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Case Report

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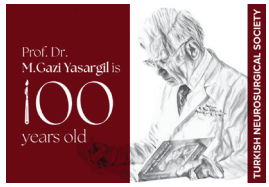

Deceptive Mimics of Trigeminal Schwannoma: Be Careful with Primary Radiosurgery

Sripartha Krishna YERRAMILLI[1], Manjul TRIPATHI[1], Chirag K AHUJA[2], Sandeep MOHINDRA[1], Rajeev CHAUHAN[3]

[1]Post Graduate Institute of Medical Education and Research, Department of Neurosurgery, Chandigarh, India
[2]Post Graduate Institute of Medical Education and Research, Department of Neuroradiology, Chandigarh, India
[3]Post Graduate Institute of Medical Education and Research, Department of Neuroanaesthesia, Chandigarh, India

Corresponding author: Manjul TRIPATHI ✉ drmanjultripathi@gmail.com

ABSTRACT

Trigeminal schwannomas are rare lesions centered on the trigeminal ganglion at Meckel's cave. The complexity and morbidity of surgery for these lesions have allowed stereotactic radiosurgery (SRS) to emerge as a safe and viable option for treatment. Various other lesions at this location must alert one to consider an alternative diagnosis before upfront SRS without histopathological correlation. We present three patients with trigeminal neuropathy with imaging suggesting trigeminal schwannoma. Primary clinicians recommended primary radiosurgery to these patients based on radiological diagnosis. Upon further evaluation and clinical suspicion, we established alternate diagnoses of non-Hodgkin's lymphoma, lepromatous trigeminal nerve involvement, and Aspergillosis involving the Meckel's cave in three cases. Each patient received appropriate treatment instead of SRS. SRS is one of the treatment options for trigeminal schwannomas. No neurosurgical ailment should be treated on its face value with primary SRS, but it must be carefully evaluated on a clinicoradiological profile. Upfront, primary SRS may be counterproductive or detrimental for inflammatory or infectious pathologies, attracting complications.

KEYWORDS: Stereotactic radiosurgery, Gamma knife, Lymphoma, Aspergillosis

INTRODUCTION

Trigeminal schwannomas are rare lesions involving the sheath of the trigeminal root, ganglion, or nerve, comprising 1% of all intracranial tumors and 0.8-8% of all intracranial nerve sheath tumors (16). These lesions are usually T1-hypointense, and T2-hyperintense with heterogeneous enhancement on contrast MRI sequences (11). Usually, trigeminal schwannomas classically show a 'dumbbell' shape when they extend into the posterior fossa or laterally beyond the confines of the Meckel's cave. Trigeminal schwannomas often show cystic areas, with or without fluid levels, suggestive of intralesional hemorrhage (17). These lesions in and around the Meckel's cave lie in a treacherously complex skull base area. The attractive ease of administration and impressive stereotactic radiosurgery (SRS) results have propelled it as a first-line therapy (6,13).

Several case reports have been published describing lesions in Meckel's cave and trigeminal ganglion that range from benign to metastatic. We present three cases referred to our institute for Gamma Knife® radiosurgery (GKRS, Elekta Ltd. Sweden). A careful clinical history, examination, and analysis of radiological appearance enabled us to suspect an alternative pathology.

CASE REPORT

Written informed consents were obtained from the individuals (and/or legal representatives) for the publication of the cases.

Case 1

A 48-year-old male was referred for GKRS with a two-month history of facial pain and numbness. He suffered from progressive diplopia and a diminution of vision in both eyes

Sripartha Krishna YERRAMILLI : 0009-0001-1179-8711
Manjul TRIPATHI : 0000-0002-5679-3439
Chirag K AHUJA : 0000-0003-0734-3252
Sandeep MOHINDRA : 0000-0002-4629-4900
Rajeev CHAUHAN : 0000-0001-6836-647X

for one month. On examination, he had reduced sensation over the right ophthalmic division (V1) of the trigeminal nerve and right abducens nerve palsy. Visual acuity was diminished in both eyes, with bilateral Frenkel grade 5 papilledema on fundus examination. A contrast-enhanced MRI showed an extra-axial, 23 x 20 mm, T1-isointense, T2-hyperintense lesion in the right cavernous sinus, with cisternal and temporal extension and intense, homogenous contrast enhancement. In dedicated T2-weighted thin sections, the tumor extended along the trigeminal nerve up to its exit from the brainstem. The provisional differential diagnosis on radiology was trigeminal schwannoma or spheno-clival meningioma (Figure 1). Inflammatory and autoimmune pathologies were ruled out in CSF and serum studies. He underwent a thecoperitoneal shunt, after which visual acuity improved.

