

Trans-nasal Trans-sphenoidal Endoscopic Resection of Spindle Cell Oncocytoma of Adenohypophysis: The First Case Report in a Child and a Review of Literature

Abstract

Spindle cell oncocytoma (SCO) is a rare tumor of adenohypophysis, arising from the sellar region. So far, about 35 cases of SCO in the sellar region have been reported. In this report, we present the first case of pediatric SCO and review the literature concerning the tumor origin, clinical presentations, radiological features, and treatment modalities. An 8-year-old male was referred to our clinic with progressive visual loss in the left eye and headache over the past 6 months. Cranial magnetic resonance imaging revealed a solid adenohypophysis mass with suprasellar extension, as well as compression and displacement of the optic chiasm. The patient underwent endoscopic trans-sphenoidal resection of the tumor. The tumor was diagnosed as SCO based on the histological study. He did not receive radiation therapy. The patient's condition remained stable, with no radiological recurrence in the past follow-up 2 years after the surgery.

Keywords: Adenohypophysis, sellar region, spindle cell oncocytoma, trans-sphenoidal

Introduction

Spindle cell oncocytoma (SCO) is a benign nonendocrine neoplasm of the sellar and suprasellar regions, which is rarely observed by neurosurgeons. It accounts for 0.1%–0.4% of all sellar tumors with no sex predilection.^[1,2] SCO was first reported by Roncaroli *et al.* in 2002.^[2] The tumor arises from the folliculostellate cells of adenohypophysis. In the 2007 classification of the World Health Organization (WHO) for the central nervous system (CNS) tumors, SCO in the neurohypophysis was described as a distinct diagnosis.^[1] This tumor has a slow growth pattern and shows clinical and radiological features similar to nonfunctional pituitary adenoma.^[3,4] To date, only 35 cases of pituitary oncocytoma have been reported in the literature.

Herein, we present the first case of pediatric pituitary oncocytoma and discuss the diagnostic and therapeutic aspects of this rare neoplasm.

Case Report

An 8-year-old male was referred to our clinic with progressive visual loss

in the left eye and headache over the past 6 months. On examination, the best-corrected visual acuity was 4 m in the right eye, and no light perception was observed in the left eye; also, the light reflex was absent in the left eye. Brain computed tomography scan showed an isodense mass in the sellar and suprasellar regions, measuring 19 mm × 30 mm, with a sphenoid sinus extension. In addition, cranial magnetic resonance imaging (MRI) revealed a solid adenohypophysis mass of 2 cm × 1.5 cm × 1 cm, with a significant suprasellar component, as well as a compressed and displaced optic chiasm, extended superiorly toward the floor into the third ventricle [Figure 1].

The pituitary function test revealed a prolactin (PRL) level of 96 ng/mL (normal: 2–17 ng/mL). Other laboratory findings were within the normal range for hormones; this finding was consistent with pituitary stalk compression by a nonfunctional tumor. However, in view of the clinical suspicion of macroprolactinoma, the PRL assay was repeated after 1:100 serum dilution and reported as 27.2 ng/mL.

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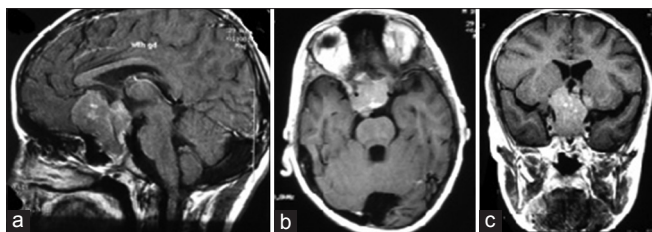


Figure 1: (a,b,c) post-contrast Magnetic resonance images (MRI) revealed an enhancing huge sellar-suprasellar mass with involvement of right cavernous sinus and optic nerve compression

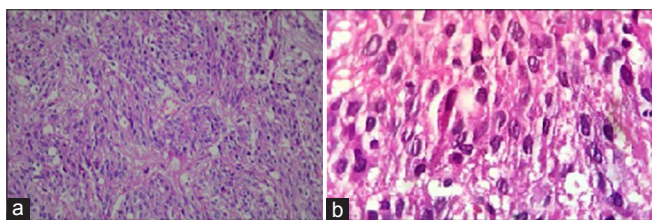


Figure 2: Photomicrograph of a hematoxylin and eosin (H&E) stained section of the tumor, showing interlacing fascicles of mono morph spindle cells and epithelioid cells. The cells had an abundant eosinophilic cytoplasm. necrosis, mitoses, or invasion was not observed. (a) Cells were spindle-shaped with round to ovoid nuclei and no mitotic activity (b)

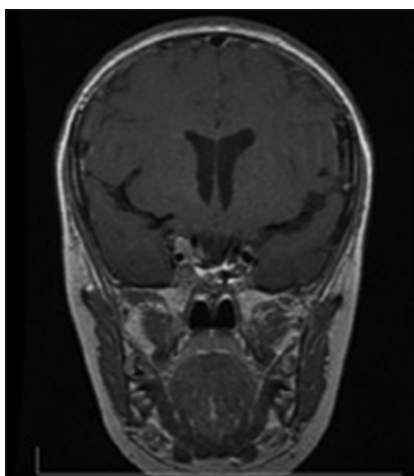


Figure 3: Post operative MRI show gross total resection of tumor with very small residue in right cavernous sinus

A tentative diagnosis for the lesion was a nonsecreting pituitary adenoma.

