Astrocytoma of the Pituitary gland (Pituicytoma): A Case Report

1Dr. Poonia Nemichand, 2Dr. Mangal Akshay, 3Dr. Singhvi Shashi
1Mch (Neurosurgery), Director & Senior Consultant, 2Junior Consultant, 3Senior Pathologist
1,2Neurocare Hospital, Vidhyadhar Nagar, Jaipur (India) 3Patho Care & Research Centre, Raja Park, Jaipur (India)

*Corresponding Author:
Dr. Akshay Mangal
Neurocare Hospital, 1/611, Vidhyadhar Nagar, Jaipur (India)

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract
Pituicytoma is a rare tumor of the sellar-suprasellar regions, arising from the pituicytes, which are specialized glial cells in the neurohypophysis and infundibulum. Due to its rarity, ambiguity persists over the diagnosis, management and prognosis of pituicytoma. It typically presents with dysfunction of the optic nerve and pituitary. The radiological characteristics are nonspecific; diagnosis is typically made on the basis of histopathological results. We present a case report of a 23-year-old female with a history of decreased vision in both eyes and amenorrhea who was found to have a sellar-suprasellar mass. The tentative diagnosis was placed as a functional pituitary macroadenoma. The patient underwent craniotomy and subtotal excision of tumor. On histopathology, the lesion consisted of elongated and plump tumor cells that were arranged in a fascicular or stororiform pattern and were positive for S-100 protein and focally positive for GFAP. The final histological diagnosis was pituicytoma.

Keywords: Neurohypophysis, Pituicyte, Pituicytoma

INTRODUCTION
Pituicytoma is a rare tumor of the sellar and suprasellar regions, originating from specialized glial cells in the neurohypophysis and infundibulum [1]. The tumor is slow growing and benign, and histologically corresponds to WHO grade I [2, 3]. Only about 90-100 cases have been reported since it was first described in 1955[4]. Due to its rarity, the clinical manifestations, radiological characteristics, histopathological features and prognoses have yet to be fully elucidated. Pituicytoma is typically challenging to distinguish from other sellar and suprasellar lesions, including granular cell tumor, pituitary adenoma, pilocytic astrocytoma and lymphocytic hypophysitis [2, 3]. Surgical treatment may be challenging owing to hypervascularity of the tumor. We report a case of histopathologically diagnosed pituicytoma in a 23-year-old female.

Case Report
A 23-year-old female presented to Neurocare Hospital, Jaipur in November 2020 with a two month history of decreased vision in both eyes and four month history of amenorrhea. The physical examination revealed bilateral finger counting at one foot. NCCT brain revealed a 28x27mm heterogeneous mass in the sellar-suprasellar region showing lateral extension towards bilateral cavernous sinus and mass effect with an intratumoral necrotic component, possibility of pituitary macroadenoma. MRI brain revealed a contrast enhanced 34x32x28mm suprasellar mass with heterogeneous signal intensity showing lateral extension into the bilateral cavernous sinuses and temporal fossa with surrounding perilesional edema. [Fig. 1] Subsequent investigations revealed increased prolactin, increased TSH and decreased...
cortisol values. A preoperative diagnosis of functional pituitary macroadenoma was determined.

Examinations of the anterior segments of the eyes and fundus were normal. A further ophthalmologic examination revealed binocular ametropia with bitemporal visual field defect. No further neurological abnormalities were identified.

A craniotomy was performed via a right pterional approach. Intraoperatively, a hard grey-white solid mass was observed. The tumor was hypervascular. The tumor was gradually resected and a sub total resection was performed owing to the extension into the cavernous sinuses.

Diagnosis of pituicytoma was subsequently confirmed following histopathological examination which revealed a spindle cell whorl forming tumor with mitotic activity. [Fig. 2] On immunohistochemical staining, the tumor cells were positive for S-100 & glial fibrillary acidic protein (GFAP). [Fig. 3] TTF-1 was predominantly negative with very few cell positivity.

Synaptophysin, Prolactin, EMA, NF, P53 & CD68 were negative in tumor cells. The positive expression rate for antigen Ki-67 was more than three percent.

