Hypothalamic hamartoma
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Section: Paediatric radiology
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 15 months, female

Clinical History:
Precocious puberty.

Imaging Findings:
This patient was reviewed in the endocrinology clinic with a 4-week history of bilateral breast enlargement and development of pubic hair. For two days there had been evidence of menarche. The patient was born at full term after an uneventful labour. Her development had been normal. On examination, she had stage 3 breast development and stage 2 pubic hair development. Her nappy was stained with a significant amount of blood. Systems examination was normal. An ultrasound examination revealed an adult type uterus. MR imaging showed a 3cm bilobular pedunculated mass arising from the floor of the third ventricle showing similar signal characteristics to grey matter, i.e. it did not show enhancement after gadolinium administration. This mass extended into the prepontine cistern. With these features, a diagnosis of hypothalamic hamartoma was made. The patient was started on GnRH analogue (Zoladex).

Discussion:
Hypothalamic hamartomas, also known as hamartomas of the tuber cinereum, are rare congenital malformations occurring in the region of the tuber cinereum or mamillary bodies. They resemble grey matter and their histological pattern resembles that of the tuber cinereum. They are composed of mature neuronal and glial tissue. Hypothalamic hamartomas of the parahypothalamic type are well-defined round lesions attached to the tuber cinereum by a stalk. Other cases occur inside the hypothalamus and are known as the intrahypothalamic type. They manifest in childhood and boys are affected more frequently than girls. The most common manifestation is isosexual precocious puberty, but patients can present with gelastic (laughing) seizures and behavioural abnormalities. The affected patients are usually large for their age. There is also bilateral breast development and pubic hair development. Menarche is evident in some children. In general, parahypothalamic type patients tend to have precocious puberty and intrahypothalamic type patients are likely to have intractable seizures and mental retardation. An initial ultrasound of examination of ovaries and adrenals should be carried out as the lesions from these sites could resemble the symptoms and signs of hypothalamic hamartoma. On CT scanning, hypothalamic hamartomas appear as a discrete, circular mass. They don’t show enhancement with contrast and appear isodense with no evidence of calcific areas. Occasionally they may have a cystic component which projects into the middle cranial fossa. Hypothalamic hamartomas show similar morphology on MR imaging and they are elegantly demonstrated because of the multiplanar capability of this technique. The lesions appear isointense to gray matter on T1 weighted images and slightly hyperintense on T2 weighted images. Intrahypothalamic and parahypothalamic hamartomas can be differentiated on MR appearances. Their size varies from a few millimetres to 3-4cm. No enhancement with intravenous gadolinium is seen. A review of recent medical literature indicates that gonadotrophin releasing hormone analogues have proved effective in the treatment of hypothalamic hamartomas and should be used as
initial management in preference to surgery. Surgery is considered an effective measure in refractory lesions.

**Differential Diagnosis List:** Hypothalamic hamartoma

**Final Diagnosis:** Hypothalamic hamartoma

**References:**

Carty H (ed)
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The NICER Institute, Oslo, Norway, p. 257 (2001).

Stewart L, Steinbok P, Daaboul J.
Role of surgical resection in the treatment of hypothalamic hamartomas causing precocious puberty.
Description: T1 axial image showing a pedunculated suprasellar mass. This mass is isointense to grey matter. Origin:
**Description:** T1 sagittal image showing a 3cm pedunculated mass between the infundibulum and the mammillary bodies. **Origin:**