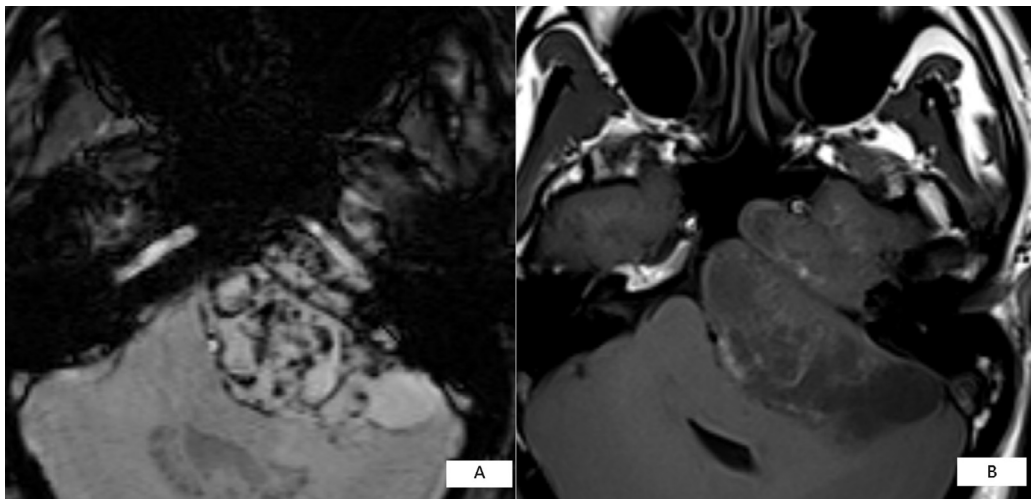

Fig. 2 – (A) Axial susceptibility weighted sequence (SWI) and T1W images show evidence of intratumoral microhemorrhage.

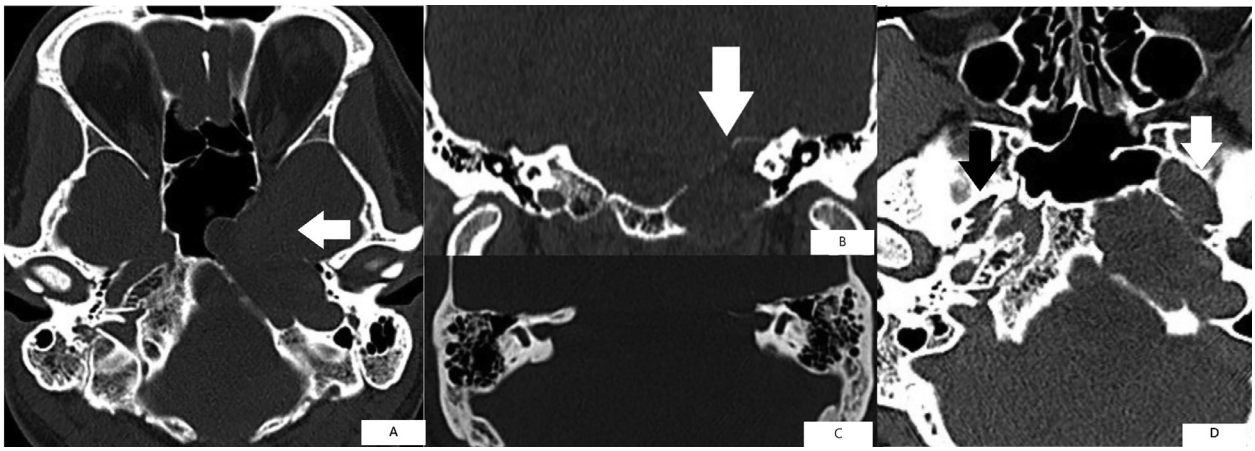

Fig. 3 – Axial (A,C,D) and coronal (B) non-contrast computed tomography (CT) brain images in bone window. Smooth erosion of left petrous apex and left side of clivus with expansion of left carotid canal (A, B; white arrow). In Figure D, there is smooth expansion of left foramen ovale (white arrow) with reference to normal contralateral foramen ovale (black arrow). The left inner ear structures are spared. Bilateral internal acoustic canals are symmetrical (C).

Features are compatible with schwannoma with regressive features.

Postoperatively, there is transient left abducent nerve palsy with exposure keratopathy, which is resolved at postoperative 3 months with ophthalmological team input. Persistent mild numbness in the left V1 territory has been reported.

Non-contrast CT brain at 3.5 months showed resolution of mass effect to posterior fossa structures and hydrocephalus (Fig. 4).

The final diagnosis was multicompartmental trigeminal schwannoma (MTS); type F as per Ramina et al. [8], type MPE as per Yoshida and Kawase [9] and type Db as per Wanibuchi et al. [6] classification.

