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Case Report

Case report: Primary ependymoma of the trigeminal nerve presenting as trigeminal neuralgia

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ABSTRACT

Background: Ependymomas are usually found in the posterior fossa originating from the fourth ventricle. Primary ependymomas arising from cranial nerves are rare with only a handful of reported cases. Trigeminal neuralgia (TN) is rarely due to space occupying lesions.

Case Description: A 20-year-old female presented with TN with a rare presentation of a pure extra-axial ependymoma involving the right trigeminal nerve in the cerebellopontine angle.

Conclusion: It is essential to explore the possibility of a mass arising from the trigeminal nerve when investigating the cause of TN.

Keywords: Ependymoma, Extra-axial ependymoma, Trigeminal neuralgia

INTRODUCTION

Trigeminal neuralgia (TN) is a disorder characterized by sudden, brief, and recurrent attacks of severe facial pain along the distribution of one or more branches of the trigeminal nerve.[14] It is caused by the demyelination of trigeminal nerve within either the nerve root, or less commonly, the brainstem.[10] Vascular compression of the trigeminal nerve root is associated with TN in about 95% of patients, whereas the remaining cases are associated with multiple sclerosis plaques, lacunar infarctions within the brain stem trigeminal system or rarely, cerebellopontine mass lesions.[17]

Ependymomas are uncommon tumors of neuroectodermal origin and are mostly found infratentorially (60%).[13] In adults, ependymomas are mostly found in the spine, whereas intracranial ependymomas account for only 4% of adult brain tumors.[7] Extra-axial ependymomas are extremely rare. Herein, we present the case of a 20-year-old female presenting with the right-sided TN with a World Health Organization (WHO) Grade 2 ependymoma involving the right trigeminal nerve in the cerebellopontine angle (CPA) before it entered Meckel's cave.

CASE DESCRIPTION

A 20-year-old right-handed female presented with a 5-month history of progressively worsening, lancinating right-sided facial pain that was unresponsive to medical management. The pain

occurred episodically and lasted for few minutes after which it would resolve. The pain was exacerbated by talking, chewing, brushing her teeth, blowing winds, and cold breezes. She reported no history of headache, watery eyes or redness of eyes, rhinorrhea, or tooth ache. The rest of the systemic history was unremarkable, and the patient otherwise had no significant medical or surgical history.

On the initial examination, the patient sat comfortably with no signs of distress. She was vitally stable, and the general examination was unremarkable. The neurological examination of the cranial nerves was significant for decreased sensation on the right in the ophthalmic (V1) and maxillary (V2) division of the trigeminal nerve. She also had House Brackmann Grade 1 facial weakness on the right side. The rest of the neurological and systemic examination was unremarkable.

A magnetic resonance imaging (MRI) scan of the brain revealed a well-circumscribed 12 × 9 mm cystic lesion in the right CPA at the site of the trigeminal nerve. The cyst returned low signals on T1WI and high signals on T2WI with no contrast enhancement and caused minimal displacement of the trigeminal nerve. Subtle relative atrophy of the visualized right trigeminal nerve was also noted. Due to the cyst, the trigeminal nerve was not completely visualized on the MRI scan. Figure 1a-f represent the MR images signifying an abnormal signal intensity lesion at the pre-Meckel cave segment of the right trigeminal nerve, which is iso- to hypointense on T1-WI and hyperintense on T2-WI with no post contrast enhancement or diffusion restriction.

The patient underwent right retrosigmoid craniotomy and exploration of the CPA. Once the thin-walled cyst was punctured (revealing clear fluid), the trigeminal nerve became more prominent, and a localized small, grayish, and soft-tissue lesion became visible on the superior surface of the nerve before it entered Meckel's cave. The mass extended over the length of the nerve and measured roughly 0.6 cm × 0.3 cm. The lesion was dissected off the nerve, and the case was completed uneventfully. The patient reported immediate improvement in the symptoms of TN and was discharged the next day. Facial nerve function also showed improvement at follow-up 2 weeks later.

Histopathology sections of the lesion revealed tumor cells arranged predominantly around central blood vessels forming perivascular pseudorosettes [Figure 2]. The neoplastic cells were rounded showing moderate eosinophilic cytoplasm with round to oval nuclei showing fine granular chromatin with inconspicuous nucleoli. Few of the nuclei exhibited intranuclear inclusions. Scattered calcifications were also present [Figure 3]. There was no significant mitotic activity, microvascular proliferation, or necrosis. Immunohistochemical stains were positive for glial fibrillary acidic protein, whereas Olig2 immunostain was negative [Figure 4a and b]. Hence, morphological and immunohistochemical features favored the diagnosis of Grade II ependymoma according to the WHO classification.

