Nasopharyngeal craniopharyngioma in an unusual location.

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Nasopharyngeal Craniopharyngioma in an Unusual Location

Nachiketa Kanungo, Norman Just, Martin Black, Gérard Mohr, Rafael Glikstein, and Louise Rochon

Summary: MR and CT are used to image a rare case of craniopharyngioma arising in the pharyngeal hypophysis, sparing the sella turcica, and spreading into the anterior fossa through the cribriform plate.

Index terms: Craniopharyngioma; Nasopharynx, neoplasms

Since 1938, nine cases of nasopharyngeal involvement by craniopharyngioma have been described in the medical literature (1–3). Six of these cases involved the sella turcica, and all but one (1) extensively involved the sphenoid sinus.

We report a case of a nasopharyngeal craniopharyngioma sparing the sella turcica. The tumor is thought to have likely arisen from the pharyngeal hypophysis. Magnetic resonance and computed tomography are used to define the tumor and its unusual spread into the anterior fossa through the cribriform plate.

Case Report

A 40-year-old woman presented with malaise and 2 months of nasal obstruction and headaches, worse on the right side.

General physical findings were normal, but rhinoscopy revealed a lobulated fleshy pink mass obstructing the right nasal cavity and filling the nasopharynx. Laboratory blood findings were normal. Plain sinus films showed diffuse opacification of all the paranasal sinuses and the right nasal cavity (Fig 1).

Axial and coronal computed tomographic scans with intravenous contrast showed a large nasopharyngeal mass with some central calcification filling the nasopharynx, the right nasal cavity, and the ethmoid air cells. Some of the enhancing tumor invaded the anterior fossa through the cribriform plate and the roof of the ethmoid sinus (Fig 2). All of the paranasal sinuses contained some soft-tissue density consistent with tumor, mucocele, or sinusitis. The sella turcica and suprasellar regions were normal.

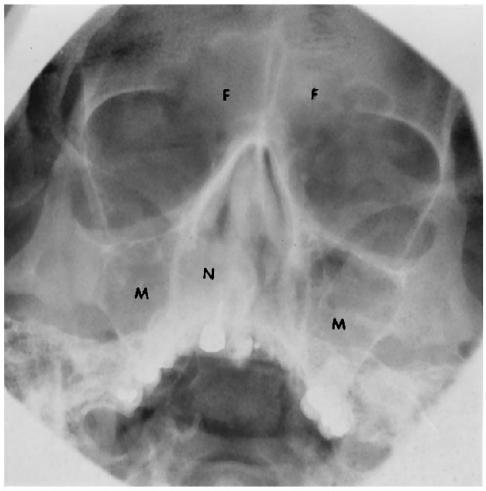

Fig 1. Water's view shows opacification of frontal (*F*) and maxillary (*M*) sinuses, as well as the right nasal cavity (*N*).

T1-weighted (plain and gadolinium-enhanced 600/11/2 [repetition-time/echo time/excitations]) and T2-weighted (3000/108/1) magnetic resonance showed the brightly enhancing, heterogeneous mass clearly to invade the anterior fossa but to spare the frontal lobes (Fig 3). The frontal sinus and portions of the posterior ethmoid air cells and sphenoid and maxillary sinuses showed signal characteristics suggesting obstruction. There was extension of the tumor through the anterior wall of the sphenoid sinus.

