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Case Report

Radiology Section

Trigeminal Nerve Choristoma- A Rare Case Report

SHIBU GARG[1], CS PRABHU[2], LOKESH KUMAR[3]

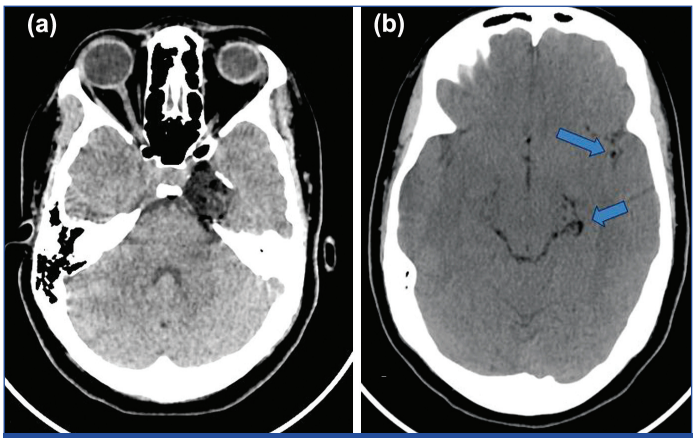
ABSTRACT

Choristomas are normal tissue in an abnormal location and are classified based on their predominant epithelial lining. They may contain fibrous tissue, skeletal muscle fibres, and nerve fibres. Neuromuscular choristoma (benign triton tumour) is a rare tumour that involves a large nerve trunk. Involvement of the cranial nerves by this lesion is exceptional; trigeminal nerve involvement is even rarer. This is a very rare case of intracranial trigeminal nerve neuromuscular choristoma which had an associated fatty component that also got ruptured, in a 30-year-old female who presented with a headache. The clinical presentation, radiological findings, pathological diagnosis for this case are discussed along with a review of the associated literature.

Keywords: Intracranial choristoma, Neuromuscular choristoma, Triton tumour

CASE REPORT

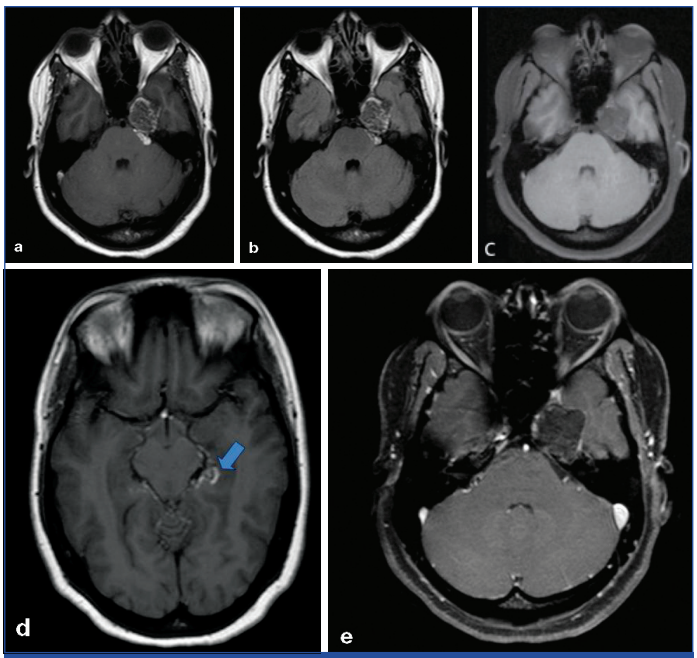
A 30-year-old female presented with worsening headache and facial numbness (left side of the face) for 2 weeks. She had a history of dull aching and progressive chronic headache for one year for which she took symptomatic treatment. There was no other significant history. On examination, her vitals were stable. Visual acuity was normal. Glasgow Coma Scale (GSC) was 15/15, with no signs of meningeal irritation and normal neurological examination except for the facial numbness. The patient was referred to the Radiology Department for imaging work-up. Non contrast Computed Tomography (NCCT) brain was done [Table/Fig-1].



