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Case Report

Pituicytoma: A rare tumor of the sella. A case report and review of literature for diagnosis and management

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ABSTRACT

Background: Pituicytomas are rare tumors of the pituitary gland derived from the ependymal cells and line the pituitary stalk and posterior lobe. These tumors are located in the vulnerable regions of the brain: Either in the sellar or suprasellar area. The location marks the difference in the clinical features of the tumor. Here, we report a case of histopathologically diagnosed pituicytoma of the sellar region. Literature is also reviewed and discussed to gain a better understanding of this rare disease.

Case Description: A 24-year-old female presented to the outpatient department with complaints of headache, diplopia, dizziness, and decreased vision in the right eye for 6 months. Computed tomography scan brain without contrast showed a well-defined hyperdense lesion in the sella without associated bony erosion. Her magnetic resonance imaging showed well defined rounded lesion in the pituitary fossa which was isointense on T1-weighted image and hyperintense on T2-weighted images. A presumptive diagnosis of pituitary adenoma was made. She underwent endoscopic endonasal transsphenoidal resection of pituitary mass. Intraoperatively, normal pituitary gland was visualized and there was a grayish-green-colored, jelly like tumor which was pulled gently. On 9th postoperative day, she presented with cerebrospinal fluid (CSF) rhinorrhea. She underwent endoscopic CSF leak repair. Her histopathology was concluded to be Pituicytoma.

Conclusion: Pituicytoma is an uncommon diagnosis. The surgical aim is to completely excise the tumor which results in complete cure, but incomplete resection may be performed due to high vascularity of this tumor. In case of incomplete excision, recurrence is common and adjuvant radiotherapy may be administered.

Keywords: Ependymal tumor, Pituicytoma, Pituitary tumor, Sellar tumor, Transnasal endoscopic tumor resection

INTRODUCTION

Pituicytomas are rare tumors of the posterior part of the pituitary gland-neurohypophysis or infundibulum.[19] A total of 174 cases of pituicytoma were reported from 1994 to December 2021.[5] Fewer cases of pituicytoma meeting World Health Organization (WHO) criteria have been reported.[17]

Embryologically, neurohypophysis is derived from the lower part of diencephalon and continues with the hypothalamus. The hypophysis is surrounded by these glial cells known as pituicytes.[21]

These are derived from the ependymal cells and line the pituitary stalk and posterior lobe.[21] Pituicytes function to regulate hormone release of oxytocin and vasopressin/Anti-diuretic

hormone (ADH) which are predominant hormones of the posterior lobe of the pituitary gland. These tumors are located in the vulnerable regions of the brain: either in the sellar or suprasellar area. The location marks the difference in the clinical features of the tumor. Tumor located in the sellar area tends to exhibit endocrine dysfunction and those present in the suprasellar region, presents with structural compression symptoms such as headache, vomiting, and visual disturbances.[4,22]

Here, we report a case of histopathologically diagnosed pituicytoma of the sellar region. Literature is also reviewed and discussed to gain a better understanding of this rare disease.

CASE PRESENTATION

A 24-year-old female presented to the outpatient department with complaints of headache, diplopia, dizziness, and decreased vision in the right eye for 6 months. Due to these complaints, she even fell multiple times and injured herself. She did not report any episodes of fits, altered level of consciousness, nausea vomiting, or any history of trauma. On examination, she was a young female, oriented to time, place, and person. Her cranial nerves' examination was unremarkable. The vision in the right eye was reduced to finger counting. The vision in the left eye was also similar. However, extraocular movements were normal bilaterally. She had normal tone, bulk, and power in all four limbs. Her sensations were also intact in all four limbs. She did not have any cerebellar signs. Her hormonal status was normal. Computed tomography (CT) scan brain without contrast showed a well-defined hyperdense lesion in the sella without associated bony erosion. Her magnetic resonance imaging (MRI) was showed well defined rounded lesion in the pituitary fossa which was isointense on T1-weighted image and hyperintense on T2-weighted images [Figure 1]. In postcontrast sagittal image, it showed thick peripheral enhancement [Figure 2].

A presumptive diagnosis of pituitary adenoma was made. She underwent endoscopic endonasal transsphenoidal resection of pituitary mass. Intraoperatively, normal pituitary gland was visualized and there was a grayish-green-colored, jelly like tumor which was pulled gently. During surgery, diaphragma sella was damaged and there was cerebrospinal fluid (CSF) leak. The sella was packed with fat and fascia which was harvested from abdomen. This was further strengthened with bone and nasoseptal flap. Postoperatively, she developed panhypopituitarism.

On 9th postoperative day, she presented with CSF rhinorrhea. She underwent endoscopic CSF leak repair with fat, flap, bone, and Hadad flap. Fibrin glue was also applied. Nasal pack was inserted.