A bifrontal craniotomy and right lateral rhinotomy were performed. En bloc resection of the tumor, olfactory nerves, the cribriform plate, and the nasal septum (perpendicular plate of the ethmoid) was carried out. The nasopharyngeal component was removed with the rest of the specimen via the rhinotomy. The anterior fossa component remained in the epidural space grossly (covered by a thinned dura) but involved the right olfactory nerve microscopically. Extensive extradural and intradural duraplasty using a pedicled pericranial flap and fibrin glue was performed after reconstruction of the bone defect with a split calvarial graft. The frontal and sphenoid sinuses were found to contain mucoceles. Drainage of these sinuses was reestablished. The bone walls of the sphenoid sinus

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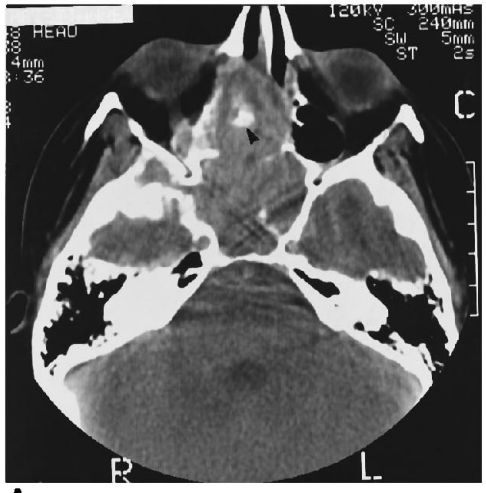
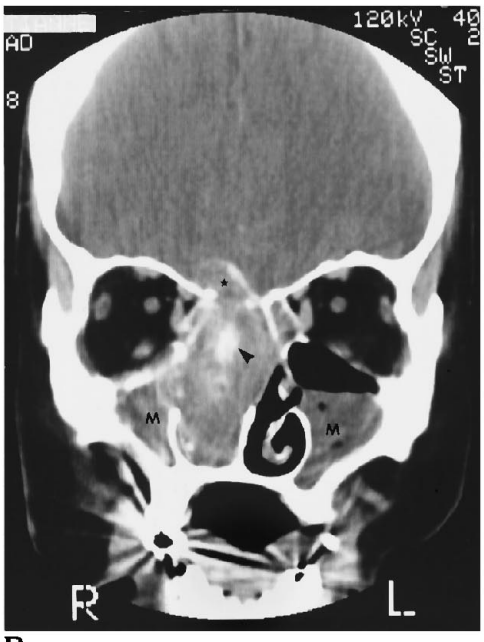


A



B

Fig 2. Axial (*A*) and coronal (*B*) computed tomographic scans show a nasal and nasopharyngeal soft-tissue mass with calcification (*arrowhead*) invading into the anterior fossa (*star*). Maxillary sinus disease is again seen (*M*) (Elscint 2400, Exel, Haifa, Israel).

were intact and free of tumor, except for erosion of the anterior wall.

All gross tumor was resected, and the patient had an uncomplicated recovery. She was discharged from the hospital 3 weeks after surgery and remains free of symptoms 6 months later.

Histologic examination revealed a classic adamantinomatous craniopharyngioma (Fig 4) with epithelial islands showing peripheral palisading and intercellular edema of central stellate areas. Cystic degeneration occurred both in the stroma and the central reticulum. The latter was frequently keratinized and associated with calcifications. In a few sections, the tumor contained bone lamellae with reactive new bone formation, presumably representing destruction of preexisting bone structures.

Discussion

Craniopharyngiomas are benign epithelial tumors, which are generally found intracranially with similar frequency in children and adults (4). Two clinicopathologic varieties have been delineated (4, 5): the papillary squamous type, which is seen almost exclusively in adults and carries a better prognosis; and the classical adamantinomatous variant, of which this case is an example.

The theory of the craniopharyngioma originating from the remnants of the obliterated craniopharyngeal duct as described by Erdheim (6) suggests that these lesions can arise anywhere along the migration of Rathke's pouch from the vomer, the roof of the nasopharynx, through the midline sphenoid bone to the floor of the sella turcica.

This course traversed by Rathke's pouch forms a cord of cells joining the stomodeal ectoderm to the future adenohypophysis in the embryo (7). The pharyngeal hypophysis (functioning adenohypophyseal tissue), which remains in adults (1, 8), is a caudal remnant of this cord, and it gave rise to one previously described craniopharyngioma (1).

