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Case Report

Sellar Embryonal Tumor: A Case Report and Review of the Literature

Abstract

Primitive neuroectodermal tumors (PNETs) are aggressive, poorly differentiated tumors in children and young adults. However, the embryonal tumor group did not include the central nervous system (CNS) PNET title and ependymoblastoma subtitle in the 2016 World Health Organization CNS tumor classification. Here, we report the case of a 6-year-old boy with a sellar embryonal tumor and present a review of the related literature. To the best of our knowledge, this is the first case of an endoscopically operated sellar embryonal tumor in the pediatric age group.

Keywords: *Embryonal tumor, endoscopy, sella*

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Introduction

Primitive neuroectodermal tumors (PNETs) are aggressive, poorly differentiated tumors in children and young adults.[1] Supratentorial PNETs represent <1% of primary central nervous system (CNS) tumors.[2] The embryonal tumor group includes CNS PNETs, which are of the following four types: CNS neuroblastoma, CNS ganglioneuroblastoma, medulloepithelioma, and ependymoblastoma, according to the 2007 World Health Organization (WHO) CNS tumor classification.[3] However, the embryonal tumor group did not include the CNS PNET title and ependymoblastoma subtitle in the 2016 WHO CNS tumor classification.[4]

Sellar embryonal tumors are exceedingly rare, and only 13 sellar PNET cases have been reported in the literature till date [Table 1]. Here, we report the case of a 6-year-old boy with a sellar embryonal tumor and present a review of the related literature. To the best of our knowledge, this is the first case of an endoscopically operated sellar embryonal tumor in the pediatric age group.

Case Report

A previously healthy 6-year-old boy was admitted to our clinic with complaints of headache and weight gain. Neuro-ophthalmological examination revealed best corrected visual acuity of 20/20 in both eyes, normal color vision, and no third cranial nerve palsy. The patient had no electrolyte or hormonal imbalance. A cranial computed tomography scan revealed a low-density lesion with irregular borders and slight hyperdense areas, which might have resulted from calcification [Figure 1]. Brain magnetic resonance imaging (MRI) showed a sellar mass with suprasellar extension. The lesion showed low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and intense enhancement after gadolinium administration. The lesion filled the suprasellar cistern and spread to the interpeduncular cistern and third ventricle. The tumor size was 45 mm × 30 mm × 25 mm [Figure 2]. Differential diagnosis of the lesion included craniopharyngioma, Rathke's cleft cyst, arachnoid cysts, chiasmatic/hypothalamic glioma, germ cell tumors, histiocytosis X, hypothalamic hamartoma, and metastatic lesions. The clinical and neuroradiological findings supported a diagnosis of germ cell tumors. Surgery was presented as the best treatment option to the parents of the child because the chance of expected symptomatic improvement was estimated to be higher than the chance of surgery-related morbidity. The patient underwent surgical removal of the lesion after his parents provided informed consent according to our institutional code of ethics. The surgical procedure involved the use of a 0° or 90° endoscope. After induction of general anesthesia and endotracheal

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Table 1: All reported cases of sellar peripheral neuroectodermal tumors till date

