

Giant Multi-Compartmental Trigeminal Neurinoma in a Child: A Case Report

Ishwar Singh* Vinit Bhai and Nisha Marwah

Department of Neurosurgery, Pt. B.D. Sharma University of Health Sciences, Rohtak, India

Abstract: We report a case of solitary giant multi-compartmental trigeminal neurinoma in a 10 year old female child, without the stigmata of neurofibromatosis. The child presented with headache, vomiting, dysarthria, diplopia and altered sensorium. Neuroimaging revealed a giant multi-compartmental trigeminal neurinoma measuring 6.8 x 4.3 x 4.0 cm in size. The tumor was successfully removed by single stage petrosal approach. It is extremely rare for a solitary giant neurinoma to occur at this site in a child without neurofibromatosis. Advantages of petrosal approach in such type of tumor are discussed. As per author's knowledge, this is the largest trigeminal neurinoma reported till date in a child.

Keywords: Child, petrosal approach, trigeminal neurinoma.

INTRODUCTION

Neurinoma^s arising from trigeminal nerve are rare tumors, though they are second most common after acoustic neurinomas [1]. Their incidence ranges from 0.07% to 0.36% among all intra-cranial tumors and 0.8% to 8% of all intracranial neurinomas [2]. They are commonly seen in the 4th and 5th decades. However a solitary neurinoma at this site in a child without neurofibromatosis is a rare occurrence [2-4]. Only a few case reports of small sized solitary neurinomas in children without neurofibromatosis are reported in the literature [2-5]. We report a rare case of a giant multicompartamental trigeminal neurinoma without neurofibromatosis in a child that was completely excised by single stage petrosal approach, with good neurological recovery.

CASE REPORT

A 10 year old female child, presented with headache associated with vomiting for the last 3 months. She gradually developed numbness of left side of face with dysarthria, ataxia and diplopia. She became drowsy 3 days prior to hospitalization. On examination there was complete sensory and motor paralysis of the 5th cranial nerve with partial involvement of 3rd, 4th and 6th to 11th cranial nerves of the left side. Bilateral papilloedema, bilateral long tract signs and left sided cerebellar signs were also present. She did not have any cutaneous manifestation of neurofibromatosis such as café au lait spots or subcutaneous nodules. There was no family history suggestive of familial neurofibromatosis. Neuroimaging revealed markedly enhancing extra-axial mass of size (6.8x4.3x4.0cm) involving left parasellar area, petrous apex, cerebello-pontine and cerebello-medullary angles, with obstructive hydrocephalus, suggestive of trigeminal neurinoma (Fig. 1).

Urgent right sided VP shunt was done to relieve the features of hydrocephalus. The patient's sensorium improved

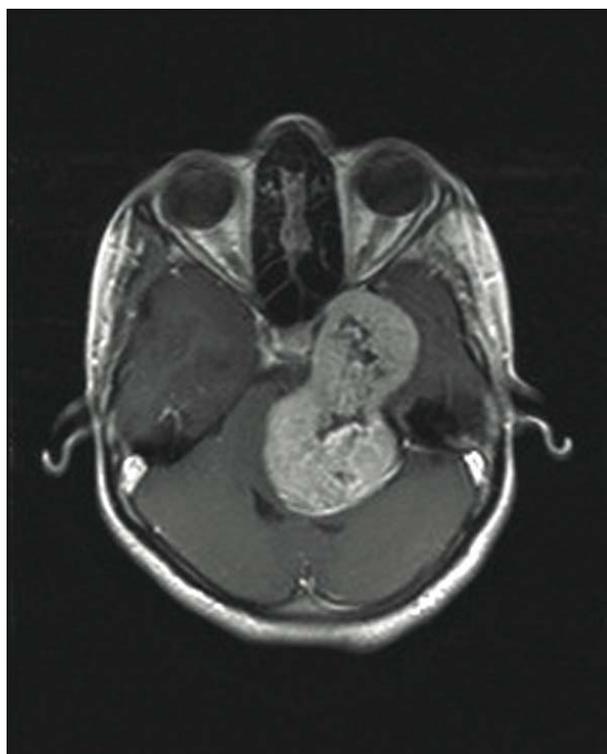


Fig. (1). Gad enhanced MRI revealing giant multi-compartmental Trigeminal neurinoma, axial section.

after the shunt surgery. Later on the trigeminal neurinoma was completely excised *via* the petrosal approach [6]. The tumor was moderately vascular and soft to firm in consistency. It was gradually debulked using bipolar cautery, suction and curette till the interface between the capsule and the surrounding neurovascular structures was identified. Capsule was then gradually separated from the brain stem and the cranial nerves. Lastly, the tumor was removed from the roof and the lateral wall of the cavernous sinus to ensure complete removal. Postoperative MRI confirmed complete removal of tumors (Fig. 2). Histopathological evaluation of the

*Address correspondence to these authors at the Department of Neurosurgery, Pt. B.D. Sharma University of Health Sciences, Rohtak, India; Tel: 01262-274200; Fax: 01262-211308; E-mail: drishwarsingh@yahoo.co.in

lesion revealed a spindle cell neoplasm with cells arranged in compact fascicles (Antoni A areas) and intervening loosely arranged cells (Antoni B areas). Cellular palisading with formation of Verroca bodies was evident at places. The tumor was richly vascular with focal perivascular hyalinisation (Fig. 3). Immunohistochemically, the tumor was positive for S-100 protein confirmed the diagnosis of Schwannoma. Patient showed complete neurological recovery at 6 month's follow-up except for persistent 50% sensory loss in V2 component of 5th cranial nerve.

