

# Primary Intracerebral Alveolar Soft Part Sarcoma in an 11-Year-Old Girl: Case Report and Review of the Literature

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Alveolar soft part sarcoma (ASPS), a rarely observed tumor, is a soft tissue sarcoma with an unidentified cell origin. It constitutes 0.5–1.0% of all soft tissue sarcomas. It may appear in various parts of the body, but mostly observed in the trunk and the extremities. It has a high metastasis potential. To the best of our knowledge, only three cases of primary intracranial ASPS without a demonstrable lesion elsewhere is encountered. An 11-year-old girl was operated because of fronto-parietal mass lesion by craniotomy. Pathological examination revealed ASPS and no primary focus was detected. In spite of radiotherapy and chemotherapy as an adjuvant therapy, after 45 months she had a second operation for recurrence of the tumor. Since it is possible to observe metastases in late phases, up to 30 years, the patients must be followed up for a long period. Although radiotherapy and chemotherapy followed by surgery is the most accepted treatment strategy, the prognosis is still poor.

**Keywords:** alveolar soft part sarcoma, childhood tumors, intracerebral, primary, surgery

## Introduction

Brain tumors are the most common solid tumor type in children and constitute 20% of total cancer cases observed in this age group.<sup>1</sup> In childhood, the distribution of central nervous system (CNS) tumors is quite different from adult counterparts. Approximately 55% of the CNS tumors in childhood period are located in posterior fossa. In adults, 10% of the intracranial tumors arise in posterior fossa.<sup>2</sup> Alveolar soft part sarcoma (ASPS), a rarely observed tumor, is a soft tissue sarcoma with an unidentified cell origin. ASPS constitute 0.5–1.0% of all soft tissue sarcomas.<sup>3</sup> They were first discovered by Christopherson et al. in 1952.<sup>4</sup> ASPS usually presented with a slowly growing painful mass lesion between the ages 15–35.<sup>3</sup> It is rarely seen in childhood with the rate of 0.8–1.8%.<sup>5,6</sup> It may appear in various parts of the body such as tongue, uterus, stomach, vagina, bones, veins, and sacrum but is mostly observed in the trunk and the extremities.<sup>3,7–11</sup> It has a high metastasis potential and generally metastasizes to lungs, brain, bones, and lymph

nodes. Since it is possible to observe metastases in late phases up to 30 years, the patients must be followed up for a long period.<sup>8,12</sup> Although intraparenchymal sarcoma and metastasis are mentioned in the literature, to the best of our knowledge seven cases of ASPS with initial brain manifestations of primary intracranial ASPS have been reported, and four of them had concomitant lung metastasis<sup>13–19</sup> (Table 1). This is the fourth case of primary intracranial ASPS without a demonstrable lesion elsewhere. Among all, presented patient has the longest follow-up period of 54 months.

## Case

An 11-year old girl with complaints of headache, involuntary movement of right arm, and paresthesia in right foot and tongue was admitted. Neurological examination revealed 4/5 monoparesia in the right arm. Magnetic resonance imaging (MRI) revealed a solitary well-defined lobulated mass lesion of left frontal lobe that was located in the subcortical white matter and extending to the deep white matter. The lesion was heterogeneously hyperintense on T<sub>2</sub>-weighted (Fig. 1A) and hypointense on T<sub>1</sub>-weighted (Fig. 1B) images. The T<sub>1</sub>-weighted imaging following gadolinium injection demonstrated the prominent and homogeneous enhancement of the lesion (Fig. 1C). There was a prominent peritumoral vasogenic edema. We decided to operate the patient for decompression and biopsy. Gross total excision was performed through left fronto-parietal craniotomy. There were no dural and bony involvement. The tumor was red-purple in color with high vascularization, and had significant invasion to brain. Post-operative MRI examination showed total removal of tumor (Fig. 1D). The post-operative period was uneventful, she was discharged on the 5th day of operation with 4/5 monoparesis in her right arm. On pathological examination, the tumor was showing alveolar nesting pattern with tumor cells containing abundant eosinophilic cytoplasm and discohesive nature. There were periodic acid schiff (PAS)-positive and diastase-resistant rod-shaped multiple crystalline materials in the cytoplasm of some tumor cells and reticulin (silver) framework surrounding the tumor nests. Tumor cells were showing focal immunoreactivity with glial fibrillary acidic protein (GFAP) and no immunoreactivity with S100 protein. Several tumor cells were showing nuclear immunoreactivity with transcription factor E3 (TFE3). These findings revealed alveolar soft sarcoma (Fig. 2A–F). On this account, we aimed to find the primary focus. The hematological and biochemical analysis, paranasal, thoracic,