Author, year	Age (year)/Sex	Location	Pathology	Management	Endocrine status (pre-/post-operative)
Sarwar, 1979[5]	31/female	Sella and parasella	Esthesioneuroblastoma	Surgery, radiotherapy	Not available
Tamura *et al*., 1982[6]	16/male	Sella and parasella	Neuroblastoma	Surgery, radiotherapy	Not available
Lach *et al*., 1996[7]	40/female	Sella	Neuroblastoma	Transcranial surgery	PRL: 68 μg/L/ normal-diabetes insipidus
Roy *et al*., 2000[8]	44/female	Sella and suprasella	Esthesioneuroblastoma	Surgery, radiotherapy	PRL: 291 μg/L/ panhypopituitarism postradiotherapy
Pang *et al*., 2001[9]	2/male	Sella and suprasella	Medulloepithelioma	Transcranial surgery, chemotherapy	Hypothyroidism/not available
Mariani *et al*., 2004[10]	35/female	Sella and suprasella	Neuroblastoma	Transsphenoidal surgery	PRL: 139 μg/L/ normal- diabetes insipidus
Idrees *et al*., 2005[1]	46/male	Cavernous sinus and sella	Ewing's sarcoma	Transsphenoidal tumor biopsy, transcranial surgery, chemotherapy	Not available/not available
Sajko *et al*., 2005[20]	57/female	Sella and suprasella	Neuroblastoma	Transsphenoidal surgery, radiotherapy	PRL: 92 μg/L/not available
Oyama *et al*., 2005[12]	33/male	Sella	Neuroblastoma	Transsphenoidal surgery	Panhypopituitarism/ panhypopituitarism
Schmalisch *et al*., 2009[13]	43/female	Sella and suprasella	Neuroblastoma	Transcranial surgery, radiotherapy	PRL: 24 μg/L SIADH/normal–SIADH
Lin *et al*., 2009[11]	40/male	Sella	Neuroblastoma	Endoscopic transsphenoidal surgery, radiotherapy	Normal/normal
Radotra *et al*., 2010[14]	29/male	Sella and suprasella	Neuroblastoma	Transcranial surgery, radiotherapy	Hypopituitarism hyponatremia/ normal-hyponatremia
Dupuy *et al*., 2012[15]	44/female	Sella	Neuroblastoma	Transsphenoidal surgery	PRL: 27 μg/L SIADH/normal–SIADH
Present case, 2016	6/male	Sella and suprasella	Embryonal tumor, NOS	Endoscopic transsphenoidal surgery, radiotherapy, chemotherapy	Normal/transient diabetes insipidus

NOS – Not otherwise specified; PRL – Prolaktin; SIADH – Syndrome of inappropriate antidiuretic hormone secretion

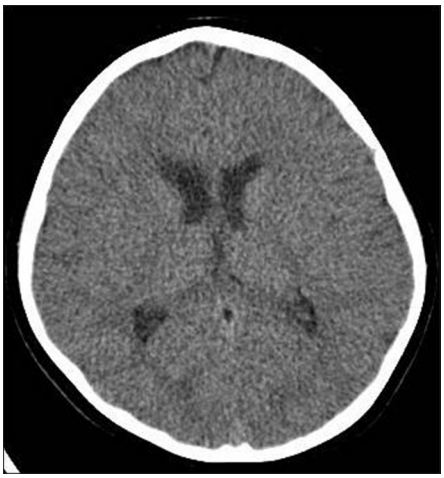

Figure 1: Preoperative computed tomography scan. In the axial section, the lesion appears to be located in the suprasellar region with a hypodense signal and slight hyperdense areas, which might have resulted from calcification

intubation, venous and arterial accesses were established. The patient's head was positioned using a Mayfield 3-point fixation system and was coregistered to a neuronavigation system. The sphenoid ostium was identified and was superior to the choana. A nasoseptal flap was raised. The sphenoid ostium was enlarged using a Kerrison rongeur. The sphenoid rostrum and posterior vomer were removed to open the whole anterior wall of the sphenoid sinus. The medial opticocarotid recess, carotid protuberance, sellar face, clival recess, and strut of bone over the superior intercavernous sinus were recognized. A micro-Doppler was used during the procedure. Sphenoidotomy was widened to include the lateral recess of the sphenoid extending lateral to the carotid canal. The exposure was then rostrally extended to expose the posterior cells of the ethmoid sinus and the planum–tuberculum junction. By reducing the floor of the sphenoid, a greater caudal-to-rostral trajectory into the suprasellar space

was created. The sphenoid sinus mucosa was removed. A combination of microcurettes and microdissectors was used to remove the tumor within and above the sella, and biopsy was performed [Figure 3]. Hypernatremia occurred on the 1st postoperative day. The patient was diagnosed with diabetes insipidus (DI), and treatment with desmopressin was initiated. His DI was transient. No visual field loss or rhinorrhea occurred in the postoperative period. Detailed histological examination confirmed CNS PNET according to the 2007 WHO classification [Figure 4]. Subsequently, in 2016, the revised WHO classification was published and the new diagnosis was embryonal tumor, not otherwise specified (NOS). We performed conventional radiotherapy and adjuvant chemotherapy postoperatively, and it was effective for tumor eradication. There was no tumor residue or recurrence on follow-up MRI performed in the 2nd postoperative year [Figure 5]. We obtained informed consent of the parents to be enrolled and have their data published. Our institute approved the study and publication.

