Case Report: Rapid Growth of Sellar Pituicytoma

Sumit Das1,2, Tibor Valyi-Nagy3, Martin K. Nicholas1 and Jinsuh Kim4

1Department of Pathology, University of Illinois at Chicago, Chicago, Illinois, USA
2Division of Neuropathology, University of Alberta Hospital, Edmonton, Alberta, Canada
3Department of Neurology, University of Illinois at Chicago, Chicago, Illinois, USA
4Department of Radiology, University of Illinois at Chicago, Chicago, Illinois, USA

Abstract

Pituicytoma is rare WHO grade I neoplasm of the sellar region that can clinically and radiologically mimic a pituitary adenoma. We present an unusual case of a pituicytoma in a 50-year-old female whose MRI images showed a heterogeneously enhancing sellar mass compressing the optic chiasm. Even with multiple partial resections, she continued to experience progressively worsening visual symptoms with subsequent images revealing interval enlargement of the mass. Pituicytomas are typically slow growing lesions that gradually grow in size over a period of years. Our case is unusual in that the interval growth between resections was actually quite rapid taking only a few months to significantly increase in size.

Keywords: Pituicytoma; Sella

Introduction

Pituicytoma is a relatively rare tumor arising in the neurohypophysis that can present as a sellar or suprasellar mass. Signs and symptoms include visual disturbance, headache, and hypothalamic dysfunction, which are typically secondary to mass effect and thus resemble other slow-growing lesions of the sellar region such as pituitary adenoma. On neuroimaging, pituicytomas are typically solid and circumscribed with homogenous contrast enhancement [1]. Histopathologic features include a fascicular arrangement or storiform pattern of bipolar spindle cells that display fusiform to plump cell processes and moderate to abundant eosinophilic cytoplasm. Pituicytoma is regarded as a slow-growing low grade (WHO grade I) glial neoplasm [2], typically growing over a period of years [3].

We present an unusual case of a pituicytoma that presented with heterogeneous contrast enhancement on neuroimaging and seemed to display relatively rapid regrowth after multiple partial resections.

Clinical History

Our patient is a 50-year-old right-handed female with a past medical history of diabetes mellitus, hypothyroidism, and diverticulitis who presented at an outside hospital with worsening visual acuity along with declining central vision on the right side and changes in peripheral vision on the left side. No other physical complaints or cognitive symptoms were reported. Neurological examination was remarkable for left temporal hemianopsia, while the right eye was only able to see peripherally. Neuroimaging revealed a heterogeneously enhancing mass within the suprasellar cistern compressing the optic chiasm (Figure 1a). Radiologic differential diagnoses included craniopharyngioma and glioma.

The patient subsequently underwent a partial resection of the mass via a right-sided craniotomy at the outside hospital. The partially resected mass was diagnosed as meningioma with atypical features. The histologic sections from the outside hospital were not available to us for examination. Postoperative course was complicated by diabetes insipidus, which was treated with vasopressin. Postoperative MRI demonstrating residual tumor is shown in Figure 1b.

Four months later she presented at our hospital with persisting visual symptoms similar to those during her initial presentation. Neuroimaging at this time confirmed the presence of a persistent suprasellar mass (2.4 × 2.0 × 1.9 cm) located posterior to the optic chiasm and displacing the optic chiasm anteriorly and the optic tracts laterally (Figure 1c). The mass was seen extending into the interpeduncular cistern displacing the basilar artery and proximal aspect of the posterior cerebral artery. The size of the mass was only slightly smaller than on prior neuroimaging that was performed before the initial resection. Also compared to the previous neuroimaging, the mass showed more central contrast enhancement and less intense peripheral enhancement. The mass also displaced the anterior commissure and invaded the anterior portion of the third ventricle with mass effect on the hypothalamus and mammillary bodies.

Given the persistent symptoms, the patient underwent a repeat craniotomy for another partial resection. Pathologic findings from this resection were in keeping with pituicytoma (WHO grade I). Microscopic sections revealed a fascicular arrangement of predominantly spindle cells along with some epithelioid cells without definite invasion into brain parenchyma. The neoplastic cells displayed mild to moderate amount of eosinophilic cytoplasm and minimal nuclear atypia. The neoplastic cells were diffusely immunoreactive for vimentin, S-100, and TTF-1 while focal immunoreactivity for EMA was seen (Figure 2).