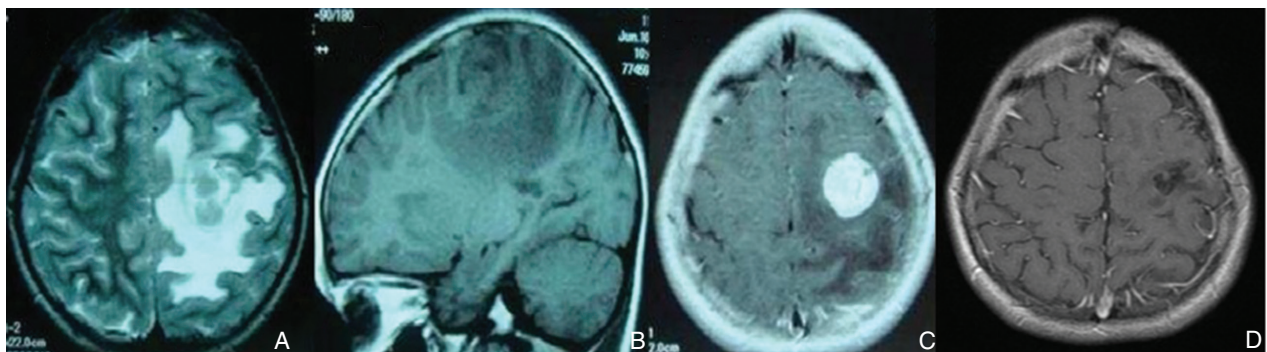
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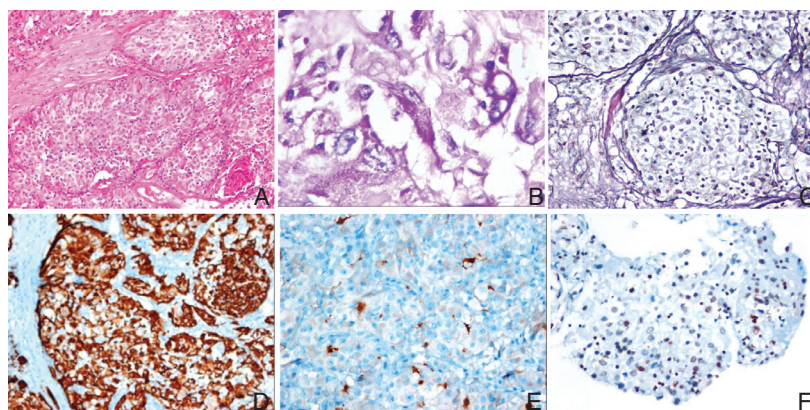
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**Table 1** Reported cases of alveolar soft part sarcoma with initial brain manifestations

