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Clinical Study

Intrasellar pilocytic astrocytomas: Clinical, imaging, pathological, and surgical findings

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ABSTRACT

A pilocytic astrocytoma is not usually considered in the differential diagnosis of an intrasellar tumor. An awareness of this tumor as primarily an intrasellar entity is important to avoid confusion during its diagnosis. We retrospectively examined the records of 631 patients treated at our institution between 2006 and 2010 who underwent transsphenoidal resection of pituitary tumors and identified those diagnosed with pilocytic astrocytoma. We excluded patients who harbored a pituicytoma. We also searched the literature for patients with a histologically confirmed diagnosis of intrasellar pilocytic astrocytoma. Only two patients in our series harbored intrasellar tumors and had a histologic diagnosis of pilocytic astrocytoma. We also found two other cases in the literature that met our criteria. Pilocytic astrocytoma should be considered in the differential diagnosis of an intrasellar lesion. An understanding of this tumor's radiological features can avoid diagnostic confusion.

1. Introduction

An astrocytoma is not usually considered in the differential diagnosis of an intrasellar tumor. Historically, astrocytomas of the neurohypophysis have been classified as pituicytomas or pilocytic astrocytomas [1]. More recent pathologic studies suggest that pituicytomas and pilocytic astrocytomas found in the pituitary fossa are distinct tumor entities. Histologically, pituicytomas lack Rosenthal fibers and eosinophilic granular bodies, which are pathognomonic, but not required, morphologic features of pilocytic astrocytomas [1]. Accurate tumor classification is important for development of future treatments and prognosis.

Pilocytic astrocytomas have been reported to occur in unusual intracranial locations, including parasellar areas [2,3]. To our knowledge, there are currently only four reported cases of pilocytic astrocytomas originating in the sella rather than extending into the sella from the supra and para-sellar regions [4–7]. Of these, only two cases were primarily intrasellar and satisfied all the histologic criteria to be classified as pilocytic astrocytomas, including the presence of Rosenthal fibers and eosinophilic granular bodies [6,7]. We present our experience with two patients with intrasellar pilocytic astrocytomas removed through the transsphenoidal approach. An awareness of this tumor as primarily an intrasellar entity is important to avoid confusion during its diagnosis.

2. Methods

Between 2006 and 2010, a total of 631 patients underwent transsphenoidal resection of pituitary tumor by three different neurosurgeons at our institution (J.M.G., T.D.P. and A.A.C.-G.). Among these patients, only two patients had a histologic diagnosis of pilocytic astrocytoma. Patients harboring pituicytomas were excluded.

Both patients were evaluated using pre-operative MRI, complete endocrinologic evaluation, and comprehensive visual field testing. Both patients underwent the same endonasal septal displacement transsphenoidal technique for removal of pituitary tumors with lateral fluoroscopic guidance as described by Jane et al. [8]. All pathologic specimens were reviewed by the same neuropathologist (J.M.B.).

Finally, we performed a literature review for "intrasellar glioma" and "pilocytic astrocytoma" using the Ovid database and selected reports describing intrasellar tumor(s). The reported cases involving tumors originating from the extrasellar space and extending into the sella turcica were excluded; only purely intrasellar tumors were considered for inclusion in this report.

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3. Results

The clinical details of the two cases reported in the literature and our two patients with pilocytic astrocytoma are summarized in Table 1. All tumors primarily involved the intrasellar space, caused expansion of the sella turcica, and demonstrated a secondary suprasellar extension.

Our two patients included one woman and one man. The average age was 35 years (range 33–37 years). One patient presented with pituitary dysfunction and was found to have subtle visual field deficits during an ophthalmologic examination. The other presented with visual field deficits and was later found to also have pituitary dysfunction. In both patients, preoperative MRI examination revealed enhancing tumors not resembling pituitary macroadenoma.

4. Case presentations

4.1. Patient 1

The clinical details of this patient are summarized in Table 1. This 37-year-old woman had experienced decreased vision in the left eye very early in life and was found to have a sellar mass at age 11, at which time she underwent transsphenoidal hypophysectomy. At that time the pathologist could not make a diagnosis, and the surgeon told the patient's parents that the diagnosis could not be made because "the tumor does not look like any tumor that can occur in this area." The patient's vision improved with surgery, but she later developed hydrocephalus and required a ventriculoperitoneal shunt.

When the patient presented at age 37, she complained of experiencing progressive loss of vision in her left eye and amenorrhea for several years. Detailed visual field tests revealed dense visual loss in the left eye and partial temporal visual loss in the right eye. An endocrinologic evaluation was consistent with panhypopituitarism. MRI of the brain revealed a primarily sellar mass with suprasellar extension, compressing the optic chiasm. The mass was irregular and multilobulated with a bright enhancement pattern (Fig. 1A, B).