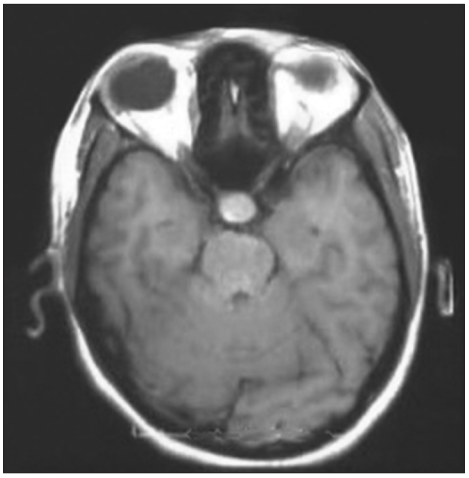

Figure 1: Magnetic resonance imaging T2-weighted image showing well-defined rounded lesion in the pituitary fossa.

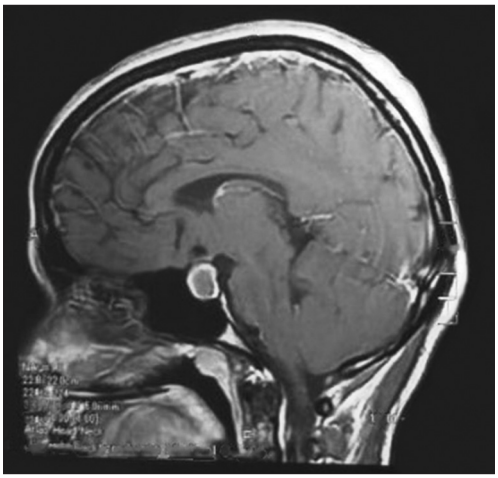

Figure 2: Magnetic resonance imaging T1 post contrast sagittal view showing thick peripheral enhancement.

Unfortunately, she developed severe headache, altered sensorium, and fever 2 days later and meningitis was suspected. She was managed with antibiotics. She gradually improved but reported CSF leak again at 5th post-CSF leak repair day. For this, a lumbar drain was inserted. The lumbar drain was kept till 6th postprocedure day and the CSF leak was resolved. She was observed for one more day. Her symptoms improved and she was discharged and her nasal pack was removed on 8th postoperative day in outpatient department. On subsequent follow-up, she has recovered completely.

Her histopathology showed abundant hemorrhage and proteinaceous material along with spindle cells. This was concluded to be pituicytoma. It was glial fibrillary acidic protein (GFAP) negative and thyroid transcription factor 1 (TTF1) positive [Figure 3]. This pathology report was

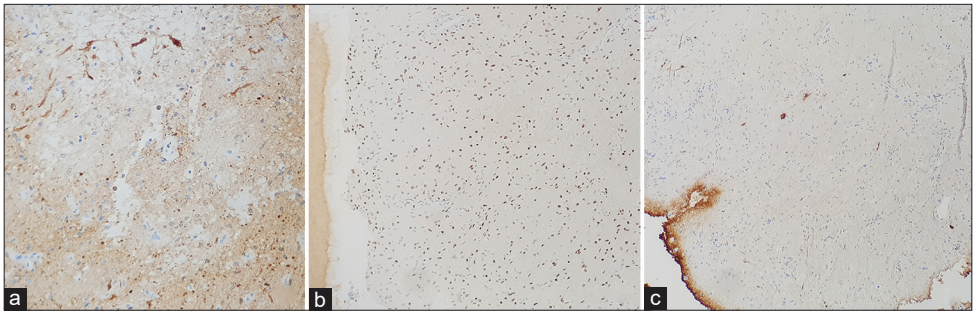

Figure 3: Tumor cells negative for S100 (a), show nuclear expression of thyroid transcription factor 1 (b), and lack glial fibrillary acidic protein staining (c).

reviewed by another pathologist who concurred with the findings.

On pathological examination, the sample showed a hypocellular lesion, composed of ovoid to spindle cells, arranged in short bundles and sheets [Figure 4]. Tumor cells were bland in appearance and showed no cytological atypia or mitoses. Cytoplasmic granularity or oncocytic changes were absent. The background was fibrillary. Immunohistochemically, nuclear expression of TTF1 was demonstrated, whereas GFAP, epithelial membrane antigen (EMA), and S100 were negative. In correlation with radiological findings, the final diagnosis of pituicytoma was established.

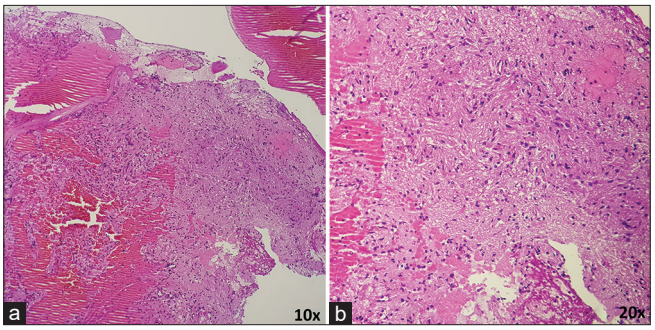

Figure 4: (a) ×10: Hypocellular lesion composed of bundles and aggregates of spindle to epitheloid cells, set in eosinophilic background. (b) ×20: High magnification showing cells with bland nuclei and eosinophilic cytoplasm. There is no significant cytological atypia or mitosis.