No.	Authors	Age, sex	Symptom	Location	Primary site (after diagnosis or treatment site)	Management	Outcome	Concomitant metastasis
1	Lewis et al. <sup>17)</sup>	25, M	Headache, vomiting	Right occipital	Found after 5 years (thigh)	Surgery, radiotherapy	Lost to follow-up	Lung
2	Perry JR et al. <sup>18)</sup>	28, M	Partial seizures	Left frontal and parietal	Not found after 18 months follow-up	Surgery, whole brain radiotherapy, chemotherapy	Alive at 18 months	Lung
3	Cohen et al. <sup>15)</sup>	28, M	Headache, declining vision (right eye)	Right suprasellar and optic tract	Found after diagnosis (shoulder)	Surgery, whole brain radiotherapy, chemotherapy	Alive at 12 months	Lung
4	Sujit Kumar et al. <sup>19)</sup>	28, M	Headache, diplopia, visual obscuration, generalized tonic clonic seizures	Left frontal, left basal ganglia and right parietal	Not found after 18 months follow-up	Whole brain radiotherapy	Alive at 18 months	Lung
5	Bodi et al. <sup>14)</sup>	39, M	Seizures	Left temporal meningeal	Not found after 10 months follow-up	Surgery	Alive at 10 months	None
6	Das et al. <sup>16)</sup>	17, F	Frontal mass	Bifrontal (right > left)	Not found after 4 months follow-up	Surgery, adjuvant radiotherapy	Alive at 4 months	None
7	Ahn et al. <sup>13)</sup>	9, F	Tinnitus, headache, vomiting, partial seizures	Cerebellopontine angle	Not found after 29 months follow-up	Surgery, radiotherapy, chemotherapy, radiosurgery	Regrowth (after 29 months)	None
8	Present case	11, F	Headache, involuntary movement of right arm, paresthesia in right foot and tongue	Left frontal	Not found after 54 months follow-up	Surgery, whole brain radiotherapy, chemotherapy, re-surgery, chemotherapy	Regrowth (after 45 months)	None



**Fig. 1** Axial T<sub>2</sub>-weighted (A) and sagittal T<sub>1</sub>-weighted (B) MR images show the left frontal intra-axial mass lesion that was hyperintense on T<sub>2</sub>-(A) and hypointense on T<sub>1</sub>-(B) weighted images. The lesion shows intense and homogeneous enhancement on postcontrast T<sub>1</sub>-weighted (C) image. MR images (A–C) reveal the prominent peritumoral vasogenic edema. T<sub>1</sub>-weighted images show no contrast enhancement was detected in the operation area (D). MR: magnetic resonance.



**Fig. 2** A: Alveolar soft part sarcoma showing alveolar nesting pattern with tumor cells containing abundant eosinophilic cytoplasm and discohesive nature (H&E, ×40), B: There are histochemically PAS-positive and diastase-resistant rod-shaped multiple crystalline materials in the cytoplasm of some tumor cells (dPAS, ×400), C: Histochemically reticulin (silver) framework surrounding the tumor nests (Reticulin, ×400), D: Tumor cells showing focal immunoreactivity with GFAP (×400), E: Tumor cells with no immunoreactivity with S100 protein (×400), F: Several tumor cells showing nuclear immunoreactivity with TFE3 (×400). PAS: periodic acid schiff, dPAS: distaste periodic acid schiff, GFAP: glial fibrillary acidic protein, TFE3: transcription factor E3.

abdominal computed tomographies (CTs), and aspiration of bone marrow were performed; however no focus was found. The positron emission tomography (PET) CT examination was also normal. She was transferred to pediatric oncology department for further treatment [radiotherapy (RT) and chemotherapy] with diagnosis of primary intracerebral ASPS. After receiving conventional RT with 54 Gy, 9 cycles of chemotherapy using the regimen of the pediatric oncology group—ifosfamide, vincristine, actinomycin—started. Her follow-up performed at 6 months intervals was uneventful. However, MRI examination performed 45 months after the initial treatment demonstrated a recurrence. The T<sub>1</sub>-weighted imaging following gadolinium injection demonstrated the prominent and homogeneous enhancement of the recurrent lesion (Fig. 3A). Re-operation was performed and gross-total excision was achieved again. There was no contrast enhancement at the postop control MRI (Fig. 3B). Pathological examination was the same with the first diagnosis. A 6 cycles of ifosfamide, carboplatin, and etoposide chemotherapy regimen was started. There was no recurrence at 9 months follow-up.