The patient underwent a repeat transsphenoidal operation for resection of the tumor. Upon opening the tumor capsule, a gelatinous yellow-gray mass was exposed and removed. The diaphragm a sella was identified and a moderate-size cerebrospinal fluid (CSF) fistula was encountered. Frozen section analysis of the lesion was consistent with an astrocytoma. Despite intraoperative repair of the CSF leak, the patient suffered from CSF rhinorrhea after surgery, but her vision improved slightly. Adjustment of her shunt valve stopped her CSF rhinorrhea. Postoperative MRI revealed a radical subtotal resection of the tumor (Fig. 1C, D).

Histologically, the tumor appeared to be a moderately cellular biphasic glial neoplasm comprised of cellular areas characterized by predominantly spindle cells with elongated nuclei, but little nuclear pleomorphism and no mitoses. Rosenthal fibers were identified (Fig. 2). In the less cellular and more sparsely populated areas, the tumor cells had a stellate appearance and there were numerous eosinophilic granular bodies. There was a prominent vascular hyalinization and frequent calcifications. Most of the tumor cells were immunoreactive for glial fibrillary acidic protein (GFAP). There was fairly intense background immunoreactivity for synaptophysin. The proliferative index was estimated at 3%. Periodic acid–Schiff stain highlighted the abundant eosinophilic granular bodies. These findings supported the histologic diagnosis of pilocytic astrocytoma.

This patient suffered a tumor recurrence 4 years later and is now undergoing radiation therapy.

4.2. Patient 2

The clinical details of this patient are summarized in Table 1 and 2. This 33-year-old man had been evaluated for infertility and diagnosed with hypopituitarism and testicular atrophy. His ophthalmologic examination was normal. MRI revealed a multilobulated enhancing solid and cystic intrasellar tumor (Fig. 3A, B). The patient underwent transsphenoidal surgery and gross total resection of the tumor. The tumor appeared grayish in color and fibrous. During surgery, a minor intraoperative CSF leak was repaired with fat graft packing.

Histologic examination revealed a cellular lesion with a biphasic pattern of alternating oligodendroglioma-like cells and more classical pilocytic astrocytes. Rosenthal fibers and numerous eosinophilic granular bodies were present (Fig. 4A). Another area of the tumor displayed cells arranged in a radial fashion around the blood vessels as well as extensive cystic degeneration with myxoid material filling the spaces (Fig. 4B). Although the diagnosis of pilomyxoid astrocytoma was considered because of the pseudorosettes and myxoid areas, the presence of abundant eosinophilic granular bodies and Rosenthal fibers supported a diagnosis of pilocytic astrocytoma. The tumor cells displayed strong GFAP immunopositivity (Fig. 4C) and a low Ki-67 proliferative index (Fig. 4D). The patient did well following resection and showed no recurrence at a 6 month postoperative follow-up examination (Fig. 3C, D).

5. Discussion

The most common neoplasms of the pituitary fossa are pituitary adenomas, although the differential diagnosis of sellar masses includes granular cell tumors, craniopharyngiomas, germ cell tumors, gliomas, meningiomas, hemangiopericytomas, chordomas, choristomas, schwannomas, neuroblastomas, lymphocytic hypophysitis, sarcoidosis, Langerhans cell histiocytosis and germ cell tumors [9–11]. Lesions in the pituitary fossa may also be benign cysts, including Rathke's cleft cyst, arachnoid cyst, epidermoid cyst, and dermoid cyst [2].

To our knowledge, the literature includes only two previous reports of primarily intrasellar pilocytic astrocytomas (Table 1). Tekkok et al. first reported an intrasellar cystic pilocytic astrocytoma resected through a transcranial approach [6]. Recently, Prasad et al.

Table 1
Patient presentation, treatment, and outcomes

Author	Age	Sex	Symptoms	Treatment and complications	Outcome
Parish et al. [current]	37	F	Bilateral visual disturbances	TPH, intraop CSF leak	Tumor recurrence 4 years later. Slight improvement in vision
Parish et al. [current]	33	M	Panhypopituitarism and testicular atrophy	TPH, intraop CSF leak	Improvement in hormonal function. No tumor recurrence at 6 months
Prasad et al. [7]	26	M	Bilateral visual disturbance	TPH, prior chemotherapy, subtotal resection, radiotherapy	Improvement in right eye vision, no recurrence at 18 months
Tekkok et al. [6]	12	M	Visual impairment, headache	Transcranial resection, radiotherapy	Symptom free 2 years after surgery

CSF = cerebrospinal fluid, F = female, intraop = intraoperative, M = male, PA = pilocytic astrocytoma, TPH = transsphenoidal hypophysectomy.