He was further evaluated with DOTANOC – positron emission tomography – computerized tomography (PET-CT), which showed a 2.9 x 1.6 mm lesion (SUV max 16.7) in the right cavernous sinus, along with tracer-avid masses in the left pterygopalatine fossa and left sixth and seventh ribs. PET-CT-guided fine-needle aspiration cytology (FNAC) from the rib lesion suggested non-Hodgkin's lymphoma (NHL). Unfortunately, due to personal reasons, he discontinued chemotherapy and succumbed to disseminated disease after three months.

Case 2

A 32-year-old male was referred for GKRS with a history of facial pain over maxillary (V2) and mandibular (V3) division distribution of trigeminal nerve with difficulty in chewing for two months. On examination, he had decreased sensation over the V2 distribution. An erythematous plaque was also noted over the left side of the face, with sharply demarcated margins, limited only to the V2 distribution (Figure 2). Contrast-enhanced MRI showed a 12 x 8 mm, extra-axial, T1-isointense,

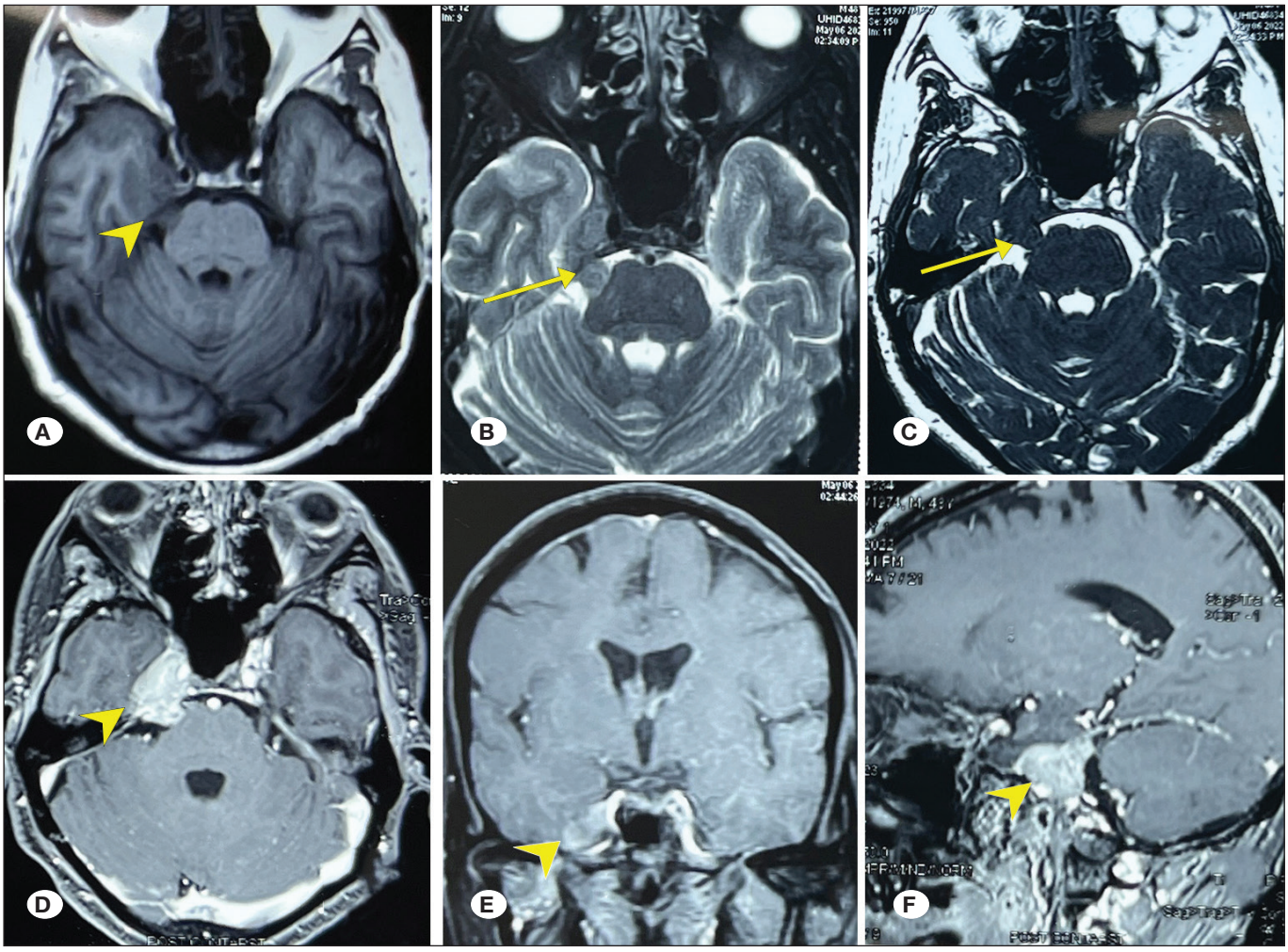

Figure 1: Magnetic resonance imaging of a 48-year-old male patient with facial pain. **A)** Axial T1 weighted imaging shows an isointense lesion arising from the right Meckel's cave, extending along the trigeminal nerve proximally (arrowhead). **B)** The lesion in hyperintense on T2 weighted axial imaging (arrow) and **(C)** its extent from the Meckel's cave, along the nerve (arrow) is seen clearly in axial constructed inference in steady state (CISS) imaging. Contrast-enhanced T1 weighted imaging clearly defines the 'dumbell' shaped tumor (arrowheads) on axial **(D)**, coronal **(E)**, and sagittal views **(F)**.

T2-hyperintense lesion in the left Meckel's cave, with intense, homogenous contrast enhancement.

A skin biopsy from the face showed the presence of multibacillary *Mycobacterium leprae*. He received multidrug therapy for multibacillary leprosy (MDT MBR) and Carbamazepine for facial pain. On follow-up, his facial pain was under control with oral medications. The erythematous plaque also resolved almost entirely 18 months after the initiation of therapy (Figure 2E, F). A repeat imaging showed complete resolution of the lesion (Figure 2C, D).