The patient underwent a postoperative neuroaxis MRI scan that revealed no residual lesion and no lesion anywhere else in the neuroaxis. She is currently asymptomatic and on close radiological surveillance.

DISCUSSION

TN is divided into either classical TN that encompasses idiopathic TN and TN caused by neuromuscular compression of the root of the trigeminal nerve; or secondary TN (STN)

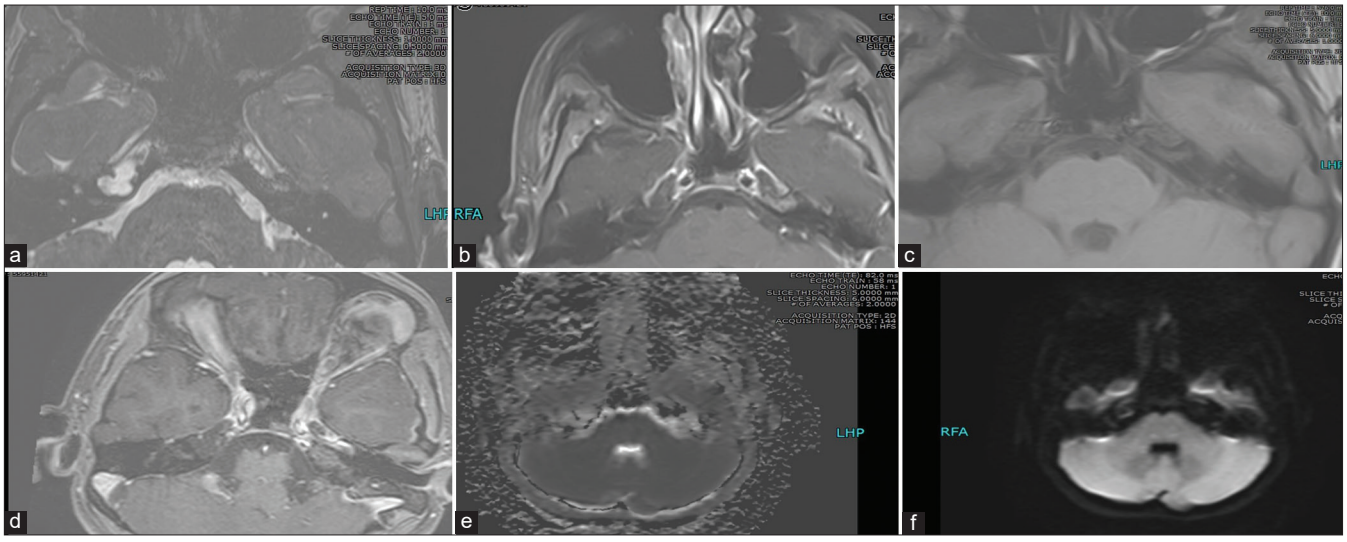

Figure 1: T2 (a), Post Contrast T1 (b), T1 (c), Post Contrast T1 (d), DW (e), ADC (f) images show an abnormal signal intensity lesion which is noted at the pre-Meckel cave segment of the right trigeminal nerve. It is iso to hypo intense on T1-WI and hyperintense on T2-WI with no post contrast enhancement or diffusion restriction.

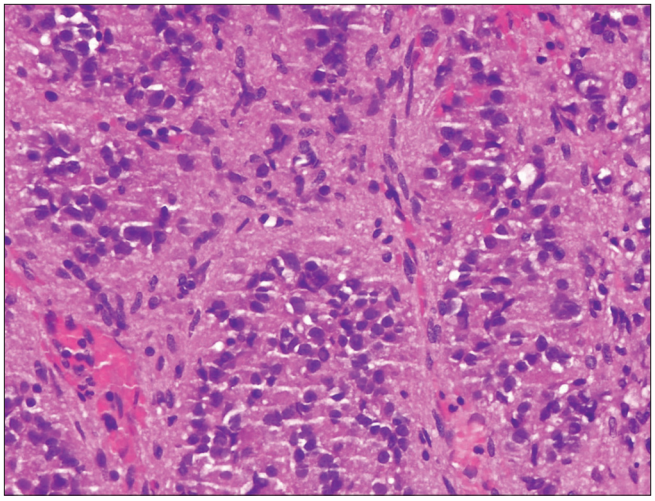

Figure 2: Image shows monomorphic tumor cells with round to oval nuclei forming perivascular pseudorosettes. There is no high cellularity, high mitotic activity, nor microvascular proliferation.