Discussion

Cystic trigeminal schwannoma is the uncommon appearance of an uncommon disease. Instead of solid enhancing tumors, the heterogeneity in appearances of cystic trigeminal schwannoma is thought to be related to an increase Antoni B to Antoni A tissue ratio, as well as more dystrophic changes due to larger tumor size [2]. Although the patient's symptoms of occipital headache and subclinical ataxia can be compatible with trigeminal schwannoma [8], the absence of innervation territory-specific symptoms such as facial pain, numbness, or masticatory muscle weakness [10] render clinical diagnosis impossible. The epidemiological characteristics of young adult male also did not provide useful clues to the final diagnosis. The interpretation of the radiologist is therefore of paramount importance in localizing and mapping the disease extent, formulating the diagnosis based on imaging features despite uncommon appearances, as well as identifying the secondary effect and/or complications.

For radiological evaluation of intracranial neurogenic tumors in general, the imaging modality of the choice would be MRI with the aid of CT study. MRI prevails in its excellent tissue contrast, with accurate delineation of tumor extension

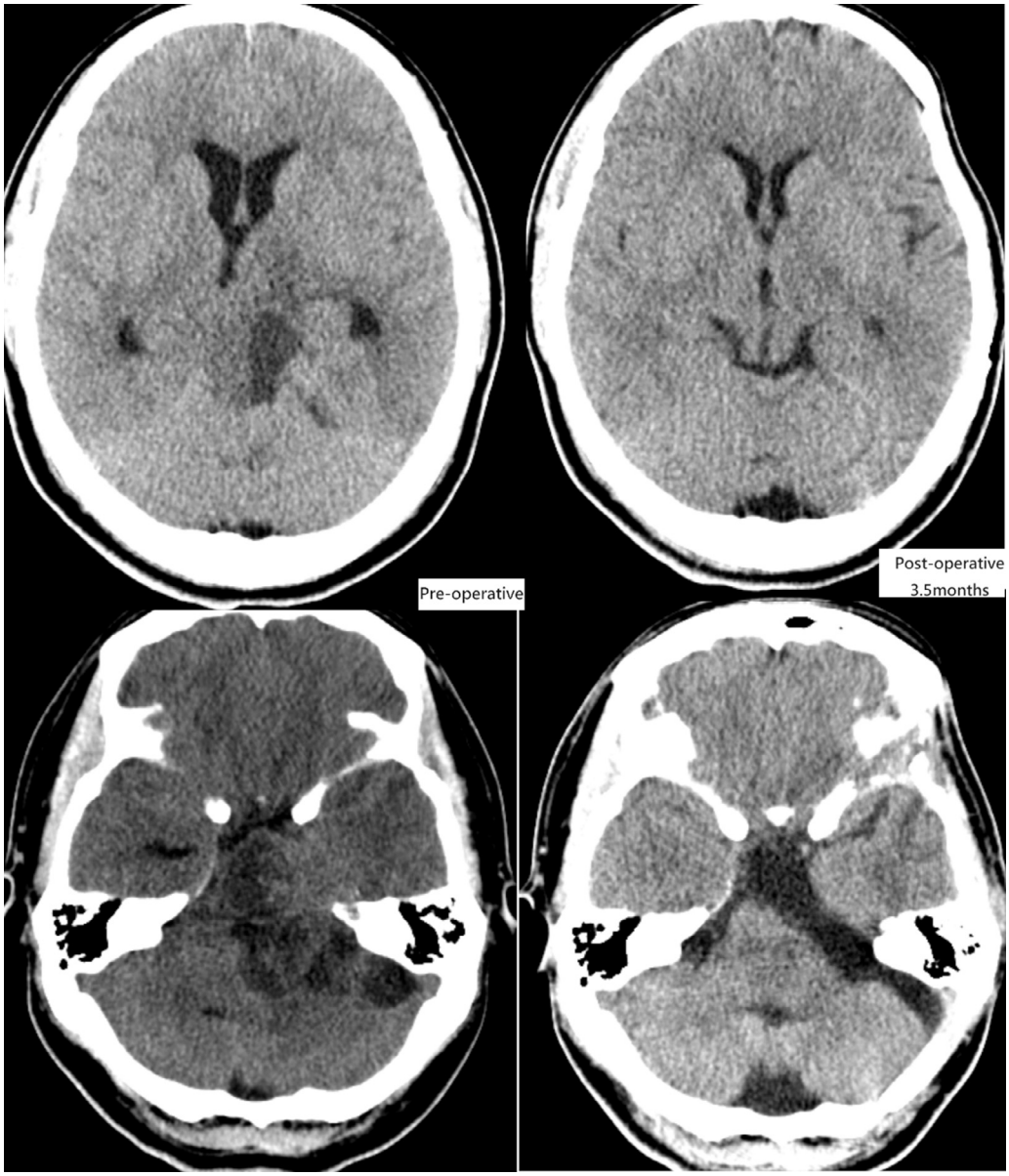

Fig. 4 – Axial noncontrast computed tomography (CT) brain study at preoperative (left column) and postoperative 3.5 months (right column), showing interval resolution of rightward midline shift and hydrocephalus together with resolution of brainstem and cerebellar compression.