Case 3

A 25-year immunocompetent married female presented with a ten-day duration of fever, headache, and right-sided facial pain with facial deviation. On examination, she had reduced sensation on the right side of her face over V2 and V3 distribution, along with right-sided facial palsy. Fundus examination showed mild papilledema. A contrast-enhanced MRI revealed a large, well-defined, dumbbell-shaped, extra-axial lesion extending from the right tentorium and Meckel's cave. The lesion was T1-isointense, T2 hypointense, with peripheral contrast enhancement and areas of central non-enhancement, along with a broad attachment over the tentorium (Figure 3). The provisional reported diagnosis was trigeminal schwannoma with hydrocephalus. However, we suspected any infection or inflammatory pathology because of rapid clinical deterioration and the recent onset of fever. Her CSF analysis and hematological evaluation were unremarkable except for slightly raised total leucocyte counts.

Repeat imaging showed an enlarging lesion. An unexpected increase in the size of the tumor pointed against the natural history of TS. The lesion was greyish, firm in consistency, and non-suckable during surgery, with no definitive plane from the adjacent parenchyma. Aseptate hyphae was present on the potassium hydroxide (KOH) smear. Histopathological analysis revealed the lesion to be consistent with Aspergillosis with *Aspergillus flavum* species identified on tissue culture. The patient unfortunately had a stormy postoperative course with postoperative edema requiring prolonged ventilation, multiple seizures, and vasculitic infarcts. She was discharged to a rehabilitative care center after a prolonged hospital stay. However, she succumbed there due to pneumonia after two weeks.

