Case Report

Pediatric sellar solitary fibrous tumor/hemangiopericytoma: A rare case report and review of the literature

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ABSTRACT

Background: Solitary fibrous tumor (SFT)/hemangiopericytoma (HPC) is a rare tumor which originates from the walls of capillaries and has historically been thought to be able to occur anywhere in the body that blood vessels are found. It is rarely found in the sellar region.

Case Description: In this report, we present the first case of this tumor occurring in the sellar region of a pediatric patient. This 12-year-old male presented with progressive vision loss which prompted surgical resection after a sellar lesion was discovered on imaging. The initial transsphenoidal approach resulted in subtotal resection and the patient experienced recurrence within 3 months. He underwent an orbitozygomatic craniotomy to achieve gross total tumor resection.

Conclusion: We conducted a literature review of intracranial SFT/HPC in the pediatric population and found it to be an extremely rare occurrence, with <30 cases reported. The incidence of SFT/HPC occurring in the sellar region for any age group was also found to be a rare entity. Treatment recommendations for this tumor are also scarce, based on retrospective chart reviews from the adult population. The role for adjuvant radiation has mixed results.

Keywords: Orbitozygomatic craniotomy, Pediatric hemangiopericytoma, Pediatric sellar tumor, Pediatric tumors

INTRODUCTION

Solitary fibrous tumor (SFT)/hemangiopericytoma (HPC) is a rare tumor derived from the wall of capillaries and has historically been thought to be able to occur anywhere in the body, though it is rarely seen in the sellar region. The term “hemangiopericytoma” was first introduced in 1942 by Stout and Murray as a vascular tumor originating from Zimmermann’s pericytes, modified smooth muscle cells within the capillary walls that form endothelial tubes and sprouts. Since that time, HPCs have undergone various iterations of nomenclature; ultimately extracranial HPCs have been moved within the spectrum of “solitary fibrous tumors” with neuropathologists retaining the term “hemangiopericytoma.” Furthermore, a theory of origination from...
translocation occurring at chromosome 12, resulting in chromosomal fusion has replaced the theory of origination from Zimmerman’s pericytes.\textsuperscript{[26]}

Head-and-neck cases make up approximately 25–33% of total HPC with <1% of these lesions occurring intracranial. HPC is rare in adults, and even more so in children, as less than 10% of HPC occur in children.\textsuperscript{[17]} Intracranial HPC for pediatric patients has rarely been reported in the literature. In this report, we describe a rare entity in the pediatric population; a sellar region lesion mimicking a nonfunctional pituitary adenoma on imaging, with a determined final pathology of SFT/HPC. To the best of our knowledge, this is the first reported sellar SFT/HPC in the pediatric population. Furthermore, we performed a literature search of pediatric intracranial HPC and general reports of SFT/HPC located in the sellar region.

CASE PRESENTATION

Our patient is a 12-year-old male who was initially being evaluated by optometry for headaches and progressive visual impairment over a 2-month period. On assessment of the patient’s vision, a diagnosis of bitemporal hemianopsia was made, prompting magnetic resonance imaging (MRI) of brain and orbits. As demonstrated in [Figure 1], it revealed a sellar lesion with suprasellar extension exhibiting homogenous enhancement. At this time, a broad differential diagnosis was considered, including those typified by sellar lesions (including sarcoid, aneurysm, teratoma, craniopharyngioma, hypothalamic glioma, meningioma, metastasis, or optic nerve glioma).

On neurological examination, the patient was neurologically intact with the exception of bitemporal visual field loss. Visual acuity was noted to be 20/20 on the right and 20/100 on the left. Serum laboratory assessment for hormonal irregularities was unremarkable. The patient was also noted to have polydipsia without overt signs of diabetes insipidus (i.e., stable serum sodium levels and remaining euvoletic). He subsequently underwent transanal transsphenoidal craniotomy for resection and decompression of the optic chiasm. Postoperatively, the patient recovered well and was subsequently discharged home without any evident complications, with plan for possible further surgery after pathology finalized.

Intraoperatively, the tumor was noted to be firm and fibrous. Complete resection was not feasible from the trans-nasal approach as the tumor was noted to be extremely adherent to surrounding structures. Frozen section pathology from the initial surgery was inconclusive. However, final pathology from this surgery was reported to be SFT/HPC, World Health Organization (WHO) Grade II.

