

# TUBER CINEREUM HAMARTOMA

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## ABSTRACT

Tuber cinereum hamartoma is a type of midline dysraphic syndrome and presents with ectopic cerebral grey matter. Hamartoma of tuber cinereum is a well known cause of central precocious puberty. A case of gelastic seizure in a 20 years old female which was subsequently diagnosed as tuber cinereum hamartoma is presented here.

## Introduction

A clinical case of uncontrolled gelastic seizure with characteristic features of tuber cinereum hamartoma in Magnetic resonance imaging is presented.

## Case Presentation

A 20 years old lady presented with gelastic seizure with typical history of uncontrolled laughter without any loss of consciousness. Following high clinical suspicion she underwent Magnetic resonance imaging with contrast study and was diagnosed to be a case of pedunculated tuber cinereum hamartoma. MRI scan revealed a small, well circumscribed pedunculated lesion in interpeduncular cistern, projecting into the suprasellar region between mammillary bodies and optic chiasm. It was isointense on T2, iso to hypointense on T1 and there was no contrast enhancement on post contrast study.

## Discussion

Tuber cinereum is a part of the hypothalamus (diencephalon), located between optic chiasm and mammil-

lary bodies. The word hamartoma is derived from a Greek word 'hamartion' which means 'bodily defect'. Tuber cinereum hamartoma is a focal non-neoplastic lesion with varying degree of differentiation.<sup>1</sup>

It is basically an ectopic cell rest of grey matter with normal neuroglial tissue but are arranged in a haphazard fashion.<sup>2</sup>

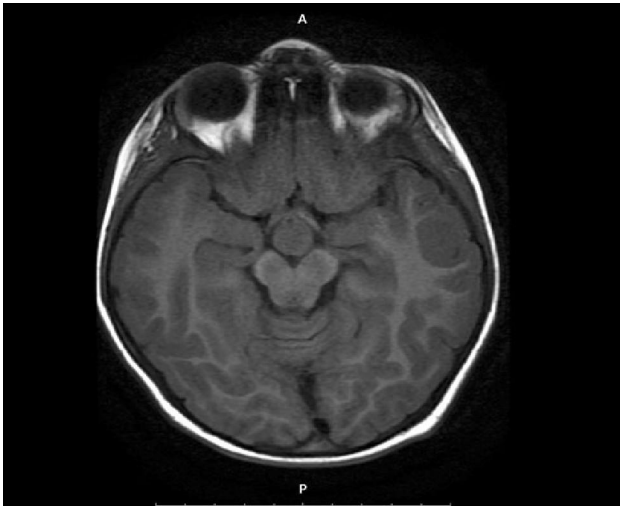
It can be of sessile and pedunculated variety and usually presents with gelastic seizure, precocious puberty or sometimes with visual impairment due to mass effect. These clinical features are noted in early life.

Structural anomalies such as corpus callosal agenesis, microgyria, polydactyly, heteropia, cardiac and facial anomalies may be associated with it.<sup>3,4</sup> It may also be associated with mental impairment, Lawrence Moon-Biedl syndrome and metabolic disturbances such as DI or GH secreting pituitary adenoma.

Plain x-ray of skull is usually normal but local erosion of tip of dorsum sella may be seen because of mass effect by the growing hamartoma.<sup>4</sup> A CT should be the next investigation of choice. A characteristic finding of tuber cinereum hamartoma is a isodense,