Discussion

PNETs are aggressive, poorly differentiated tumors in children and young adults. PNETs show a peak incidence in the second decade and have male predominance.[1] CNS PNET included CNS neuroblastoma, CNS ganglioneuroblastoma, medulloepithelioma, and ependymoblastoma in 2007 WHO classification.[3] However, WHO removed the term CNS PNET from the 2016 CNS tumor classification.[4] Embryonal tumors now include medulloblastomas (genetically defined, histologically defined, and NOS), embryonal tumors (with multilayered rosettes C19MC-altered and multilayered rosettes NOS), medulloepithelioma, CNS neuroblastoma, CNS ganglioneuroblastoma, CNS embryonal tumors NOS, atypical teratoid/rhabdoid tumors, and CNS embryonal tumors with rhabdoid features.[4] Ewing's sarcoma is classified under mesenchymal and nonmeningothelial tumors as Ewing sarcoma/PNET.[4]

Intrasellar neuronal neoplasms are extremely rare and generally well-differentiated gangliocytomas or ganglioneuromas.[16] Primary intracranial neuroblastomas are extremely rare[7] and generally arise in the

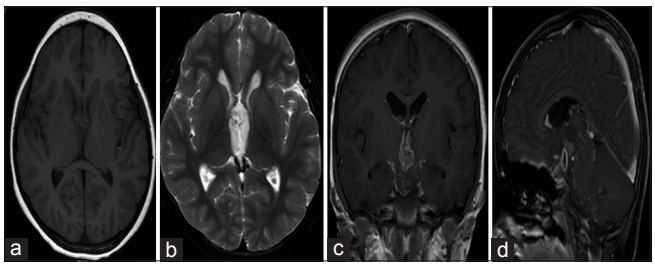

Figure 2: Preoperative magnetic resonance images. The lesion shows low signal intensity in T1-weighted axial images (a), high signal intensity in T2-weighted images (b), and intense enhancement after gadolinium administration on coronal (c) and sagittal (d) sections

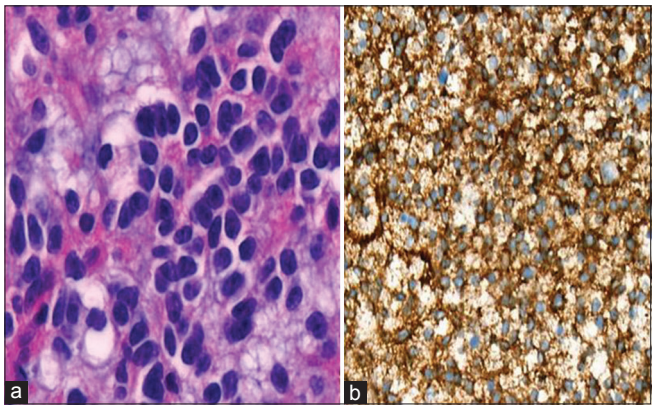

Figure 4: Histological examination demonstrates a cellular tumor involving small embryonal cells with oval irregular dark nuclei and narrow cytoplasm (a). In addition, slightly molded embryonal cells are noted. Mitotic activity is 10 mitoses per 10 high-power fields. There are no Homer-Wright rosettes, necrosis, or vascular endothelial proliferation. In immunohistochemical studies, the tumor cells demonstrate diffuse cytoplasmic synaptophysin (b) and Glial fibrillary acidic protein expression. In addition, some of the cells show S100 staining

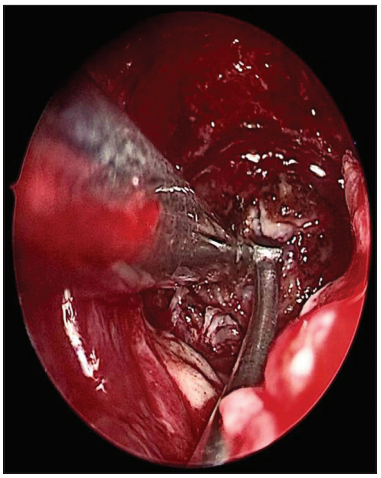

Figure 3: Image captured from the intraoperative video. Biopsy performed using the transsphenoidal endoscopic approach