On 3-month follow-up, imaging demonstrated progressive and enlarging lesion, as demonstrated in [Figure 2]. Furthermore, the patient also complained of worsening vision in the left eye, with visual acuity now at 20/200. The remainder of cranial nerves were intact. At this time, the patient returned to the operating room for a left-sided orbitozygomatic craniotomy. Tumor was visualized in the suprasellar region and resected in a piecemeal fashion with the aid of an ultrasonic aspirator. The tumor has a similar consistency to the first time – firm and fibrous with regions of high vascularity. Microsurgical techniques were used to dissect the tumor capsule from all neural elements. The surgery proceeded without complications.

Postoperative imaging, as visualized in [Figure 3], demonstrated gross total resection (GTR) of the mass. This time, the patient’s postoperative course was complicated with the development of diabetes insipidus requiring desmopressin. The patient also had transient loss of vision in the left eye which returned to preoperative baseline before discharge from the hospital. [Figure 4] demonstrates the pathology obtained from the patient; [Figure 4a-c] displays the hematoxylin and eosin staining which demonstrates a mesenchymal neoplasm with moderate cellularity and nuclear pleomorphism. Tumor cells were noted to form into sheets and clusters with fibrous stroma, as well as abundant vasculature. Furthermore, staghorn appearance and additional strong positivity for CD34 was noted along endothelial cells with no definitive staining of tumor cells [Figure 4d] as well as strong nuclear staining for

![Figure 1](image1.png)

**Figure 1**: Magnetic resonance imaging brain (a) sagittal, (b) coronal demonstrating a sellar lesion with suprasellar extension exhibiting homogenous enhancement. Red arrows demonstrate a sellar lesion with suprasellar extension with homogenous enhancement.

![Figure 2](image2.png)

**Figure 2**: On 3-month follow-up, magnetic resonance imaging (a) sagittal and (b) coronal sequences demonstrated progressive and enlarging lesion. Red arrows demonstrate the progressively enlarging lesion.
signal transducer and activator of transcription 6 (STAT6) [Figure 4e], confirming the diagnosis of SFT/HPC. The final diagnosis of HPC was agreed on by outside consultation to the Children's Hospital of Los Angeles, who reviewed the samples.

DISCUSSION

WHO classification

Extracranial SFT/HPCs have been reclassified as a spectrum of SFTs, whereas the HPC continues to be used among neuropathologists. Both entities share the 12q3 inversion and fusion of the NGFI-A-binding protein 2 (NAB2) and STAT6 genes with the STAT6 nuclear expression visible on immunohistochemistry.[20] This inversion and fusion juxtaposes NAB2 and STAT6 in an initiating event found to be a disease-specific gene fusion.[23] This finding may have contributed to the change to the 2016 WHO classification of central nervous system tumors, assigning three grades to the “SFT/HPC” class, breaking the tradition of assigning names to the classes (e.g., glioblastoma as Grade IV);[8] the specific descriptions of each grade is displayed in [Table 1]. In the prior 2007 WHO classification, “Hemangiopericytoma” was distinct from SFT and anaplastic HPC.[12] For our case, a WHO Grade II SFT/HPC is the same as the prior “hemangiopericytoma” classification, thus the literature search was limited to this term.

Pediatric SFT/HPC

HPCs have the potential to occur anywhere in the body as they originate from blood vessels but the most common location is in the lower extremity in the pediatric population.[9] Intracranial SFT/HPC is a rare entity in the pediatric population. Limited cases were identified in the literature as can be found in [Table 2]. Moreover, from

**Table 1:** Three grades to the “solitary fibrous tumor/hemangiopericytoma” class, breaking the tradition of assigning names to the classes (e.g., glioblastoma as Grade IV). The specific descriptions of each grade are depicted above.

| WHO Grade (2016) | Description | Prior WHO (2007) Name |
|-----------------|-------------|-----------------------|
| I               | Highly collagenous, relatively low cellularity, spindle cell lesion | Solitary fibrous tumor |
| II              | More cellular, less collagenous tumor with plump cells and “staghorn” vasculature | Hemangiopericytoma |
| III             | Five or more mitoses per 10 high-power fields | Anaplastic hemangiopericytoma |

