Pediatric alveolar soft part sarcoma of the orbit: a case report and literature review

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Abstract
Alveolar soft part sarcoma (ASPS) of the orbit is exceedingly rare but little is known regarding its radiologic features. Here, we reviewed the CT and MRI findings of one case of ASPS of the orbit with emphasis on its salient imaging features.

Keywords: alveolar soft part sarcoma, orbit, radiology

INTRODUCTION
Alveolar soft part sarcoma (ASPS), which accounts for approximately 0.5–1% of all soft tissue neoplasms, was firstly described by Christopherson et al. in 1952[1]. Although it is found in some unusual locations[2], ASPS of the orbit has been extremely rarely reported. Most previous reports have mostly focused on the pathological features of the disease while few papers have focused on the imaging features. Here, we present one case of ASPS of the orbit with emphasis on CT and MRI findings of the disease that could help the differential diagnosis of ASPS.

CASE REPORT
A 10-year-old girl presented with a 2-month history of slowly increasing proptosis of the left eyeball with progressively decreased visual acuity. She denied any past medical problems, especially neurological or ophthalmological complaints. Physical examination revealed visual oculus dexter 0.4, visual oculus sinister 0.3, left side proptosis and limited external movement. The remainder of the ophthalmologic evaluation was unremarkable.

CT examination performed 3 days before hospitalization showed a well-circumscribed, irregular mass measuring 15 × 20 × 18 mm in the lower outer quadrant of the left orbit. CT value of the mass was approximately 52 hounsfield units (HU) with no obvious bone destruction. MRI examination performed 2 days before hospitalization indicated that the mass showed heterogeneous isointensity on T1WI, heterogeneous iso-to slightly hyperintensity on T2WI, and heterogeneous enhancement after contrast administration. Flow void signals were seen on T1, T2, and post-enhanced images (Fig. 1A to 1E).

Five days after hospitalization, the tumor was resected through a trans-conjunctival inferior orbitotomy. Gross examination revealed a firm, grayish white tumor, measuring about 15 mm in diameter. Microscopic examination showed that the tumor was composed of round cells with mild nuclear pleomorphism and the tumor cells were arranged in nests; sheet structures were separated by septa of vascularized fibrous connective...
tissue (Fig. 1F). Immunohistochemical stains disclosed positive staining of the tumor cells for myogenic determination factor1 (+) and destin (+), and negative staining for smooth muscle actin (-), neuron specific enolase (-), S-100 protein (-), and CD34 (-) (Fig. 1G and 1H). The morphological and immunohistochemical features are consistent with the diagnosis of ASPS.

Postoperatively, the patient was referred to oncologists for further advice on the management of ASPS. Seventeen days after the procedure, the first round of chemotherapy began with the DVCC regimen (Doxorubicin, 30 mg/m², ivd, d1, d8; vincristine, 1.5 mg/m², ivp, d0, d7; cyclophosphamide, 300 mg/m², ivd, d1–d3; cisplatin, 90 mg/m², ivd, d0 ). The IVE regimen ( ifosfamide, 1.5 g/m², ivd, d1–d5; vincristine, 1.5 mg/m², ivp, d0, d7; etoposide, 100 mg/m², ivd,d1–d5) was the alternate chemotherapy used at approximately monthly intervals.

Follow-up MRI examinations were performed at 3, 6, and 12 months after the procedure, respectively. Both the clinical and MRI evaluation revealed no local recurrence or distant metastases (Fig. 2).

**DISCUSSION**

Most previous reports of orbital ASPS have focused on the pathological features with few reports on its radiological features. Here, we followed up a pediatric case of orbital ASPS and reviewed the diagnosis and differential diagnosis from the view of radiologic imaging. In

**Fig. 1 Radiological and pathological examination of a 10-year-old girl with alveolar soft part sarcoma of the orbit.** Axial T1WI and T2WI MR show a left orbit mass which contains flow void signal (a–b). CT shows a well-circumscribed, irregular mass in the lower outer quadrant and no obvious bone destruction is found (c). Axial and coronal contrast-enhanced fat-suppressed T1WI MR shows markedly heterogeneous enhancement (d–e). Microscopic examination shows that the tumor is composed of round cells with mild nuclear pleomorphism, and the tumor cells are arranged in sheets separated by septa of vascularized fibrous connective tissue (H&E, magnification, × 100) (f). Immunohistochemistry shows positive staining of the tumor cells for MyoD (+) and destin (+) (magnification, × 100) (g–h).

**Fig. 2 Serial axial T1WI, T2WI and enhanced T1WI demonstrate no local recurrence at 12 months after procedure.**
a previous study, Rebecca et al.\(^6\) indicated that pediatric ASPS is denser than muscle on plain CT and demonstrates significant peripheral contrast enhancement. On MRI, ASPS may appear to have iso-to hyperintensity on T1WI, and hyperintensity on T2WI because of the abundant, but slow, blood flow within the tumor. Meanwhile, flow void signal may also be seen due to the dilated vessels in and around the lesion. Similarly, an ASPS mass in the extremity also demonstrated iso-intensity on T1WI, hyperintensity on T2WI and heterogeneous enhancement after contrast administration \(^7\). In our present report, the orbital ASPS mass showed as a hyper-dense mass on plain CT, with isointensity on T1WI, T2WI, and heterogeneous enhancement after contrast administration. Meanwhile, flow void signals were seen on T1W, T2W and enhanced images. All these image findings are similar to those described in previous reports\(^{6-7}\).

Radiologically, the most important differential diagnosis for ASPS is orbital solitary fibrous tumor (SFT). Orbital SFT is a well-defined, non-encapsulated tumor with a patternless arrangement of spindle cells. By MRI, orbital SFT commonly shows as isointensity on both T1WI and T2WI, and exhibits moderate enhancement after contrast administration. However, most SFTs are located in the superior portion of the orbit and less flow void signal is seen, compared with ASPS\(^6\). In addition to SFT, some other rare tumors, such as rhabdomyosarcoma, capillary hemangioma, or giant cell angiofibroma should also be considered.

ASPS of the orbit is an extremely rare malignancy, accounting for a small proportion of orbital tumors. The definitive diagnosis of this disease depends mainly on the pathologic examination, especially immunohis-tochemical staining. The most appropriate choice of treatment for ASPS is early surgery; the roles of radiotherapy and chemotherapy still need to be evaluated.

In conclusion, we report a case of orbital ASPS, mainly from the perspective of the imaging features. ASPS should be considered when a highly vascularized orbital lesion is seen and CT and MRI should be performed as part of a comprehensive analysis to narrow down the differential diagnosis.

References


