Synchronous Clival Chordoma and Sellar-Suprasellar Germinoma in a Pediatric Patient: Case Report and Review of Literature

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Abstract

Pediatric brain tumors are the leading cause of cancer in children. While children more likely present with craniopharyngiomas, germinomas or optic gliomas, synchronous intracranial lesions are highly uncommon. Here, we describe the case of a 10-year-old female who presented with central Diabetes Insipidus (DI). MRI revealed enhancing sellar/suprasellar mass. The patient was taken to the operating room for endonasal resection. Pathology was consistent with sellar/suprasellar germinoma and contiguous clival chordoma. To the best of our knowledge this is the first reported case of association between these two rare tumors.

Keywords

Sellar/Suprasellar germinoma, Clival chordoma

Introduction

Though occurring at one-tenth the frequency of adult brain tumors, pediatric brain tumors are the leading cause of cancer related morbidity and mortality in children [1]. Synchronous intracranial lesions are highly unlikely, particularly those of separate, distinct cell types. Patients most at risk are those with neurocutaneous disorders, prior history of tumors, or history of chemotherapy and radiation.

Pediatric suprasellar tumors are distinct from their adult counterparts. While adults are more likely to have meningiomas or pituitary adenomas, children are more likely to present with craniopharyngiomas, germinomas, or optic gliomas [2]. Pathology determines appropriate treatment, and endonasal approaches, when appropriate; limit morbidity associated with surgery [3].

Case Report

A Ten-year-old female with history of cognitive delay, autism spectrum disorder and attention deficit hyperactivity disorder presented to her primary care provider with polydipsia, polyuria, enuresis, and 20 lb weight loss in the last year. Review of systems was positive for headache and daily vomiting. Past surgical history, social history, and family history were non-contributory. Vital parameters demonstrated tachycardia and hypertension, and the rest of her exam was unremarkable. Complete Blood Count (CBC) and urinalysis demonstrated no abnormalities. Complete Metabolic Panel (CMP) was significant for hypernatremia, hyperchloremia, and non-anion gap acidosis. Serum osmolality was high. The patient was directly admitted for work up of Diabetes Insipidus (DI), with renal, and endocrinology consults.

As an inpatient, ddAVP test demonstrated central etiology for her DI and hormonal work up showed panhypopituitarism. Subsequent MRI brain with and with-
out contrast revealed an enhancing sellar/suprasellar mass with a possible cystic non-enhancing clival component measuring 2.2 × 1.2 × 1.7 cm (Figure 1). Serum beta-HCG and alpha-FP were found to be within normal limits. CSF beta-HCG was 22 while CSF alpha-FP was < 0.5. Differential diagnosis included craniopharyngioma, germ cell tumor, atypical pituitary adenoma, or Langerhans histiocytosis. The possibility of two separate tumors was considered, although thought to be less likely. Ophthalmologic exam demonstrated no visual field defects or optic disc edema/pallor.

The patient was medically optimized and taken to the operating room by otolaryngology and neurosurgery for endoscopic endonasal, transsphenoidal approach and resection. A lumbar drain was placed prior to surgery. Intraoperatively, the sphenoid sinus was opened widely and the clivus was drilled between the carotids. An extradural, brown mass was noted and multiple biopsies demonstrated chordoma on pathologic section (Figure 2). Image guidance was used to aid in further surgery of this area and the tumor was resected without a Cerebrospinal Fluid (CSF) leak. The bone overlying the sella turcica and suprasellar area at the level of the tuberculum sellae was removed. A thin portion of pituitary gland was noted, and this was dissected until a soft and friable tumor was encountered. The suprasellar dura was opened and the tumor was not to be tracking toward the third ventricle, thus giving the tumor a classification of Type IIIa [4]. Frozen and later permanent section was consistent with germinoma (Figure 3). Tumor was resected to the third ventricle, where visualization became more difficult. At this point, given the patient’s age and pathology, the decision was made to conclude the resection. The patient’s skull base defect was reconstructed with inlay and onlay Dura Matrix (Stryker Corporation, Kalamazoo, Michigan), anasoseptal flap, and resorbable packing.