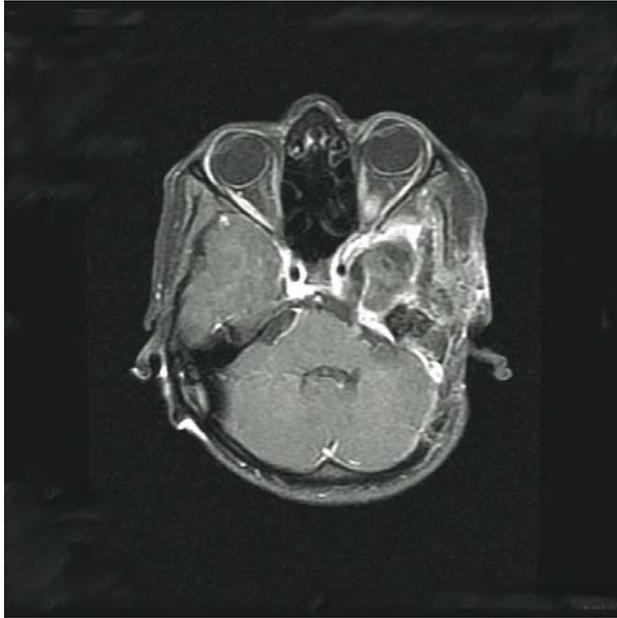


Fig. (2). Post op Gad enhanced MRI revealing complete excision, axial section.

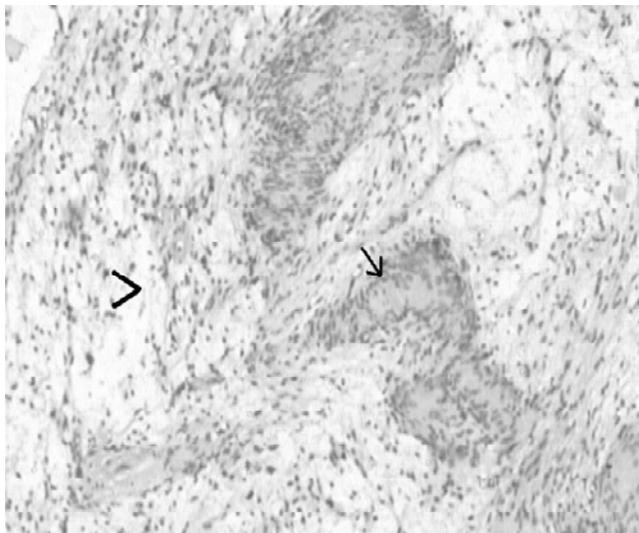


Fig. (3). Microphotograph showing neurinoma with Antoni A (arrow) and Antoni B areas (arrowhead). (H & E x 100x).

DISCUSSION

Trigeminal neurinoma is an uncommon tumor and is usually seen in association with multiple intra-cranial neuro-

nomas as a manifestation of neurofibromatosis [2]. It is mostly seen between 4th and 5th decades of life with slightly female preponderance [2]. The sensory symptoms in distribution of the trigeminal nerve are the commonest presentation [1, 2, 7]. Other signs and symptoms are produced as per size and location of the tumor. Trigeminal neurinoma is classified into six types according to the tumor location [2, 8]. Type M, P and E represent middle fossa, posterior fossa and extra cranial space respectively; type MP (middle fossa and posterior fossa), ME (middle fossa and extra cranial space) and MPE (middle fossa, posterior fossa and extra cranial space) are tumors involving multiple compartments. Surgical approach to these tumors depends upon their anatomical location. Hence accurate pre-operative delineation of the tumor mass is essential. Multi-compartmental tumors are removed by different combined approaches [7, 9, 10]. The tumor that straddle the middle and posterior cranial fossae and do not extend below the internal auditory canal can be removed through a middle fossa approach [2, 11]. The combined approach is employed for large dumbbell shaped tumors that extend ventral to the lower brainstem and below the internal acoustic meatus [2, 11]. Various reports have stressed the need for radical surgery so as to achieve higher cure rates. Moreover the recurrence rate with partial resection is reported to be higher in trigeminal neurinoma than acoustic neurinoma [10]. Major impediment to complete removal is inadequate exposure due to deep location of the tumor and close proximity to vital neuro-vascular structures. The ideal surgical approach should be shortest and direct as well as wide and low so as to avoid the need for prolonged and excessive brain retraction. Verstappen *et al.* [11] reported a case of trigeminal neurinoma of maximum diameter of 53mm, centered over Meckel's cave in a 6 year old girl removed by single stage pterional approach. However with pterional approach it is difficult to remove large posterior fossa component as was in our case. Moreover pterional approach requires a lot of temporal lobe retraction leading to increased postoperative morbidity. In our patient, a giant multi compartmental trigeminal neurinoma with a large posterior fossa component was completely removed in a single stage using petrosal approach. This combined approach has following advantages:-

- 1) Cerebellar and temporal lobes are retracted minimally
- 2) Operative distance to the tumor is less
- 3) Surgeon not only has direct line of sight to the lesion, but also of the anterior and lateral aspect of the brainstem
- 4) Neural and the otological structure including the facial nerve, cochlea and labyrinth are preserved
- 5) The transverse and sigmoid sinus along with the vein of labbe and basal veins are preserved
- 6) Tumor supply is interrupted early in the procedure
- 7) Multiple axes of dissection are available.

Injury to the trigeminal nerve is universal and nearly always inevitable, but nontraumatic microsurgical dissection of fibers that are not incorporated in the tumor might even preserve or improve trigeminal nerve function [9, 11]. Our patient had complete trigeminal nerve paralysis which was improved except for 50% sensory loss in v2 component.

CONCLUSION

Trigeminal neurinoma is very rarely reported in children. Although several multi-staged strategies have been reported in the past, the large tumor in our case was completely excised by single stage petrosal approach, with good neurological recovery.

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