DISCUSSION

Adenomas arising from adenohypophysis are the most common pituitary tumors. Pituicytomas, germinomas, meningiomas, hamartomas, and craniopharyngiomas are categorized among posterior pituitary tumors originating from neurohypophysis.[1,22] Among these, pituicytomas further include granular cell tumors or choristomas and pilocytic astrocytomas which are often misdiagnosed and it is difficult to differentiate them from other brain neoplasms located in the same region.[7]

CNS tumors of glial origin are benign and were classified by the WHO in 2006 where it termed them pituicytoma differently from astrocytomas under low Grade-1 glial tumors.[13] This was revised in 2016, and these Grade-1 glial tumors are now classified as Group I of TTF1-expressing tumors.[10] Pituicytomas are noninfiltrative, solid, and well-defined tumors arising from posterior pituitary or its stalk and specifically comprise of glial cells known as pituicytes. These appear to be benign, slow-growing tumors which clinically present symptoms from mass effect. Immunohistochemical components; EMA, GFAP, S-100 protein, and vimentin further confirms the diagnosis of pituicytoma from other posterior tumors.[7]

A cohort study on 93 diagnosed cases of pituicytoma from 1958 to 2020 concluded the most favorable findings suggestive of the diagnosis includes: (1) presence of hypopituitarism, (2) CT showing isodense tumor without calcification, (3) MRI revealing a globular mass located in the suprasellar region separating the pituitary gland, and lastly (4) an isointense image on T1-weighted MRI due to marked vascular proliferation and intense contrast-enhancement.[14]

The histological criteria for diagnosis of pituicytoma was proposed by Brat *et al.* in 2000.[1] It is characterized by four main features: (1) having bipolar spindle cells with interlacing fascicles, (2) eosinophilic and homogenous cytoplasm with granulation or vacuolization, (3) oval nuclei with mild irregularity or atypia, and (4) nuclei with no mitotic activity.[1] These features fit well with the findings in our case. Another characteristic feature which is unique to neurohypophyseal architecture is the arrangement of glial cells around blood vessels in the posterior pituitary.[13,23,24] The histogenesis is further elaborated to its immunohistochemical characteristics which includes immunoreactivity for the glial markers vimentin, S-100,[24] and GFAP, TTF1[11,25] with occasional positivity of cytoplasmic EMA in favor of pituicytoma.[1] However, our case showed negative staining for GFAP, EMA, and S-100. Although, variable immunohistochemical expression is well known, S-100 negativity is unusual and may represent analytical limitation due to very low yield

of tumor cells in the biopsy sample.[1,15] Recently, positivity of vascular endothelial growth factor (VEGF) has been described which has diagnostic and therapeutic utility.[11]

The preoperative clinical presentation includes mass effect symptoms including visual impairment such as bitemporal hemianopsia, headache, endocrine disorders like hypopituitarism being the most common,[12,18] hyperprolactinemia, and diabetic insipidus which is often suggestive of pituitary stalk lesions.[25] In some cases, the patients are often asymptomatic and tumor gets detected incidentally.[12,18]

The mainstay of treatment for these tumors is surgical excision either by trans-sphenoidal or transcranial route.[23] The surgical approach varies depending on the location and extent of the tumor.[13] Although a benign tumor, pituicytoma faces certain challenges in the treatment strategies due to its vascularity and firmness of its content. Literature shows that incomplete resection sometimes results due to vascularity of tumor, and this is associated with recurrence.[16] To overcome recurrence with partial resection of the tumor, it is recommended to prescribe targeted therapy of the ligand of VEGF-R following surgery, for example, bevacizumab-VEGF antibody.[6] Similarly, somatostatin receptors analogues are also recommended. They undergo down regulation of protein biosynthesis and induce cell arrest which leads to apoptosis within the stroma of the tumor. It also affects the vascularization of the tumor.[3,8]

Gross total resection (GTR) is the cure for the disease based on a large series of cases reviewed in 2021.[23] It was observed that open transcranial was likely to result in more postoperative complications especially with visual function.[2] Partial resection of the tumor is only recommended for large sizes where GTR will most definitely damage the pituitary gland resulting in endocrinopathies. In such cases, residual tumor needs to be observed carefully. If tumor regrows, it is recommended to reoperate[23] or prescribe radiotherapy[9] to eliminate the tumor.[16]

Brat *et al.*, proved that complete excision of the tumor was the most significant predictor of recurrence.[1]

CONCLUSION

Pituicytoma remains an uncommon tumor in neurosurgical specimens. The tumor may be detected incidentally or may present with a spectrum of symptoms ranging from mass effect symptoms to endocrine abnormalities. The surgical aim is to completely excise the tumor which results in complete cure, but incomplete resection may be performed due to high vascularity of this tumor. In such cases, it is recommended to follow patient with MRI and offer either re-operation or radiotherapy if any recurrence is identified.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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