**Table 2:** Limited cases of intracranial solitary fibrous tumor/hemangiopericytoma identified in the literature.

| Pediatric cases of solitary fibrous tumor/hemangiopericytoma |
|-------------------|-----|-----|-----|
| References        | Year | Patients | STAT6 | CD34 |
| Herzog et al.     | 1995 | 2     | N    | N    |
| Cole and Naul     | 2000 | 1     | N    | N    |
| Bunai et al.      | 2008 | 1     | N    | Y (pos) |
| Fernandez-Pineda et al. | 2011 | 5     | N    | N    |
| Kerl et al.       | 2011 | 1     | N    | Y (pos) |
| Chen et al.       | 2012 | 9     | N    | N    |
| Trabelsi et al.   | 2015 | 1     | Y    | Y (neg) |
| Pang et al.       | 2015 | 2     | N    | N    |
| Silva da Costa et al. | 2017 | 1     | N    | N    |
| Yilmaz Semerci et al. | 2017 | 1     | N    | Y (neg) |
| Roth and Constatini | 2017 | 1     | N    | N    |
| Total             | 25   |       |      |      |
the available literature, there are no reports that were of HPC located in the sellar region. The largest case series for pediatric HPC reported nine cases with the cranial convexity and parasagittal region being the most common locations.\textsuperscript{10}

Two forms of HPC have been identified during childhood; infantile type which occurs during the first 12 months of life and the adult type which occurs after the age of 12 months old.\textsuperscript{4,17} Pediatric HPC consists of both entities and distinction between the two which is essential for treatment strategies. Infantile HPCs are considered congenital and have a favorable prognosis. Similar to adult HPC, infantile HPC has been classified at times in the literature as “infantile myofibromatosis” with studies suggesting both as different stages of the same entity.\textsuperscript{16,25} Adult HPCs behave similar in the pediatric population as the adult population. This distinction is made from the clinical standpoint rather than histological.\textsuperscript{9}

A recent study\textsuperscript{24} from three pediatric hospitals reevaluating prior pathology from the past 30 years found that 18 tumors total were classified as SFTs. However, when immunohistochemistry was performed on the tissue, only three tumors from two patients were found to be STAT6 positive; the remainder tumors required reclassification. This demonstrates that the diagnosis of SFT, and HPC, may be rarer than previously thought. Indeed, in the past, both SFTs and HPCs were referred to histologically as “angioblastic meningiomas” or “hemangiopericytic variant of meningioma,” before the advent of STAT6 immunopositivity studies.\textsuperscript{1} Our patient’s tissue was confirmed to be STAT6 positive [Figure 4e]. Furthermore, the implications of this study are vast as much of the prior epidemiological studies may have included false-positive diagnosis.

### STAT6 for diagnosis of SFT/HPC

STAT6 immunohistochemistry has been found to be an exclusively nuclear immunostaining signal with a strong ability to verify SFTs and HPCs, with recent studies showing all non-SFT/HPCs unanimously STAT6 negative and 98.7% agreement among observers.\textsuperscript{13} In the past, CD34 has been the most characteristic immunohistochemical stain for SFTs, however, it is also nonspecific.\textsuperscript{1,20} Very little was known about the molecular genetics of this tumor. Around 2013, immunohistochemical staining for STAT6 was presented in the literature as a more specific marker for SFTs and HPC.\textsuperscript{1,3,11,19} A NAB2-STAT6 gene fusion, results in an inversion at the 12q13 locus, produces a chimeric protein in which a repressor domain of NAB2 (EGR1-binding protein 2) (NAB2) is replaced with a carboxyterminal transactivation domain from signal transducer. The activator of transcription 6 and interleukin-4 induced (STAT6) acts as a transcriptional activator through early growth of the tumor and has been recently believed to be highly sensitive and specific for SFTs. Several journal articles have confirmed the validation of the diagnosis of intracranial and extracranial SFT.\textsuperscript{6}

