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AJNR Am J Neuroradiol 1990, 11 (2) 403-406
<http://www.ajnr.org/content/11/2/403.citation>

This information is current as
of May 13, 2025.

Granular Cell Tumor (Choristoma) of the Neurohypophysis: Two Cases and a Review of the Literature

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Primary tumors of the neurohypophysis and infundibulum are extremely rare. Two types are known: glioma, also known as pilocytic astrocytoma or pituicytoma; and granular cell tumor, also known as choristoma, myoblastoma, or granular cell myoblastoma. Several authors [1–7] suggest that they are a fairly common incidental finding at autopsy, but reports of masses large enough to cause symptoms are few; the last report in the radiologic literature was in 1956 [3]. We are unaware of previous descriptions of the MR appearance of granular cell tumors. We present two cases of this rare neoplasm and a review of the literature.

Case Reports

Case 1

A 62-year-old man presented in February 1984 with sudden diplopia. Physical examination was significant for a left sixth nerve palsy. A CT scan showed a 1.2×1.2 cm enhancing mass in the posterior suprasellar region, superior and adjacent to the posterior clinoid processes. The mass was hyperdense on an unenhanced scan (Fig. 1A) and showed intense homogeneous enhancement with IV contrast material (Fig. 1B). There was no associated bony erosion or destruction. Angiography revealed no abnormality. An endocrine workup was unremarkable. Because of the small size of the patient's sphenoid sinus, transsphenoidal resection was not undertaken. A right temporal craniotomy was performed; the mass was very vascular and not completely resectable. The mass was biopsied; the histology was described as that of choristoma.

The patient did well after surgery except for mild diabetes mellitus. He was treated with 6000 rad of external beam therapy in June 1984. In December 1984, he developed fatigue and unsteadiness. Neurologic examination was negative except for unsteady gait. In January 1986, his mental status began a slow deterioration. CT scan showed the mass to be unchanged; there was evidence of a right temporal craniotomy and right temporal lobe encephalomalacia (Fig. 1C). By May 1988, the patient was demented. MR showed a 1.1×1.2 cm suprasellar mass isointense with brain on 600/20/2 (TR/TE/excitations) (Figs. 1D and 1E) and 2000/80 (Fig. 1F) images. Right temporal encephalomalacia was unchanged.

Case 2

A 56-year-old woman first presented in 1983 with a history of confusion and memory changes of several months' duration. Neurologic examination was significant for slowed serial 7's and a questionable bitemporal hemianopia. Laboratory studies revealed a slightly elevated prolactin and a very low luteinizing hormone; these findings suggested secondary hypogonadism, presumably from pituitary stalk compression.

CT performed in July 1983 at an outside hospital showed a hyperdense, enhancing, well-circumscribed 5×5 cm suprasellar mass (Figs. 2A and 2B). Angiography (Figs. 2C and 2D) revealed a suprasellar tumor blush persisting throughout the venous phase. Feeding branches arose from the meningohypophyseal trunks and superior hypophyseal arteries bilaterally. There was downward displacement of the right posterior communicating artery, the A1 segments of the anterior cerebral artery, and the left choroidal artery. There was no evidence of vascular encasement or of arteriovenous shunting. These findings were suggestive of meningioma.

A right frontal craniotomy was performed. The tumor was well-encapsulated; it extended to the hypothalamus and displaced the chiasm, optic tracts, and optic nerves superiorly. The carotids and A1 and M1 segments were also displaced by the mass. It was deemed unresectable. Extensive bleeding was encountered when it was biopsied. Frozen and permanent pathology sections were described as granular cell myoblastoma.

Postoperatively, the patient had a left lower quadrantanopia and persistent lethargy. She received 5000 rad to the sella, but her condition did not improve, and she was eventually transferred to a nursing home.

In November 1986, she developed acute hydrocephalus. A CT scan at that time showed a 3.8-cm enhancing suprasellar mass. She was treated with ventriculoperitoneal shunt. Therapy for mild hypopituitarism was also instituted.