Postoperative MRI performed two days later (Figure 1d) restated the suprasellar mass which confirmed partial resection of the tumor and still displayed heterogeneous enhancement within the tumor.

The patient’s bi-temporal field defects showed little change. Her panhypopituitarism manifested as adrenal insufficiency, hypothyroidism, and diabetes insipidus which were managed with hydrocortisone, levothyroxine, and desmopressin.

The patient was started on adjuvant radiation therapy for tumor remnant but unfortunately, her symptoms continued to worsen. Her radiation treatment was therefore interrupted at this point and she was admitted to our hospital for further neurosurgical evaluation. Follow-up imaging performed four months after the last neuroimaging revealed interval enlargement and centripetal filling of an enhancing T1 isointense and T2 hyperintense suprasellar mass (2.4 × 2.1 × 2.0 cm) with persistent mass effect on the optic chiasm (Figure 1e). Given the progression of the lesion and progressively worsening symptoms, the patient underwent another surgical resection via a pterional craniotomy along with decompression of the optic apparatus. Microscopic sections from
this resection specimen revealed a spindle cell neoplasm whose morphologic features and immunohistochemical profile resembled that of the prior resection. This tumor was therefore also diagnosed as a pituicytoma (WHO grade I).

Discussion

Pituicytoma is a very rare neoplasm of the neurohypophysis with approximately 80 cases reported in the English literature thus far [4–9]. It was not until 2007 that pituicytoma was recognized as a distinct pathologic entity [5]. Clinically these can mimic a pituitary adenoma presenting with a headache, visual symptoms and panhypopituitarism. Bitemporal hemianopsia can be present when the optic chiasm is compressed by the mass, as was the case with our patient. Compression of the infundibulum can interfere with the hypothalamic delivery of dopamine resulting in hyperprolactinemia, amenorrhea and decreased libido [10]. In a review by Convington et al. [11] that included thirty-five cases of pathologically confirmed pituicytomas (18 male, 17 female), the most common presenting symptom was visual disturbances (18/35) including bi-temporal hemianopsia and decreased visual acuity as was the case in our patient. The next most common presenting symptom was a headache (16/35) followed by fatigue (8/35), decreased libido (7/35), and hypopituitarism (6/35).

To our knowledge, there are currently no unique radiologic characteristics that can distinguish pituicytoma from the more common pituitary adenoma. Neuroimaging typically shows a hypointense or isointense lesion in T1-weighted images with homogenous contrast enhancement and is hyperintense on T2-weighted images [7,12,13]. The radiologic differential diagnoses include intrasellar and suprasellar mass lesions with positive enhancement: pituitary adenoma, pilocytic astrocytoma, granular cell tumors, craniopharyngioma, meningioma, ganglioglioma, germ cell tumors, sarcoidosis, and metastatic tumors. Occasionally the tumor will show heterogeneous enhancement, which was seen in our case. A pathologic diagnosis is currently the only way to definitively distinguish a pituicytoma from pituitary adenoma and other homogenously enhancing lesions that can arise in the sellar region.

Grossly, the tumor is a demarcated mass with firm rubbery texture. Histologically, pituicytomas are characterized by bipolar, short to elongate, plump to angulated spindle cells arranged in compact fascicles or storiform pattern. The nuclei show little to no atypia and mitotic figures are rare to absent [5]. Herring bodies and axons at the periphery representing posterior pituitary and stalk may be seen. Neoplastic cells are typically immunoreactive for vimentin, SMA, and TTF-1 as was seen in our case. EMA may show patchy cytoplasmic reactivity [3]. Variable GFAP immunoreactivity may also be noted [14]. Cells are usually negative for synaptophysin and pituitary hormones.