Her lumbar drain was removed on Post-Operative Day (POD) 7 after no concern for CSF leak. Radiation

Figure 1: Preoperative MRI Head, T1 with contrast, sagittal view demonstrating an enhancing sellar/suprasellar mass with a possible cystic non-enhancing clival component measuring 2.2 × 1.2 × 1.7 cm.

Figure 2: The clival lesion consists of cells with bubbly, vacuolated cytoplasm. These cells showed strong cytokeratin expression (not shown). (Hematoxylin and eosin, ×400 original magnification).
noid sinus. Her drain was removed on POD 5, and there
was no evidence of recurrent leak.

Twenty months after her first surgery, surveillance
imaging demonstrated recurrence of her clivalchordoma. Revision endonasal endoscopic resection of the
clival lesion was performed by extending the previous
bony defect and further resection in this area. Postop-
erative recovery was uneventful.

Thirty-two months she again was reported to have
a progressive enlargement of the clival mass on serial
MRSIs. The patient’s records were reviewed by an out-
side Proton Therapy Center and was determined to not
be a candidate for proton beam radiation and surgery
was recommended. A third surgery was performed at
an outside institution thirty-five months after initial sur-
gery. An abdominal fat graft was harvested for repair of
the skull base defect and abdominal JP was left in place
which was discontinued on POD2. On POD7 she was
taken back for removal of the Merocels. Her postopera-
tive course was complicated by worsening of her diabe-
tes insibidus, requiring increasing her standing DDAVP
dosing. She was discharged home on POD8.

Discussion

We discuss the case of a 10-year-old female with a
sellar/suprasellar germinoma contiguous with a clival-
chordoma. Germinomas represent 1% of all intracranial
tumors, and of these, 20% are suprasellar [5]. They
typically develop from undifferentiated embryonal cells.
These classically present with the triad of diabetes in-
sipidus, panhypopituitarism, and visual changes [6]. Our
patient did not exhibit the latter, however given her age
and cognitive deficit, it is possible she could not articu-

Figure 3: An intra-operative smear from the sellar/suprasellar mass confirms sheets of large atypical polygonal cells mixed
with numerous small benign lymphocytes. (Hematoxylin and eosin, ×400 original magnification).

Figure 4: Postoperative MRI Head, T1 with contrast, sagittal
view demonstrating fat packing of the clival operative cavity
and no discrete residual or recurrent tumor identified.
late these issues. Typically, treatment includes surgery, but these tumors are also chemo and radiation sensitive [7].

Chordomas develop at the ends of the notochord remnant near the sacrum and clivus. Although a preponderance in the sacrum in the elderly, chordomas mostly affects the skull base in the pediatric population [8]. Though histologically low grade, they are locally invasive, highly recurrent and may metastasize [8]. Pediatric chordomas account for 5% of all chordomas, and typically present with atypical features and more aggressive pathology [8-10]. There are relatively low rates of complete resections and local recurrence may occur in up to 2/3rds of patients [8]. As a result, post-operative radiation is a mainstay of therapy [8].

Both germinomas and chordomas are regarded as congenital or vestigial tumors. Whether clival or spinal in location, there has only been few reported cases in the literature of chordomas in association with other neoplasms. There was a report of a clival chordoma found in association with a craniopharyngioma [11]. This was resected via open approach with resulting complete left cranial nerve III palsy. A more recent case, demonstrated sacrococcygeal chordoma synchronous with myxopapillary ependymoma identified on permanent section using immunochemistry [12]. To the best of our knowledge, this is the first reported case of association between chordoma and germinoma tumors. We suspect that because of the rarity of the two tumors, it would be unlikely that this is a pure coincidence. Further cytoanalysis or genetic analysis is needed to clarify the link between these two tumors.

Conflicts

There are no financial disclosures to be made.

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References