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MR Imaging of Large Hypothalamic Hamartomas in Two Infants

Hamartomas of the hypothalamus are uncommon congenital tumors. Most originate in the area of the tuber cinereum and extend into the basal cisterns. They rarely exceed 2 cm in diameter.

Case Reports

Case 1 was an 11-month-old boy referred for the evaluation of apnea and seizures, which he had had since birth. He had syndactyly, polydactyly, a cleft epiglottis, and developmental delay. He was hypoglycemic, hypothyroid, and deficient in growth hormone. Case 2 was a 6-month-old boy referred because of mild developmental delay. A large suprasellar mass was found in both patients on CT and MR (Figs. 1 and 2). Because of the central location of the masses, neither patient had a biopsy. Clinically, the infants remained unchanged 1 year after their initial visits.

Discussion

Histologically, hamartomas of the hypothalamus most closely resemble the posterior hypothalamus. The masses typically are small and pedunculated and look like a collar button. They fill the space between the optic chiasm and pons [1]. Signs and symptoms usually are seen early in life. The most frequent presentations are precocious puberty (35–70%), seizures (48%), and developmental changes [2]. Usually, no focal neurologic signs are seen. Precocious puberty rarely occurs with large hamartomas [3].

CT typically shows a mass situated posterior to the dorsum sellae and the pituitary stalk, anterior to the pons and cerebral peduncles, between the two internal carotid arteries and medial surface of the two temporal lobes. The mass is the same density as the surrounding cerebral tissue and does not change after administration of contrast material [4].

MR is helpful because of the ability to obtain direct sagittal and coronal images. As seen in our two cases, T1-weighted sagittal images are superior to CT scans for assessing the origin, extent, and mass effect of the tumors [5]. T2-weighted images are important to show that these lesions are isointense with the neighboring temporal lobes. This should help to differentiate the mass from other tumors that occur in the region [6].

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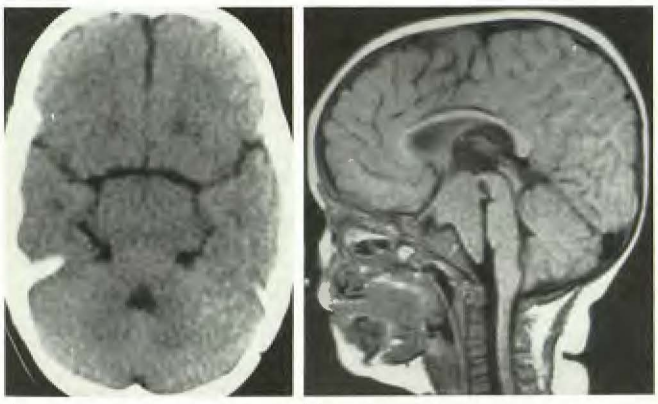

Fig. 1.—Case 1: Hypothalamic hamartoma in 11-month-old boy.
***A*, CT scan shows suprasellar mass 2 × 3 cm.**
***B*, Sagittal T1-weighted MR image, 750/30, shows mass originating from hypothalamus and extending down along clivus, with compression of pons.**

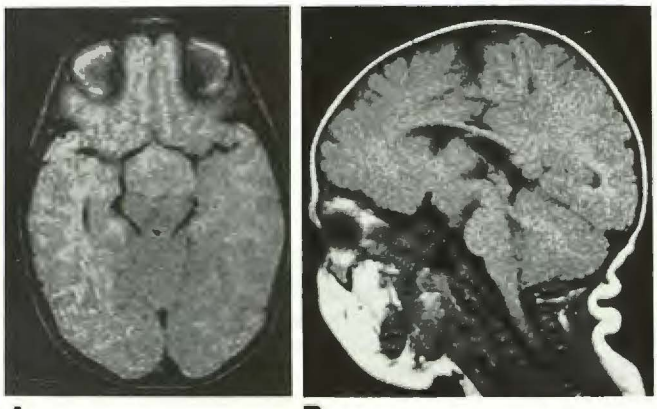

Fig. 2.—Case 2: Hypothalamic hamartoma in 6-month-old boy.
***A*, Axial T2-weighted MR image, 2100/100, shows a homogeneous suprasellar mass 2 × 3 cm with same signal intensity as surrounding temporal lobes. Effacement of pons is present.**
***B*, Sagittal T1-weighted MR image, 750/30, shows mass extending down from hypothalamus to level of mid pons.**