DISCUSSION

Trigeminal schwannoma and other lesions abutting the cavernous sinus are commonly referred to as primary SRS. These lesions are often advised upfront radiation without a histopathological diagnosis. Our series demonstrates three

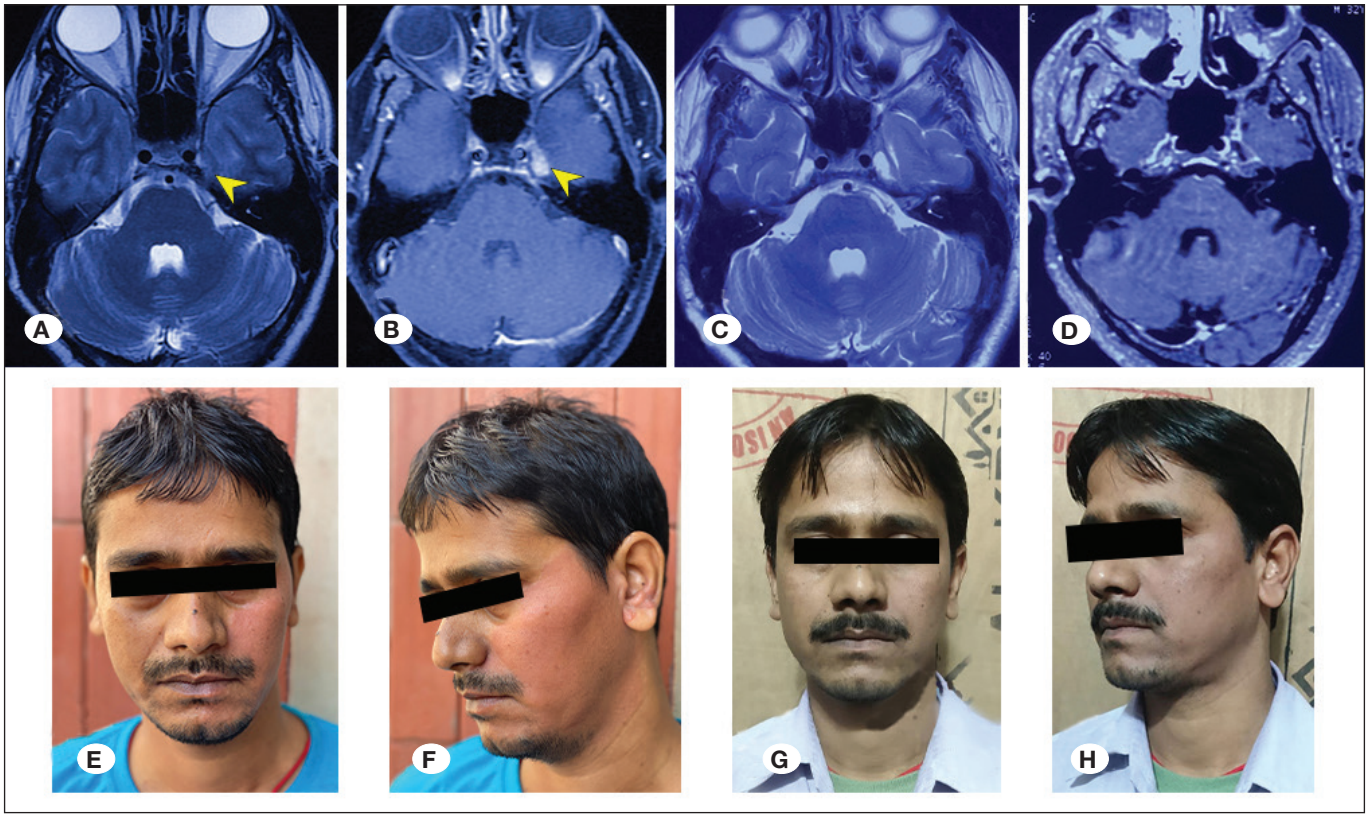

Figure 2: Radiological and clinical images of a 32-year-old male with facial pain. **A)** T2 weighted axial magnetic resonance imaging (MRI) shows a hyperintense, regular lesion (arrowhead) involving the trigeminal ganglion in the Meckel's cave, **(B)** with homogenous gadolinium uptake (arrowhead). MRI done one year after initiation of multidrug therapy (MDT) shows complete resolution on T2 weighted imaging **(C)**, with no residual contrast uptake **(D)**. There is a well-defined erythematous plaque over the skin distribution of the left ophthalmic division **(E, F)**, with complete resolution after 1 year of MDT **(G, H)**.