In November 1987, she suffered a respiratory arrest. CT scan showed bleeding around the tumor and into a dilated fourth ventricle. The third ventricle was obliterated. The patient's course was marked by repeated fever spikes without an identifiable source; they were thought to be of central origin. Comatose, she underwent a tracheostomy and was returned to the nursing home.

Received December 30, 1988; revision requested February 17, 1989; revision received June 2, 1989; accepted June 4, 1989.

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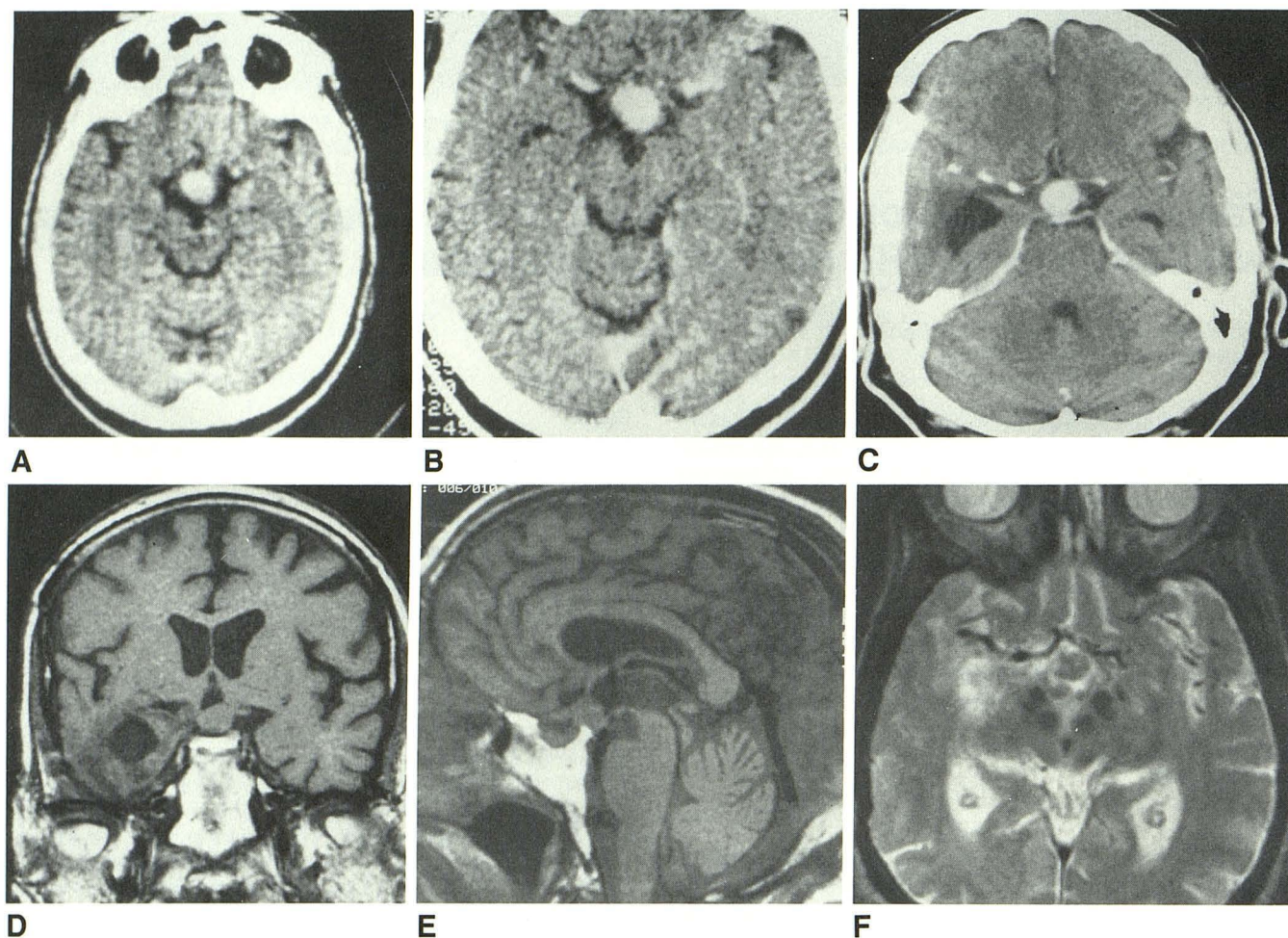


Fig. 1.—Case 1.