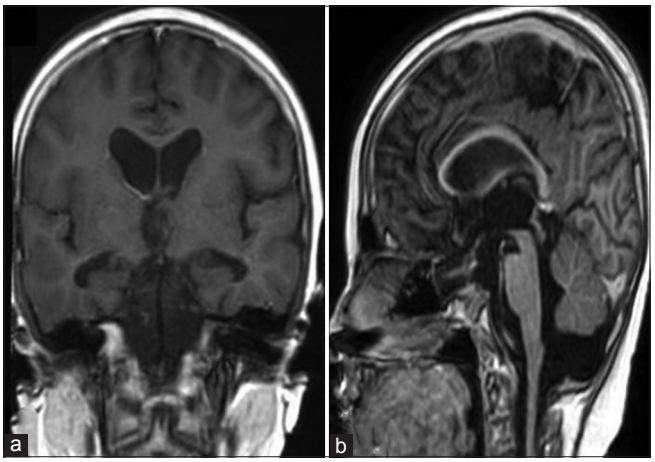

Figure 5: Postoperative magnetic resonance images obtained 2 years after surgery. T1-weighted coronal (a) and sagittal (b) images obtained after gadolinium enhancement show that there is no further pathological enhancement

supratentorial parenchyma or paraventricular region.[12] Sellar neuroblastomas are even rarer; hence, the decision to diagnose these tumors based on hematoxylin and eosin staining results alone can cause misdiagnosis of pituitary adenoma and recurrent surgical interventions. The histological diagnosis should be confirmed with immunohistochemical and ultrastructural studies if the tumor recurs within only 16 months.[12] Esthesioneuroblastomas were first described as originating from the neural crest with similarities to neuroblastomas of the sympathetic region and are known as olfactory neuroblastomas.[17] Primary cerebral neuroblastomas tend to occur in children, and olfactory neuroblastomas have a bimodal peak in the second and fifth decades of life.[18] On the other hand, primary spinal PNETs are uncommon tumors, and 95 cases have been reported till date. Intracranial metastases were detected in 11 of the 95 primary spinal PNET cases.[19]

We searched the English literature and identified 13 cases of sellar embryonal tumors [Table 1]. The mean age of the patients was 34.3 years, and the male:female ratio was 6:7. The management was primarily surgical resection.[15] All patients were surgically treated through transcranial or endoscopic approaches. Transcranial surgery was preferred in the pediatric age group.[6,9] Endoscopic transsphenoidal surgery has been performed in only one adult patient.[11] The present case is the second sellar embryonal tumor case that has been endoscopically operated. To the best of our knowledge, our case is the first case of an endoscopically operated sellar embryonal tumor in the pediatric age group in the literature.

Postoperative radiotherapy was performed in seven of the previous cases. As neuroblastomas are radiosensitive tumors, adjuvant radiotherapy has been suggested as initial therapy by several authors.[5,8,11,13] Oyama *et al*. reported second recurrence in a patient who received gamma knife radiosurgery.[12] We performed radiotherapy and chemotherapy in our patient; however, there is no consensus on chemotherapy.[15] Although some authors suggested adjuvant chemotherapy in patients with subtotal resection or recurrence of solid neuroblastomas, adjuvant chemotherapy has not been recommended for primary sellar neuroblastomas.[15]

The histological diagnoses of the published sellar PNET tumors were as follows: Nine cases of neuroblastoma, two of esthesioneuroblastoma, one of Ewing's sarcoma, and one of medulloepithelioma [Table 1]. Our patient was diagnosed with embryonal tumor, NOS.

The normalization of hyperprolactinemia was marked by postoperative courses in all cases. Postradiotherapy, panhypopituitarism occurred in only one patient.[9] Preoperative syndrome of inappropriate antidiuretic hormone secretion persisted during the postoperative period in two cases.[1,13] DI occurred in two cases during the postoperative period.[7,10] Transient DI occurred in the present case.

Conclusion

Embryonal tumors are extremely rare in the sellar region. After biopsy and histological diagnosis, the tumor can be treated with adjuvant radio/chemotherapy. In addition, an endoscopic approach might be appropriate in such patients. Neurosurgeons should consider an embryonal tumor in the differential diagnosis of a sellar mass.

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Conflicts of interest

There are no conflicts of interest.

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