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CASE REPORT

Pediatric central nervous system solitary fibrous tumor: case report

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Abstract
Introduction Solitary fibrous tumors are mesenchymally derived masses most commonly originating from the lung pleura.
Case report Herein, we report a 6-month-old presenting with syndrome of inappropriate antidiuretic hormone secretion (SIADH) and a suprasellar mass. The mass proved to be a solitary fibrous tumor. This case and salient literature are reviewed.
Conclusions To our knowledge, this is the youngest patient to be described with a mass of this type within the central nervous system.

Keywords Tumor · Brain · Intracranial · Suprasellar · Third ventricle · Hypothalamus · Pediatric

Introduction

Solitary fibrous tumors are mesenchymally derived masses most commonly originating from the lung pleura [1]; however, they can arise from virtually any site in the body [2]. Solitary fibrous tumors of the central nervous system (CNS) were first placed into a separate pathological entity in 1996 when they were defined as a rare mesenchymal, nonmeningothelial tumor [3]. The finding of a solitary fibrous tumor in the CNS is a rare occurrence [4] possibly due to relative absence of true connective tissue [5]. It has been shown however that the development of these masses from dural fibroblasts may go on to invade brain parenchyma, nerve roots, or the skull base [6, 7]. A review of the literature by Centeno et al. reported that intracranial solid fibrous tumors can occur across a range of ages from 11 to 73 years with a median age of 47.6 years and approximate equal gender distribution [8]. Bisceglia et al. [9] reviewed the literature regarding 220 cases of solitary fibrous tumors of the central nervous system and found that most are dural-based and intracranial in location.

Case report

We present a 6-month-old boy presenting with diarrhea and vomiting who was found to have hyponatremia (Na 119). A septic workup was performed and empiric antibiotics begun. The patient was full term with a history of bronchiolitis at 2 months of age. There was a family history of Hashimoto's thyroiditis and lupus. CSF was positive for enterovirus and VRP positive for paraflu. Antibiotics were stopped after 3 days. The patient's persistent syndrome of inappropriate antidiuretic hormone secretion (SIADH) without a clear etiology led to MRI of the brain, which occurred approximately 1 month after presentation. A large (5×5×3 cm), enhancing, suprasellar mass in the third ventricle was found (Fig. 1a–c). One month later, MRI showed minimal but measurable growth of the lesion. A biopsy was performed and was consistent with a solitary fibrous tumor. A craniotomy was performed a month later with the goal of radical debulking. Histological sections (Fig. 2) demonstrated a mildly to moderately hypercellular neoplasm with prominent collagenized

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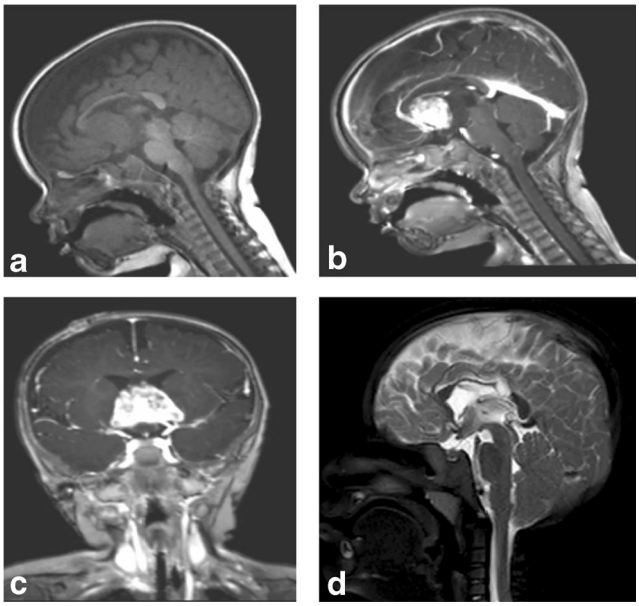

Fig. 1 **a** Preoperative sagittal MRI without contrast demonstrating 5×5×3 cm tumor of the third ventricle of case reported herein. Note the tumor's upper limit under the genu of the corpus callosum. **b** Preoperative sagittal MRI with contrast. Note the relationship of the tumor to the anterior cerebral arteries. **c** Preoperative coronal MRI with contrast. Note the pyramidal shape of the tumor in the coronal plane and suprasellar location. **d** Follow up T2-weighted sagittal MRI image at 1.5 years follow up noting gross total resection cavity without enhancing tumor

stroma. The tumor was composed of fascicles of cells with slightly spindled to fusiform nuclei and a moderate amount of cytoplasm. Mitotic figures were not readily found. There were no neuropils in the specimen. The tumor cells were diffusely positive for CD34. An immunostain for CD99 showed focal positivity in the tumor cells. The bcl-2 immunostain only highlighted occasional cells. The Ki-67 (MIB-1) proliferation index stain was positive in <1 % of tumor cells. Control slides reacted appropriately. No data were available to suggest that chemotherapy might be effective in this tumor type. The patient's postoperative course was complicated by transient diabetes insipidus, then by persistent SIADH, which was treated with fluid restriction. A ventriculoperitoneal shunt was placed 4 months post operatively and at 1.5 years follow-up, and the child is developing normally with no recurrence on repeat imaging (Fig. 1d).