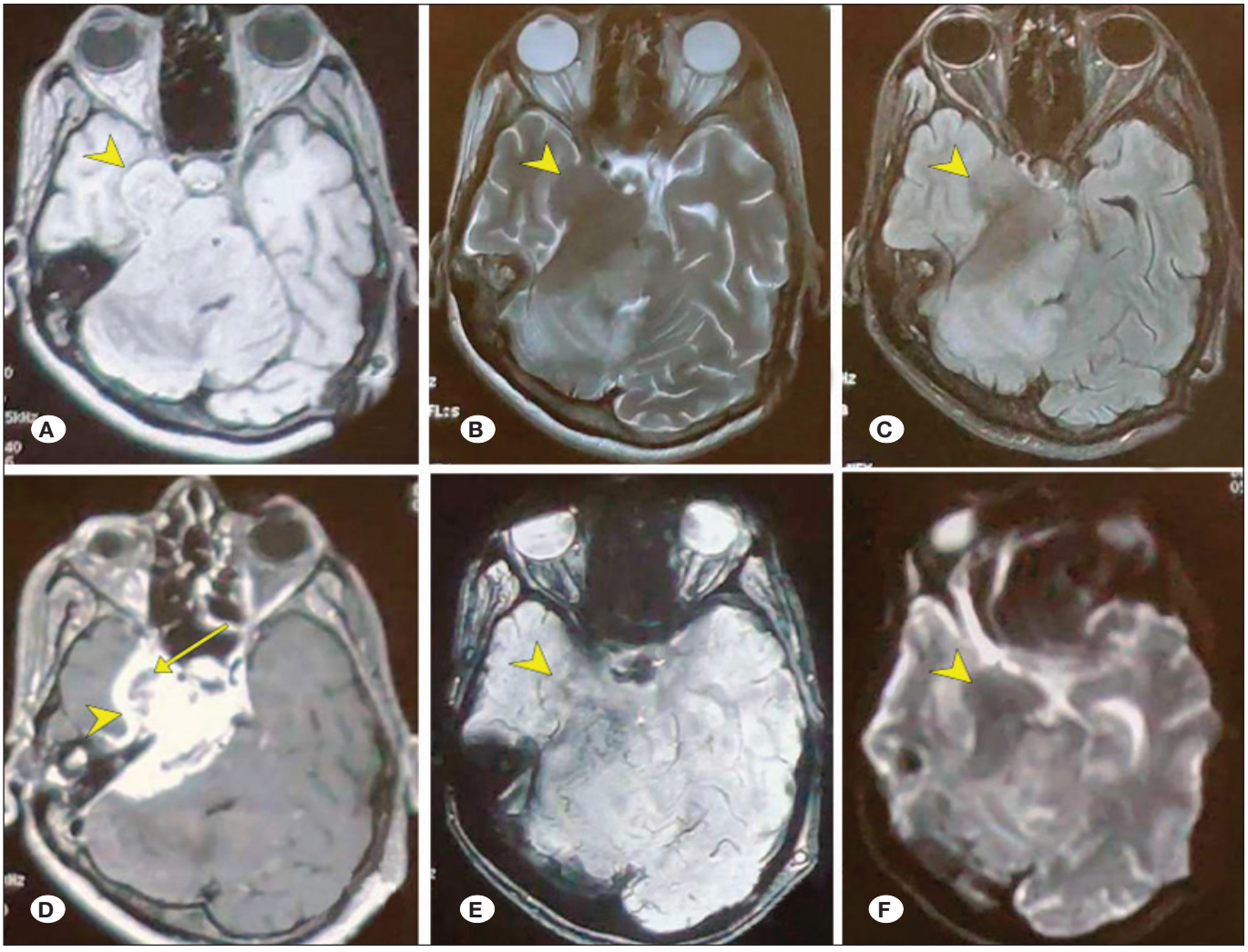

Figure 3: Magnetic resonance imaging of a 25-year-old female with facial pain. **A)** T1 weighted imaging shows an isointense lesion (arrow) along the middle cranial fossa, extending into the posterior fossa, which is hypointense on T2 weighted axial imaging **(B)** and FLAIR axial imaging **(C)**, with perilesional edema. The lesion shows strong contrast enhancement (arrowhead) with areas of non-enhancement in its center (arrow) **(D)**. Susceptibility-weighted axial imaging **(E)** and diffusion-weighted axial imaging **(F)** show the lesion to be relatively avascular with no diffusion restriction.

patients with trigeminal neuropathy, referred for primary SRS. The diagnosis of trigeminal schwannoma was doubtful, given the clinicoradiological mismatch.