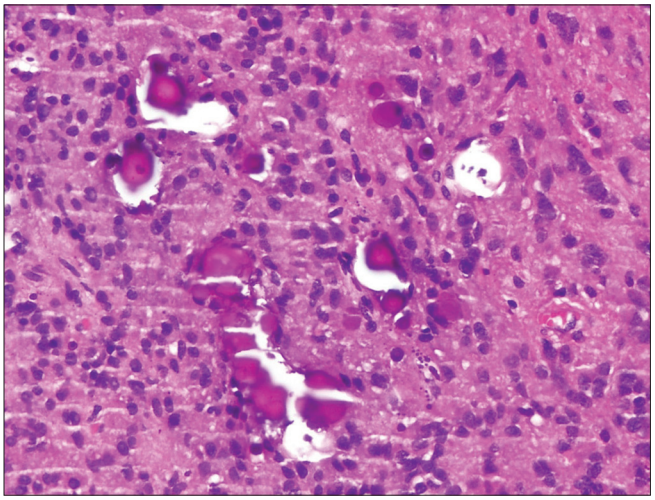

Figure 3: Calcifications are noted in this image. The image also shows monomorphic tumor cells with round to oval nuclei forming perivascular pseudorosettes. There is no high cellularity, high mitotic activity, nor microvascular proliferation.

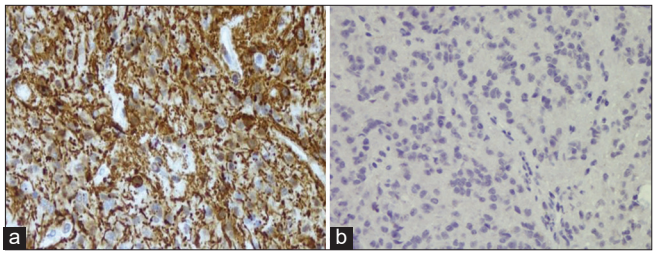

Figure 4: (a,b) GFAP immunostain shows positive staining in tumor cells with stronger expression in perivascular tumor cells. Olig2 immunostain is negative.

caused by multiple sclerosis or a space occupying lesion such as a tumor, cerebral aneurysm, or a megadolicho basilar artery.[11] The authors present a case of STN with a biopsy proven WHO Grade 2 ependymoma. Ependymomas are rare tumors of neuroectodermal origin arising from ependymal cells of the ventricular system, choroid plexus, filum terminale, and central canal of the spinal cord.[1] These rare tumors account for a mere 3–5% of adult intracranial neoplasms.[2,12] About 60–70% of the intracranial ependymomas in the cranium are localized in the posterior fossa and the majority of those originate from the fourth ventricle.[6] Ependymomas can present as intra-axial or intra-axial-extra-axial lesions, while a complete extra-axial intracranial ependymoma presentation is rare.[15] Berhili *et al.* reported in 2017 that there were only 14 adult cases in the English literature of intracranial extra-axial ependymomas.[3] Although ependymomas may extend into the CPA or subarachnoid space, a primary extra-axial intracranial ependymoma in the CPA is extremely rare.[8] Ebrahimi *et al.* highlighted the importance of considering ependymomas as a differential diagnosis among CPA tumors as six adult cases of CPA ependymomas had been reported in the English literature by 2020.[5] Rare cases of CPA ependymomas extending into unusual locations such as into the cavernous sinus and pineal region have been reported.[4,16] In one case, Torun *et al.* reported the tumor extending from the CPA into Meckel's cave.[15] In another case, Zhao *et al.* reported the first case of bilateral primary posterior fossa ependymomas originating from the CPA and extending to the internal auditory canals, with spinal cord metastasis.[18] To the best of our knowledge, there are only two previous case reports similar to ours, wherein the origin of the tumor is from the sheath of the trigeminal nerve.[9,15]

CONCLUSION

TN is a relatively common disorder with possible rare differentials which include infratentorial ependymomas. To the best of our knowledge, only two cases have been previously reported, in which the tumor has originated from the sheath of the trigeminal nerve. We have reported one more case of a pure extra-axial ependymoma arising from the sheath of the trigeminal nerve resulting in TN.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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