### Sellar location SFT/HPC

To date, a total of 15 cases of sellar region SFT/HPC were identified in the English literature, as listed in [Table 3]. However, none have been reported in the pediatric population. The youngest patient in literature was an 18-year-old female who also had similar imaging – homogenously enhancing mass concerning for pituitary adenoma.\textsuperscript{26} The surgical approach utilized in these cases was transphenoidal, transcranial, or the former followed by latter after diagnosis was revealed and significant residual tumor or recurrence occurred. Many of these patients presented with endocrinopathies that would result from pituitary compression, as well as visual change from compression of the optic apparatus. The most common pediatric tumor in this location is a craniopharyngioma (6–9%), followed by hypothalamic glioma (4–8%), germ cell tumors (1–2%), and pituitary adenoma (0.5–2.5%).\textsuperscript{10,25}

### Treatment

Given the rarity of HPC, information about management specific to intracranial HPC for pediatric population is scarce. Moreover, the distinction between infantile and adult HPC is paramount to the clinical course. The infantile variant, occurring <12 months of age, is chemoresponsive. There are reports of extracranial infantile HPC exhibiting spontaneous regression.\textsuperscript{4,14} Moreover, this variant has a propensity to mature into benign neoplasms, such as hemangioblastoma, and has been shown to be responsive to neoadjuvant chemotherapy.\textsuperscript{7} One report of a neonate with intracranial HPC was treated with neoadjuvant anthracycline-based chemotherapeutic agent which was able to decrease tumor size before surgical resection.\textsuperscript{8} Another series of two infantile HPCs were treated with surgical resection without adjuvant therapy with no evidence of recurrence at time of report.\textsuperscript{9,14,21} Large series specific for intracranial infantile HPC are lacking given limited cases.

On the contrary, adult HPC requires a more aggressive approach. Surgical resection followed by adjuvant radiation therapy has been recommended in the past for treatment from adult literature.\textsuperscript{21} In a 29-patient series report for intracranial adult HPC, the 5-, 10-, and 15-year overall survival rates were 85%, 68%, and 43%, respectively, for patients undergoing this treatment route.\textsuperscript{13} Another series of 43-patient cases reported 1-, 5-, and 10-year overall survival at 100%, 94.4%, and 72.2%, respectively.\textsuperscript{21} Outcome is directly related to the presence of metastatic disease and adequacy of local control; GTR is advocated as the 5-year local disease control rates were 84% when compared to only 38% for subtotal resection (STR).\textsuperscript{18}
However, a recent meta-analysis of 563 cases found that GTR alone provided superior survival, with or without adjuvant radiation therapy; patients receiving >50 Gy of radiation had worse survival outcomes. This study included pediatric patients, with age ranging from 1 month to 80 years, however, the distinction between pediatric and adult population was not made in their analysis nor was the number of patients under age 18 reported. A single-institution retrospective review of 43 patients reported adjuvant radiation can hinder tumor progression but appears to have no effect on overall survival. Patients with GTR alone were compared to patients with STR + radiation and the latter group had longer progression-free interval. However, when comparing overall survival for the same groups, GTR alone had better results.

Data specific to pediatric population for intracranial disease is lacking; however, given the similar clinical behavior of extracranial disease, we believe that these results can be extrapolated to the pediatric population with intracranial adult HPC. Surgical intervention with GTR should be the goal with conflicting evidence for adjuvant radiation therapy. Given the benefit of improved recurrence-free interval with radiation therapy, there is likely some benefit to adjuvant radiation therapy if total dose is kept below 50 Gy. This dose limit may need to be decreased in younger population given proximity to optic apparatus, further research is needed in this area.

**CONCLUSION**

The authors present a report of a SFT/HPC occurring in a 12-year-old male patient in the sella. This lesion mimicked a nonfunctioning pituitary adenoma both clinically and on imaging, with a surprising final pathological result; this is the first report of HPC occurring in the sellar region in the pediatric population. Moreover, recent literature with regard to STAT6 positivity suggests that many prior diagnoses of HPC may be false positives, making it an even rarer entity. Data are lacking for evidence-based treatment recommendations for pediatric HPC; from adult data, goals favor GTR for best overall survival for adult-type HPC, which occurs in the pediatric population after the age of 1 year. The role for adjuvant radiation therapy is unclear at this time, with benefits seen with in patients only able to undergo STR with regard to the progression-free interval. Prospective trials are needed to further characterize specific treatment modalities.

**Declaration of patient consent**

Patient’s consent not required as patients identity is not disclosed or compromised.

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**Conflicts of interest**

There are no conflicts of interest.

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