A histologic differential diagnosis to keep in mind and which was strongly considered in our case is spindle cell oncocytoma. The clinical presentation of spindle cell oncocytoma can be similar to
that of pituicytoma as well as pituitary adenoma. Neuroimaging of spindle cell oncocytoma usually shows a circumscribed contrast enhancing mass similar to both pituicytoma and pituitary adenoma. Since spindle cell oncocytoma and pituicytoma consists of overlapping immunohistochemical profiles, one must therefore return to the morphology of the tumor itself to determine a diagnosis. In our case, pituicytoma was the favored diagnosis in light of the spindle cell morphology and lack of oncytic appearance along with the immunopositivity of the neoplastic cells for TTF-1, which is more commonly noted in pituicytoma [15]. Recent literature, however, has revealed positive TTF-1 immunohistochemistry in cases of spindle cell oncocytoma, and hence one may argue this cannot be completely ruled out [7, 16]. Multiple authors have actually suggested that the TTF-1 immunoreactivity observed in both pituicytoma and spindle cell oncocytoma indicates that these entities are actually both derivatives of pituicytes [8, 17]. Interestingly, TTF-1 is encoded by the NKX2-1 gene and believed to play a role in the development of the basal forebrain and posterior pituitary [18]. Presently there is no characteristic genetic signature for pituicytoma but one case report demonstrated losses of chromosome arms 1p, 14q, and 22q and gain of 5p by comparative genomic hybridization analysis [19].

Given the rarity of pituicytomas, there are very few studies on their treatment and are primarily focused on surgical management [20–22]. Gross total resection via transsphenoidal approach has been documented by some authors [4, 10, 22]. Feng et al. [23] suggested that an endoscopic endonasal trans-sphenoidal and transplanum approach carries the best chance for total excision and the least morbidity. Conversely, craniotomy, which was the mode of treatment used in our patient, may be associated with a higher risk of visual loss or other neurologic deficits [24]. Partial resections were performed in our patient rather than total resection because of the close relationship of the tumor to the optic nerves and chiasm and it was therefore felt that risk of total resection outweighed its possible benefit. Attempted resection of pituicytoma has also been reported to be associated with unexpected intraoperative bleeding [11, 22]. The primary goal of the surgery was to relieve pressure on those vital structures in hopes of relieving the patient’s symptoms. To our knowledge data regarding targeted treatment for pituicytoma not amenable to full resection are also limited. In a study by Mende et al [25] that included ten samples of pituicytoma (7 male, 3 female; mean age = 57.8 years), all were found to be positive for vascular endothelial growth factor (VEGF) while seven cases were positive for somatostatin receptors (SSTR3 and SSTR5), representing possible targets for therapies in cases where surgical treatment alone remains insufficient. Further research and larger data sets, however, are necessary to accurately determine whether tumor growth can be inhibited by this mode of therapy. Partial resection may be associated with gradual regrowth over a period of years [3]. Our case is unusual in that the tumor seemed to regrow much more rapidly, occurring over the course of a few months after partial resection. A recent case series by Zygourakis et al. [8] of five patients diagnosed with pituicytoma and two with spindle cell oncocytomas, raised the possibility of more aggressive growth of spindle cell oncocytoma. This finding along with TTF-1 and EMA

**Figure 2:** Hematoxylin and eosin stain sections revealed a spindle cell neoplasm (A) with foci of epithelioid cells (B). Tumor cells are immunopositive for vimentin (C) and TTF-1 (D) with focal immunopositivity for EMA (E).
immunoreactivity not only makes it more difficult to completely rule out spindle cell oncocytoma but also suggests that pituicytoma and spindle cell oncocytoma exhibit similar histogenesis and are both derived from pituicytes. As mentioned previously, however, it was the histologic features in conjunction with the results of immunohistochemistry that led to the favored diagnosis of pituicytoma.

In conclusion, we present a rarely diagnosed case of pituicytoma in the sellar region where clinical and radiologic presentation closely mimic that of pituitary adenoma and exhibited fairly rapid growth. Although rare, pituicytoma, as well as spindle cell oncocytoma, should be kept in mind as part of the differential diagnosis of a circumscribed contrast enhancing sellar mass along with the more common entities such as pituitary adenoma, meningioma, and metastatic tumor. Follow-up studies examining the clinical course of partially resected pituicytommas would be helpful in guiding clinical management of such patients.

**Conflict of Interest**

The authors have no conflict of interest to disclose.

**References**


*Corresponding author: Sumit Das, Division of Neuropathology, University of Alberta Hospital, Edmonton, Alberta, Canada, E-mail: sumit.das@ahs.ca*

Received Date: June 23, 2017, Accepted Date: July 26, 2017, Published Date: August 08, 2017.

Copyright: © 2017 Sumit Das, et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.