Considering the size of the tumor, positive visual signs, and observable stretching of the optic chiasm, it was decided to treat the tumor, using an extended endoscopic endonasal transtuberulum/transplanum approach. A binostril approach was adopted. A right-middle turbinectomy with resection of the posterior nasal septum, along with a wide sphenoidotomy, was carried out. The tuberculum sellae, posterior portion of the planum sphenoidale, and sellar floor were removed, and the tumor was completely resected.

Postoperatively, the patient developed panhypopituitarism, which was managed by hormonal replacement. Paraffin sections of the tumor revealed interlacing fascicles of

spindle cells and epithelioid cells. The cells had an abundant eosinophilic cytoplasm, with round or oval nuclei and inconspicuous nucleoli. The mitotic activity was inconspicuous, and necrosis was absent [Figure 2]. The immunohistochemical staining profile showed a diffuse immunoreaction with epithelial membrane antigen (EMA), Vimentin, and S-100. The final histopathological diagnosis was confirmed as pituitary oncocytoma. He did not receive radiation therapy. The patient did not present any clinical or radiological signs of progression on the past follow-up 2 years after the surgery [Figure 3].

Discussion

SCO is a rare slow-growing tumor, which mimics nonfunctional pituitary adenoma and other sellar lesions. According to the WHO classification, these tumors are Grade I tumors of CNS, originating from the anterior pituitary.^[5-7] A total of 35 cases of SCO were reported in 2003. These tumors usually affect adult patients with the mean age of 56 years at presentation.^[6-26] Nineteen out of 35 patients published in the literature were female, and 16 were male. The patients were in the age range of 26–80 years. Dahiya *et al.* reported the youngest case of SCO (a 26-year-old male patient).^[5] Our case is the first case presenting in a child.

The process of differential diagnosis can be difficult, as the clinical and neuroradiological features of SCO are not definitive.^[14] Patients with SCO may present with a wide range of neurological symptoms, such as headache, visual complaints, and panhypopituitarism. Among 35 patients with SCO, the visual loss was the most common clinical problem (21 patients), whereas 15 patients had panhypopituitarism, and 15 patients had headache. Syncope occurred in one patient, and three patients experienced weight loss.^[9,10,27] Furthermore, epistaxis was reported in one patient, and one patient presented with an altered level of consciousness.^[25]

Preoperative imaging studies are generally inconclusive and nonspecific for the lesion and do not differentiate SCO from pituitary adenomas.^[3] Recently, Hasiloglu *et al.* described the characteristic radiological findings for SCO, which includes hypointense foci and linear signal-void areas on T1-weighted and T2-weighted imaging. On dynamic contrast-enhanced MRI, these tumors showed intense contrast enhancement in the early stage of contrast administration (Hasiloglu's *et al.* sign),^[28] as reported in our case.

Cavernous sinus invasion or clival and sellar floor destruction has been rarely reported.^[7] The definite diagnosis of oncocytoma is best achieved postoperatively through histopathological and immunohistochemical studies. The classic immunohistochemical feature of these tumors is the coexpression of EMA, Vimentin, and S-100, along with galectin-3.^[1] The generally accepted

management plan includes complete tumor resection whenever possible. During surgical resection, tissue texture can vary from soft and creamy to exceptionally adherent to the surrounding structures. In some cases, total resection of tumor is difficult due to tumor hypervascularity, causing significant intraoperative bleeding and increasing the risk of injury to the adjacent neurological structures due to tumor adhesion.

In cases with incomplete resection of the lesion, adjuvant postoperative radiotherapy has been suggested, although tumor recurrence after radiotherapy has been also reported. There is little evidence in the literature regarding the sensitivity of SCO to radiotherapy. Therefore, no recommendations can be made at this time regarding the effectiveness of adjuvant radiotherapy for SCO. Although oncocytoma is a benign tumor, only one case of malignant transition has been reported so far.^[28,29] Accordingly, long-term close follow-ups, along with continuous regular imaging studies, are necessary for these patients.

Conclusion

We present the case of an 8-year-old male, presenting with headache and visual loss. To the best of our knowledge, this is the first case of pediatric SCO. A combination of histopathological, immunohistochemical, and ultrastructural examinations is required to reach a definite diagnosis. In the absence of any definitive morphological or predictive prognostic factors, extended regular follow-ups, along with an aggressive treatment protocol, are essential.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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