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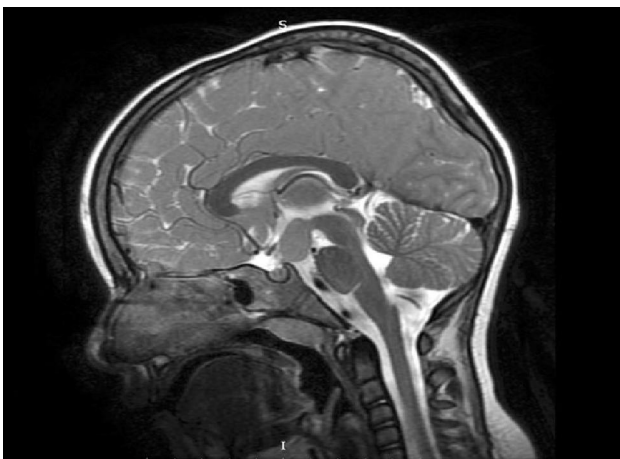
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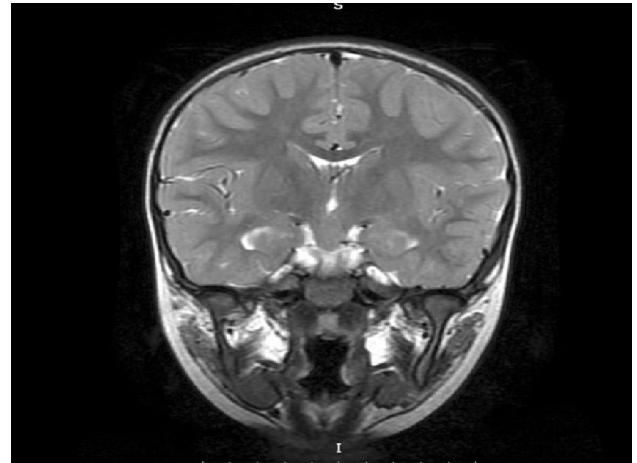
**Figure 1:** T1 AXIAL (showing a well circumscribed mass lesion in interpeduncular cistern which is iso to hypointense as compared to the cerebral grey matter)



**Figure 2:** T2 AXIAL (showing a well circumscribed mass lesion in interpeduncular cistern which is isointense to cerebral grey matter)



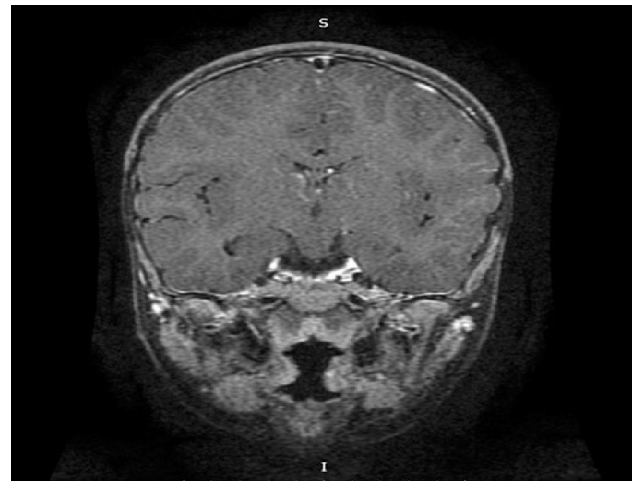
**Figure 3:** T2 SAGITTAL. <TR-4300,TE-100> (showing a pedunculated mass lesion extending from tuber cinereum into suprasellar region)



**Figure 4:** COR T2, <TR-4542,TE-98> (showing a mass lesion extending from base of the brain to suprasellar region without causing any significant mass effect)



**Figure 5:** T1 SAGITTAL+C, <TR-1900,TE-75> (post contrast study shows no obvious contrast enhancement of the aforesaid mass lesion)



**Figure 6:** T1FS COR +C (post contrast study shows no contrast enhancement of the mass lesion)

well circumscribed collar-button shaped soft tissue mass projecting into post chiasmatic region, between the infundibular stalk and mamillary bodies. On MRI the mass appears isointense on T1 and slightly hyperintense on T2, T2 hyperintensity may increase if the lesion contains more glial tissue, occasionally signal characteristics can be heterogenous with central hypointensity and peripheral rim of isointensity on T1 and T2. No calcification or contrast enhancement is noted.<sup>5</sup>

A few suprasellar mass lesions should be included in differential diagnosis such as glioma, infundibuloma, pituitary adenoma, craniopharyngioma and chiasmatic glioma.

Hamartoma has no tendency of neoplastic evolution. So, medical therapy of symptom is targeted. Surgery is reserved for the cases of intractable epilepsy.<sup>6</sup>

## Conclusion

Tuber cinereum hamartoma is haphazardly oriented ectopic rest of normal cerebral grey matter with low morbidity and mortality. Characteristic presentation is central precocious puberty or gelastic seizure. MRI brain should be the imaging investigation of choice, however histopathological diagnosis is confirmatory.

## Aknowledgements

**Competing interests-** We have no financial or personal interest in writing this report.

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