achieved with conventional MRI sequences and multi-planar reformatting of isovolumetric data [11]. Additional advanced MRI sequences (e.g., DWI, SWI) are useful to aid lesion characterization and prioritizing differential diagnoses. Dedicated MR neurography can further delineate the anatomical relationship between the lesion from the nerve [12] or nerve related injury [13]. Cranial nerve tractography, like diffusion tensor imaging (DTI), has also been shown to be able to delineate the course of individual nerves within the posterior fossa [14] with a potential role in planning nerve preservation surgery [15]. While computer tomography (CT) has a limited role in soft tissue characterization of intracranial neurogenic tumors, it is useful in providing an initial assessment of tumor complications (e.g., intratumoral hemorrhage) and the associated secondary effects like hydrocephalus and herniation in emergency settings [16]. The presence of well-defined boundaries, displacement rather than infiltration of adjacent structures, and smooth remodeling of skull base foramina [4] can be clues to a non-aggressive and slow-growing disease nature. Effortless identification of calcification is also an advantage of CT study [17].

To evaluate voluminous intracranial tumors with multicompartmental involvement, radiologists should not only focus on the major tumor bulk (i.e., cerebellopontine angle in the current case) but also the complete extent of the lesion. While trigeminal schwannoma is not among the top differential diagnoses of cystic cerebellopontine angle tumors [18], foramen ovale and petrous apex involvement should give clues to the diagnosis. Tumor bulk extending across the porus acousticus without expanding the ipsilateral internal acoustic canal

(Fig. 3C) renders the more common vestibular schwannoma as well as cochlear and facial nerve tumors unlikely. The rare cause of endolymphatic sac tumors is also excluded with a normal nondilated vestibular aqueduct.

Tumor aggressiveness is another aspect that warrants early assessment. While the absence of parenchymal infiltration and perilesional edema in MRI are supporting features, the smooth bone remodeling at the skull base in CT suggests benignity for lesions of this large size. These render malignancies, such as chondrosarcoma and metastases, unlikely in the current case. However, these features may not be applicable to smaller lesions [19].

The mixed cystic solid appearances in conventional MRI sequences essentially exclude more common cystic lesions like arachnoid and epidermoid cysts, as well as petrous apex cholesterol granuloma, from the differential diagnoses.

Petrous apex meningiomas may be difficult to differentiate from neurogenic tumors due to their variable appearances in different types of bone involvement and the degree of tumor invasion [20], though meningioma with cystic components is also a rare disease entity [21]. Features of the dural base soft tissue component, CT hyperdensity and homogeneous enhancement in the solid component and dural tail [22] are usually useful features for diagnosis. In more ambiguous cases, a higher ADC value in large neurogenic tumors may be a distinguishing feature from other cerebellopontine angle tumors including meningiomas, which is related to a larger proportion of loose-textured and cystic areas (i.e., Antoni B tissue) [23,24]. It is also suggested that the presence of non-calcification microbleeds, which is also present in our reported case, as well as a myoinositol peak in magnetic resonance spectroscopy, are features for differentiating schwannoma from meningioma [25].

Plexiform trigeminal neurofibroma may be a close differential diagnosis after establishing the neurogenic tumor nature, although major cystic components are not common imaging feature [26]. The rare differential diagnosis of carotid canal sympathetic plexus schwannoma is also unlikely for the inferolateral rather than supralateral displacement of petrous L ICA [27] and the axis of growth from cerebellopontine angle to foramen ovale in the reported case, despite demonstrating carotid canal smooth expansion.

After establishing the diagnosis, recognizing surgical classifications can improve our understanding of the surgeon's perspective. The most cited classification for trigeminal schwannoma was proposed by Jefferson in 1953 [28], who divided these lesions by location, namely middle fossa lesions (Type A), posterior fossa cranial nerve roots (Type B), dumbbell lesions (Type C), and lesions with extracranial extension (Type D). Since then, multiple other classifications of trigeminal schwannoma have been proposed, with 3 of them included above in the case presentation. Some authors have proposed modifications based on prior classifications. For example, Jeong et al. [29] put further emphasis on the dominant disease components in the middle (Mp) or posterior (Pm) fossa in dumbbell lesions. These classifications are usually coupled with surgical approach recommendations to cater to the anatomical characteristics of different trigeminal schwannomas.

While trigeminal schwannomas may have variable disease extents, the European Association of Neurosurgical Society (EANS) skull base section expert consensus recommends maximal safe resection with preservation of nerve function as the management aim of the disease [30].

Therefore, it is essential for radiologists to accurately localize the disease, establish the diagnosis, and effectively communicate findings to our fellow referring surgeons (with or without familiar classifications) to guide early management, especially in cases with significant mass effects, as illustrated. Interdisciplinary communication can be facilitated when radiologists and clinicians speak common languages, which may potentially improve patient care [31].

Conclusion

Sound anatomy knowledge and high vigilance in the disease extension along cranial nerve together with comprehension of imaging features of differential diagnoses are key to arrive at preoperative diagnosis of the uncommon entity of cystic MTS centered at the cerebellopontine angle, which potentially alters the management approach and improves the patient's outcome. Advanced magnetic resonance imaging techniques may provide further value in tissue characterization and surgical planning.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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