Post-operatively the patient’s visual acuity and visual field improved. The patient went into Diabetes Insipidus and developed hypernatremia which was resolved in seven days with adequate fluid management. The patient’s post-operative hormonal panel was suggestive of hypopituitarism and was therefore started on steroids and thyroid replacement therapy. The patient was referred to a radiation oncologist for further management of the residual tumor.

Discussion

Pituicytomas are rare, primary tumors originating from the so-called pituicytes in neurohypophysis and pituitary stalk [2]. Pituicytes are glial cells that support the large axons of vasopressin and oxytocin producing hypothalamic neurons. The first case of this type of glioma was identified in the posterior lobe of the pituitary gland and was described by Scothorne in 1955[4]. Brat et al. reported nine cases of low-grade glioma of the neurohypophysis in 2000, for which the term pituicytoma was proposed [2]. Pituicytoma was previously regarded as a condition with a wide clinical spectrum, including pituitary astrocytoma, posterior lobe glioma, choristoma and infundibuloma in the sellar and suprasellar regions. However, the tumor was named as a separate entity in the 2007 WHO classification of CNS tumors. [3]

Pituicytomas occur predominantly in adults with a male: female ratio of 1.3:1[5]. The most common reported symptoms of pituicytoma are vision & visual field disorders, headache and hypopituitarism. The clinical symptoms are typically attributable to the local effects of tumor, and therefore, depend on the tumor size and location. The duration of symptoms prior to the diagnosis range from a few months to several years. [6, 7] Pituicytomas arise along the distribution of the neurohypophysis, including the infundibulum and posterior pituitary. Accordingly they may be located within the sellar, the suprasellar region or both. The imaging characteristics of pituicytomas are nonspecific. On MRI examination, pituicytomas commonly present as well-defined, solid, round or oval masses in the sellar region with or without suprasellar extension.

The tumors usually appear hypointense-isointense on T1, low-moderately hyperintense on T2 and with homogenous or heterogeneous contrast enhancement [8]. The differential diagnoses include pituitary adenoma, meningioma, craniopharyngioma, granular cell tumor and pilocytic astrocytoma.

Surgical resection is the preferred treatment for pituicytoma with an extremely low recurrence rate following complete resection. Majority of pituicytomas are well demarcated and of a benign nature. Hypervascularity is a common intraoperative challenge, it can hinder the success of gross total resection. Current surgical approaches include the frontotemporal craniotomy and the transsphenoidal approach. The most common post-operative complications include diabetes insipidus, hypopituitarism, visual impairment and hypothyroidism [9]. The accurate diagnosis of pituicytoma continues to depend on histopathological evidence.

Microscopically, pituicytomas are composed of round to spindle-shaped cells with a fascicular or storiform growth pattern. The tumor cells have abundant eosinophilic cytoplasm and a rich capillary network is visible. Tumor cell nuclei are round to oval with usually no evidence of atypia or mitotic figures. On
IHC they are usually positive for S-100 and vimentin protein, show nuclear staining for TTF-1, negative or low–moderately positive for GFAP, and negative for EMA, Syn, chromogranin, cytokeratin and neurofilament protein. They usually have a low proliferation index indicating that pituicytomas are consistently benign [10]. The recurrence interval following subtotal resection is usually long and no instances of malignant transformation or CSF dissemination has been reported.

**Conclusion**

Pituicytoma is an extremely rare tumor of the neurohypophysis. Since the radiological characteristics and clinical manifestations are nonspecific; they are liable to be initially misdiagnosed. The definitive diagnosis depends on pathological examination. The surgical resection should be designed for optimal functional preservation. Considering local recurrence following subtotal resection, postoperative radiotherapy should be recommended in patients where gross total resection is not feasible. Even for those patients undergoing complete tumor resection, a close MRI follow-up is essential.

**References**


Images

Fig 1 Initial MRI T2 sagittal image showing a heterogeneous intensity pituitary mass

Fig 2 Histopathology slide showing spindle cell whorl forming tumor with mitotic activity

Fig 3 Immunohistochemical staining showing A) S-100 positivity B) GFAP positivity