Intracranial ectopic pituitary adenoma

Case report

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The authors report a unique case of ectopic intracranial pituitary adenoma, associated clinically with generalized seizures and aggressive behavior. The lesion presumably arose from cells in the pars tuberalis and did not involve the sella turcica.

KEY WORDS - ectopic pituitary adenoma - adenohypophysis - chromophobe adenoma - pituitary adenoma

Potential extrasellar sites for the development of pituitary adenomas include intracranially the pars tuberalis, and ventral to the sella the body of the sphenoid and the posterior surface of the nasopharynx. A unique case of an adenoma arising ventral to the sella within the sphenoid was reported by Erdheim in 1909. According to Russell this tumor presumably arose within “an embryonic nest at the site of the obliterated craniopharyngeal duct.” To the authors’ knowledge, our case is the first reported example of an intracranial ectopic pituitary adenoma.

Case Report

This 15-year-old boy was admitted to Downstate Medical Center for evaluation of a hypothalamic mass. His growth and development had been normal until the age of 12, when he developed generalized seizures. He was initially controlled with myoline and dilantin but in the months prior to the current admission the seizures became more frequent and were associated with outbursts of violent behavior and headaches. A recent work-up at another hospital, including pneumoencephalography and cerebral angiography, reportedly revealed a hypothalamic mass for which surgery was not recommended.

Examination. The patient’s intellectual function appeared normal although no formal testing was performed. He was subject to outbursts of violent behavior and abused the hospital staff on several occasions following admission. The cranial nerves were normal except for papilledema without hemorrhages. His gait was mildly ataxic. He had a mild right hemiparesis with right hyperreflexia. Plantar responses were flexor bilaterally. Sensation was intact. An electroencephalogram from the previous hospitalization was interpreted as consistent with a convulsive disorder with bilateral cerebral dysfunction. A sonogram revealed a 2 to 3 mm right-to-left shift of the midline structures.
Plain skull films revealed the sella turcica to be normal. Angiography done by means of selective left vertebral catheterization revealed slight local displacement of a thalamo-perforating artery which arose from the basilar artery. Pneumoencephalography confirmed the presence of a round mass indenting the inferior aspect of the third ventricle just posterior to the infundibular recess (Fig. 1). Frontal and lateral tomography during pneumoencephalography revealed no continuity between the mass and the sella turcica (Fig. 2). The smooth round configuration of this lesion seen radiographically suggested a cystic mass, and preoperatively a craniopharyngioma or possibly a cystic hypothalamic tumor were considered most likely, given the location and age group.

Operation. The patient was operated on and the mass removed by way of a right frontotemporal approach. At surgery the mass was noted to be invaginating the inferior aspect of the third ventricle just superior to the interpeduncular fossa, and compressing the medial surface of the right temporal lobe. There was no connection between the mass and the sella turcica. A gross total removal of...
Pathological Findings. Microscopic examination of the first tissue fragment removed showed uniform small chromophobe cells with round nuclei and scanty agranular cytoplasm, arranged in cords and nests between prominent thin-walled sinusoids (Fig. 3). This sinusoidal arrangement is one of the characteristic patterns seen in chromophobe adenomas of the pituitary.

The second group of tissue fragments removed showed the microscopic features of normal adenohypophysis. Large granular and smaller agranular cells are arranged in acini (Fig. 4). Periodic-acid-Schiff (PAS) and Orange-G stains demonstrate PAS-positive cells (basophils), Orange-G-positive cells (acidophils), and cells with pale staining cytoplasm (chromophobes).

Postoperative Course. Immediately after surgery the patient developed transiently a right third nerve palsy and diabetes insipidus. Endocrine evaluation subsequently revealed persistent evidence of hypothyroidism and hypoadrenalism secondary to hypothalamic or pituitary insufficiency.

Fig. 3. Photomicrograph of pituitary adenoma, showing agranular cells with round nuclei arranged in cords and masses around large sinusoids. H & E, X 160.

Fig. 4. Left: Photomicrograph of anterior pituitary tissue, showing normal acinar arrangement. Light staining, agranular cells are chromophobes, and cells with darker staining cytoplasm are acidophils. PAS-Orange-G reticulin stain, X 250. Right: Higher power view of adenohypophyseal tissue, showing chromophile and chromophobe cells in acinar arrangement. PAS-Orange-G hematoxylin stain, X 400.
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Comment

This was a unique case of extrasellar pituitary adenoma. The tumor presumably originated from cells in the pars tuberalis. There was histological evidence in the surgical specimen of a normal pituitary gland and development of hypopituitarism postoperatively. The question is therefore raised as to the possibility of an extrasellar location of the normal adenohypophysis.

References


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