The present tumor almost certainly arose in the nasopharyngeal hypophysis, because it eroded only the anterior wall of the sphenoid bone but spared the remainder of the sphenoid (including the sella turcica) despite its large size.

Despite its uncommon location and unusual route of spread, radiologic and histologic appearance are typical of craniopharyngioma. Esthesioneuroblastoma, nasopharyngeal carcinoma, and juvenile angiofibroma should be considered in the radiologic differential diagno-

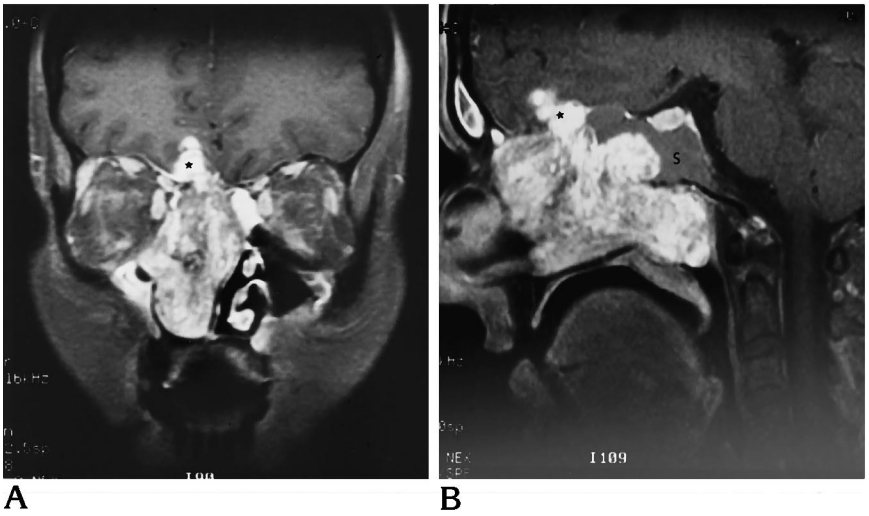


Fig 3. Coronal (*A*) and sagittal (*B*) 600/11 magnetic resonance scans with gadolinium show a heterogeneously enhancing nasopharyngeal mass with extension into the anterior fossa (*star*). Much of the sphenoid sinus (*s*) contents do not enhance, which is consistent with mucocele. Other sections showed the frontal and maxillary sinuses to be opacified by secretions and not by tumor (1.5-T, GE Signa, Milwaukee, Wis).

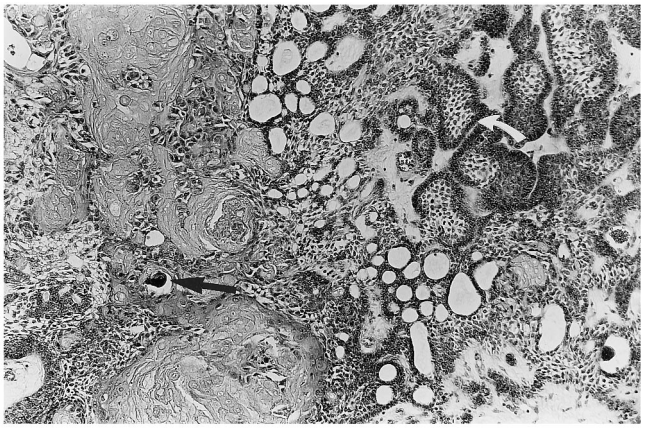


Fig 4. Classic adamantinomatous craniopharyngioma with peripheral palisading (*white arrow*), dyskeratosis, ghost cells, and calcification (*black arrow*). Multiple microcysts are seen in the center of the field (hematoxylin and eosin, original magnification ×125).

sis, although the latter occurs almost exclusively in young boys. The location of the tumor is supportive of the theory of adenohypophyseal embryogenesis as suggested by Erdheim in 1904.

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