Our first case, a 48-year-old male, had facial pain and a contrast-enhancing lesion on MRI. Bilateral optic nerve involvement with bilateral papilledema and abducens palsy raised the suspicion of any alternate pathology. Twelve cases of primary lymphomas affecting the trigeminal nerve have been reported in the literature (7). A majority of these patients presented with facial pain (10 of 12), and their ages ranged from 29-77 years. Akaza et al. have described a case demonstrating the role of SRS in a 60-year male diagnosed with trigeminal schwannoma based on a clinicoradiological picture alone. Though improvement of symptoms was observed initially he had a relapse of symptoms after 11 months, with multiple periventricular lesions on repeat imaging (1).

Thickening of the peripheral nerves is a well-known phenomenon in leprosy (8). Trigeminal and facial nerve involvement is relatively common amongst cranial nerves. These usually present as lesions at the level of respective nuclei in the brainstem and contrast enhancement of the nerve (12). A demarcated facial hyperpigmentation and hypoesthesia restricted to V2 distribution in the second case raised the suspicion of leprosy.

Falcioni et al. (5), Lao and Tsai (10), and Chang et al. (4) have described 'dumbbell' shaped tumors involving the Meckel's cave. Though this appearance is classical to trigeminal schwannoma (11), all three cases had an alternative histopathological diagnosis. Our third case had a lesion confined to the prepontine cistern with an extension into the cavernous sinus, giving a dumbbell-shaped appearance. However, hypo intensity on the T2 weighted scan and acuteness of the clini-

cal course prompted us towards a possible fungal pathology. In most clinical cases in our practice, Aspergillosis presents with predominant rhino-cerebral involvement. In this case, the paranasal air sinuses were clear of such pathology.

Some authors have reported sarcoidosis involving the trigeminal nerve, either as an isolated involvement (2,14), or as a secondary manifestation of systemic sarcoidosis (3). The clinical presentation and imaging of trigeminal neurosarcoidosis closely mimic trigeminal schwannoma, leading to surgical excision of these lesions, often an avoidable intervention. These masses respond well to immunosuppressive therapy alone.

Raswoli et al. describe a case of a 67-year-old woman misdiagnosed with trigeminal schwannoma who underwent SRS and later succumbed to the progression of the malignant lesion (15). This highlights the need to consider the pathological diagnosis of any trigeminal schwannoma, which does not follow a favorable clinical course, especially following SRS.

Reports of more vicious entities masquerading as trigeminal schwannoma have been described in the literature, including a case of grade 4 diffuse astrocytic glioma arising from pons, with perineural spread, giving the characteristic radiological appearance of trigeminal schwannoma (15). Metastases to Meckel's cave have been described in about 20 patients across literature from various sources, including cutaneous squamous cell carcinoma (15,18), small cell lung cancer (19), renal cell carcinoma (20), follicular thyroid cancer (9), breast cancer and colon cancer.

CONCLUSION

Through our series of three cases and a brief literature review, we have tried to highlight various lesions commonly misdiagnosed as trigeminal schwannoma and referred for SRS. Compared to previous generations, there is diminishing stress on clinical findings and dwindling interest among clinicians in detailed clinical examination. This article highlights the possible complications with primary radiosurgery when the clinicoradiological correlation is not strong.

Uncommon clinical features, such as cranial nerve deficits beyond the usual confines of the Meckel's cave and cavernous sinus, and a rapid progression of symptoms or unusual clinical course, must always alert the surgeon about the possibility of an alternative diagnosis.

Declarations

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Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

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AUTHORSHIP CONTRIBUTION

Study conception and design: MT, CA
Data collection: SKY, RC
Analysis and interpretation of results: MT, SM
Draft manuscript preparation: SKY, MT, CA
Critical revision of the article: SM, MT
Other (study supervision, fundings, materials, etc...): RC
All authors (SKY, MT, CA, SM, RC) reviewed the results and approved the final version of the manuscript.

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