Discussion

Solitary fibrous tumors are rare masses that develop from fibroblasts most commonly in the pleura of the lungs [1] but have been described in multiple different body areas including, albeit rarely, the dura mater [5–8, 10]. There have been multiple locations of origin described including the clinoid process, middle cranial fossa, falx cerebri, lateral ventricle, and cerebellopontine angle [11–13]. Interestingly, while a

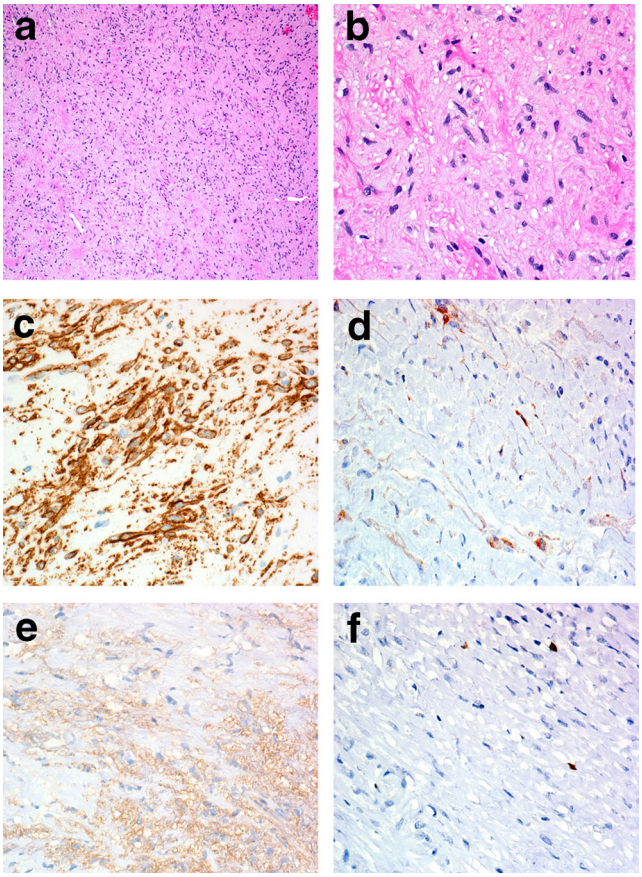

Fig 2 Histological images of tumor resected **a** HE (100×); **b** HE (400×); **c** CD34 (400×); **d** Bcl-2 (400×); **e** CD99 (400×); **f** MIB-1 (400×)

majority of intracranial solitary fibrous tumors contain dural attachments, approximately two-thirds of cases found in the spinal region lack any dural connection [14, 15]. Histologically, hemangiopericytoma and solitary fibrous tumors are considered the same, especially with NAB2-STAT6 fusion but behave differently clinically [16]. Solitary fibrous tumors are benign and if removed completely, do not return. However, hemangiopericytoma tend to be aggressive tumors with the potential for local recurrence and metastases [10]. A newer marker for hemangiopericytoma/solitary fibrous tumor is Stat-6. Unfortunately, this new marker was not available for our case. Vimentin, CD34, BCL2, and CD99 are consistently the most positive markers for solitary fibrous tumors [9].

Treatment for solitary fibrous tumors is complete removal. The most dependent factor for recurrence described at other anatomical sites was whether a complete excision was achieved [17]. In fact, approximately 25 % of solitary fibrous tumors recurred in cases where there was incomplete resection and patients were followed for at least 1 year [18]. This finding implies that solid fibrous tumors grow more slowly than atypical meningiomas and are less aggressive than hemangiopericytomas [4]; however because of the paucity of pediatric solitary fibrous tumors reported in the literature, appropriate treatment and prognosis is unclear.

Intraoperatively, these tumors can be adherent and vascular making dissection away from the brain parenchyma difficult. In addition, these masses are complicated by the potential for postoperative hemorrhage. In spite of this, gross total resection is usually possible [19]. If complete excision cannot be completed, the addition of radiotherapy has been utilized and even chemotherapy in cases where the cellular proliferation was found to be high [20, 21].

Conclusion

In this case report, we describe a case of a 6-month-old child who presented with a solitary fibrous tumor of the central nervous system. To our knowledge, this is the youngest patient to ever have been described with a mass of this type within the central nervous system [22]. Although mostly benign, recurrence and malignant transformation have been reported [22, 23]; however, because a case of solitary fibrous tumor has not been described previously in a patient so young, the natural course is not well understood.

Conflict of interest The authors have no conflicts of interest to report.

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