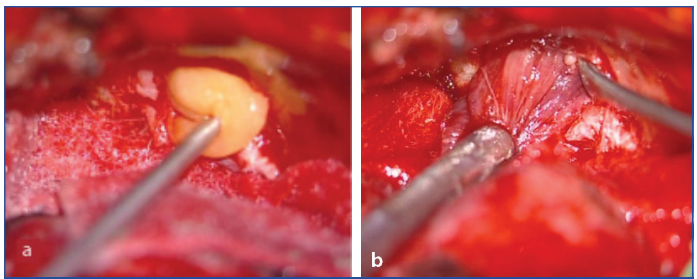
[Table/Fig-1]: (a) Axial CT scan shows a heterogeneous hypodense lesion with predominant fat density. It is centered in Meckel's cave. (b) Multiple fat density globules were noted in the subarachnoid spaces in the basal cisterns and sulci (blue arrows in b).

The MRI Brain with contrast was done [Table/Fig-2]. MRI suggested a provisional diagnosis of ruptured dermoid and trigeminal schwannoma. The fat density globules favoured a ruptured dermoid. The typical location and the lack of intense enhancement favoured a schwannoma. She underwent left fronto-temporo-orbito-zygomatic craniectomy with incision of fronto-temporal dural band. The lesion was excised using an extradural approach. Anterior, posterior, and lateral walls of the tumour were well defined in the posterior aspect of the left cavernous sinus. Tumour had a fatty component that was yellowish and muscular components which were brownish. The tumour was not vascular. However, the entire lesion could not be resected from the nerve. The resected part was sent for histopathological analysis [Table/Fig-3]. The patient tolerated the procedure well.

Histopathological sections of the fragments of the lesion showed mature skeletal muscle tissue (disorganised) and mature adipose tissue, with absent epithelial components, suggesting a diagnosis of choristoma. A final diagnosis of left trigeminal neuromuscular choristoma was established.



[Table/Fig-2]: MRI plain and contrast in axial sections. T1/T2 FLAIR shows a heterogeneously hypointense lesion in the left Meckel's cave with a peripheral rim of hyperintensity and a focal hyperintense area (a and b) that is suppressed on fat saturated sequence (c), representing the fatty component. However, the macroscopic fatty component seen on the CT image could not be identified on MR images. The cranial section shows fat density globules in the ambient cistern (blue arrow in d). The lesion is extending posteriorly into the left prepontine cistern. However, the trigeminal nerve could not be visualised separately on heavily T2W TSE. There is no evidence of enhancement (e).



[Table/Fig-3]: (a) Intraoperative images showing fatty components and (b) the muscular strands.

DISCUSSION

Neuromuscular choristomas are mesenchymal tissue that is incorporated in nerve sheaths during embryogenesis [1]. It can present in different age groups (predominantly in younger patients), with a 3:1 female to male predisposition [2,3]. It is a rare benign tumour composed of skeletal muscle fibres and neural elements [4,5]. It can involve cranial nerves and peripheral nerves [6,7].

In 1972, Louhimo I and Rapola J first reported these tumours as an intraneural muscular hamartoma. Hamartoma, however is abnormal tissue in a normal location [8]. Thus, in 1983 Bonneau R and Brochu P gave the term "neuromuscular choristoma" because the muscle fibres are not normal constituents of peripheral nerves [9]. They have been reported to regress, remain stable, or recur [10]. Symptoms depend upon the site and extent of involvement. The lesion was arising from the trigeminal nerve in this case and the signs and symptoms were attributed to the dysfunction of the nerve.