A and B, Noncontrast (A) and contrast-enhanced (B) CT scans of hyperdense suprasellar mass show no significant contrast enhancement.

C, Postoperative CT scan shows contrast-enhancing suprasellar mass and evidence of right temporal craniotomy and encephalomalacia from surgery.

D–F, Suprasellar mass is seen to be isointense with brain in these 600/20 (D), 600/20 (E), and 2000/80 (F) (TR/TE) MR images.

Discussion

Pathology

Choristoma, granular cell tumor, pituicytoma, and myoblastoma are names given to a benign tumor occurring in the neurohypophysis. The tumor has a characteristic histologic appearance, and is composed of large polygonal cells with small nuclei and granular cytoplasm, usually staining positively with PAS [1, 2, 4, 5, 7–9]. The variety of names given to this tumor indicates the uncertainty regarding the cell of origin. Some authors believe that the tumor originates from the pituicyte, which represents a modified astrocyte [5, 7–9], and a few use the name pituicytoma [6, 10]. Histologically, the tumor is very different from the tumor occurring in this region called pilocytic astrocytoma, which is undoubtedly of astrocytic origin [1, 6, 10, 11]. Choristoma is another name given to the tumor reported here, but this is a term used to indicate ectopic rests of normal cells [12]. Rubenstein [5] and Russell

and Rubenstein [6] cite a case of glioma containing choristoma cells. To complicate the matter further, a tumor identical to the type reported here has been found occasionally in the tongue, subepidermal tissue of the upper body, breast, and gastrointestinal, genitourinary, and biliary tracts [1, 2, 4, 5, 7–9, 12]. The names granular cell tumor and granular cell myoblastoma have also been used. There is no evidence that the tumor originates from muscle cells or from Schwann cells [12], as has been considered. Until further evidence becomes available regarding the origin of this tumor, the purely descriptive name of granular cell tumor seems to be the most appropriate.

Clinical and Radiologic Findings

Very few cases of granular cell tumor of the neurohypophysis are described in the clinical or radiologic literature.