Similar cases in the literature showed that patients showed features that varied from migraine-type headaches, impaired sensation, numbness, and difficulty in opening the mouth [2,7]. On imaging, the lesion was predominantly hypointense with minimal hyperintense areas which were suppressed on T1 Fat saturated sequence [Table/Fig-2c] and partially suppressed on FLAIR sequence. There was no significant postcontrast enhancement [Table/Fig-2e]. Earlier reported cases have shown few varied appearances like hypointense appearance on T2 and mild homogeneous enhancement [1,2]. The appearance depends on the predominant cellular component present. To the best of our knowledge, an associated fatty component in addition to the muscular elements has not been reported previously. The choice of treatment between conservative surgical management or a complete resection is debatable.

Tumours of the trigeminal nerve are rare [2]. Only a few cases of trigeminal choristomas have been reported in the literature [Table/Fig-4] [1,2,7,11,12]. The common differentials of a lesion in the Meckel's cave are enlisted [Table/Fig-5] [7,13,14].

Studies	Year	Age and Gender	Symptoms	CT/MRI	Histopathology
Zwick DL et al., [12]	1989	29 months/ Male	Left eye redness	Extra axial dumbbell shaped mass	Rhabdomyoma
Vandewalle G et al., [11]	1995	4 years/ Male	Progressive right sided facial paresis	Enlargement of the roots of the right seventh and eighth cranial nerves. Enhancing mass emerging from the porus acousticus internus	Rhabdomyoma
Vajramani G et al., [1]	1999	4 years/ Male	Impaired sensation on the left side of the face, with swelling in the left temporal region and difficulty in opening the mouth.	Large hyperdense, nonenhancing mass lesion in the left infratemporal fossa, extending intracranially near to the petrous apex, partly in the middle fossa and partly in the posterior fossa	Neuromuscular hamartoma
Castro DE et al., [2]	2005	15 years/ Male	Asymptom-atic	Well-circumscribed mass centered in Meckel's cave and extending into the foramen ovale along the V3 division.	Neuromuscular hamartoma
Tobias S et al., [7]	2006	36 years/ Female	Long standing headache	Enhancing mass lesion in right Meckel's cave, enlarging the trigeminal ganglion and extending through the foramen ovale.	Neuromuscular hamartomas
Present case	2021	30 years/ Female	Headache and facial numbness on left side.	Heteroge-neous non-enhancing mass lesion centered in the left Meckel's cave involving the trigeminal nerve with an associated fatty component.	Neuromuscular choristoma

[Table/Fig-4]: Choristomas of the cranial nerves.

Differential diagnosis	T1	T2	DWI	Postcontrast Enhancement
Schwannoma	Hypointense	Hetrogenously Hyperintense	-	Heterogeneous
Meningioma	Isointense	Hypointense	-	Avid
Paraganglioma	Heterogeneous (Salt and Pepper)	Heterogeneous	-	Avid
Epidermoid	Isointense	Hyperintense	Present	Peripheral rim
Choristoma	Hypointense	Heterogeneous	-	Minimal

[Table/Fig-5]: Differential diagnosis of lesions in the Meckel's cave region [7,13,14].

CONCLUSION(S)

Trigeminal choristoma, being a rare tumour is often misdiagnosed. MRI helps in diagnosing, and differentiating it from other lesions. The clinical, radiological and histological findings support the diagnosis of choristoma. Hence, it should be considered in the differentials of tumours affecting the trigeminal nerve with atypical characteristics, particularly in young patients.

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PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Student, Department of Radiodiagnosis, Mahatma Gandhi Medical college and Research Institute, Pondicherry, India.
2. Associate Professor, Department of Radiodiagnosis, Mahatma Gandhi Medical college and Research Institute, Pondicherry, India.
3. Associate Professor, Department of Radiodiagnosis, Mahatma Gandhi Medical college and Research Institute, Pondicherry, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. CS Prabhu,
Associate Professor, Department of Radiodiagnosis, Mahatma Gandhi Medical College and Research Institute, Pillayarkuppam-607402, Pondicherry, India.
E-mail: radio.prabhucs@gmail.com

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