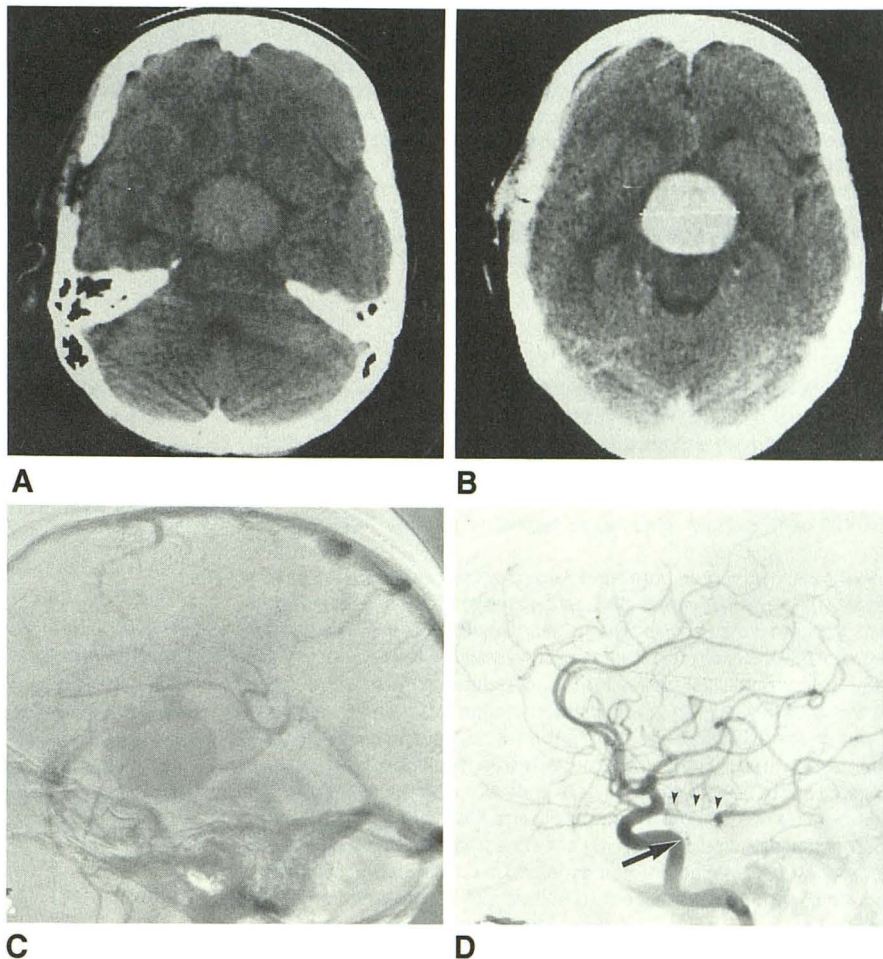
Headache, decreased vision, decreased libido, and infertility

Fig. 2.—Case 2.

A and B, CT scans without (A) and with (B) IV contrast enhancement show large, enhancing suprasellar mass.

C, Angiogram of venous phase of right internal carotid artery shows prolonged blush in location of the mass.

D, Angiogram of arterial phase shows hypertrophic meningeohypophyseal feeders (arrow) and inferior displacement of posterior communicating artery (arrowheads). (Angiograms courtesy of S. M. Wolpert and E. S. K. Kwan.)



are the most frequently mentioned presenting symptoms; in fatal cases, features of increased intracranial pressure develop [2, 3, 4, 7–9]. Examination usually reveals a bitemporal hemianopia with optic nerve atrophy, and, in male patients, decreased body hair. In older patients [2, 4, 7], plain films and pneumoencephalography have revealed supra- and/or intrasellar masses, often eroding bone and elevating the anterior third ventricle. Angiography has been performed in three cases. Doron et al. [9] reported elevation of the left anterior cerebral artery and a definite tumor stain. The anterior cerebral artery was also elevated in case 1 of Symon et al. [4]. These authors also noted slight unfolding of the right carotid siphon, but no evidence of tumor stain or neovascularity. Their second case, however, showed sellar feeding branches from the parasellar carotid and a suggestion of intrasellar pathologic circulation. In most cases, craniotomy was the procedure of choice, and though profuse hemorrhage was always encountered, the tumors, though small and noninvasive, tended to be quite firm and difficult to remove. Decompression and partial excision was usually employed [3, 4, 7, 9], frequently with good results. Radiation therapy in one case [3] had no effect; it is difficult to assess the effects of radiation therapy in the case reported by Doron et al. [9]

because it was employed after partial excision, and no follow-up radiologic examinations were described.

The MR scan in our first case showed an isointense mass in T1-, proton-density, and T2-weighted images. This may help in the differential diagnosis of sellar and suprasellar masses. However, the similarity to the MR appearance of meningioma suggests that the neurosurgeon be prepared to encounter a potentially hemorrhagic and unresectable granular cell tumor when operating for suprasellar meningioma.

ACKNOWLEDGMENTS

We are grateful for the assistance of the Radiology Departments at St. John's Hospital, Lowell, MA.; Lowell General Hospital, Lowell, MA.; and New England Medical Center Hospital, Boston, MA. We thank Eddie S. K. Kwan, and Samuel M. Wolpert for supplying the angiographic images, and Daniel E. Leonard for his assistance with the preparation of this article.

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