## Discussion

ASPS is a rare form of soft tissue sarcoma. The disease affects primarily younger patients and occurs more commonly in women.<sup>3</sup> In adults it is more frequent in the extremities whereas in children the head and neck region is affected more commonly. Ogoose et al. reported that approximately 10% of these tumors may occur as primary bone tumors.<sup>20</sup>

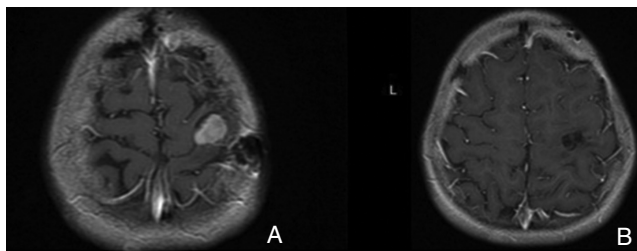
The patients generally present with pain and mass, and since the tumor is growing slowly, metastases are present at the time of diagnosis in significant number of patients. The tumor is rich in terms of vessels; sometimes murmur can be heard as a clinical sign.<sup>3</sup> In differential diagnosis arteriovenous malformations and benign tumors in head-neck region should be regarded.<sup>21</sup>

In radiological differential diagnosis, other aggressive lesions of bone such as lymphoma, plasmacytoma, Ewing sarcoma, malignant fibrous histiocytoma, and other aggressive bone lesions should also be keep in mind. ASPS commonly demonstrates increased signal intensity on both T<sub>1</sub>-weighted and T<sub>2</sub>-weighted images and can be misdiagnosed as hemangioma.<sup>22</sup> Unlike the hemangioma, in ASPS peripheral contrast enhancement and central necrosis can be

observed in CT. Chu et al. reported a case with ASPS mimicking hemangioma.<sup>23</sup> Heterogeneous contrast enhancement after gadolinium administration is another MRI finding that can suggest ASPS. Due to more prominent arterial structure in ASPS, areas with low signal intensity are more apparent compared to hemangioma. In the differential diagnosis, tumors showing hyperintensity on T<sub>1</sub>-weighted images, such as clear cell sarcoma, metastatic melanoma, liposarcoma, and soft-tissue tumors associated with hemorrhage should also be considered.<sup>22</sup> In the presented case, MRI findings were similar; increased signal intensity and contrast enhancement was determined. Scintigraphic methods can be used in order to evaluate far metastasis of tumor and bone invasion.<sup>24</sup> The identification rate of conventional diagnostic procedures in patients with primary lesion is low. This low rate can be attributed to several reasons. The major reasons are small primary lesion and the loss of primary lesion as a result of apoptosis after metastasis.<sup>25</sup> In their study to evaluate the accuracy of FDG PET in primary tumor detection, Delgado-Bolton et al. estimated the sensitivity at 87% (50–100%) and the specificity at 71% (45–100%).<sup>26</sup>

The macroscopic appearance of the tumor is a round or lobulated soft mass that can be seen in a yellowish-white color.<sup>27</sup> ASPS may be mistaken as and must be differentiated from renal cell carcinoma (RCC), adrenocortical carcinoma (ACC), hepatocellular carcinoma (HCC), pediatric age RCC with TFE3 gene fusion, malignant melanoma, paraganglioma, alveolar rhabdomyosarcoma, perivascular epithelioid cell tumors (PEComas) especially epithelioid angiomyolipoma, and granular cell tumor.<sup>3,9,28,29</sup> Immunohistochemical markers can be used in order to differentiate these entities from each other. RCC can be differentiated from ASPS with its positive staining for epithelial membrane antigen, pancytokeratin (CK), and RCC marker. ACC shows immunoreactivity with inhibin, melan-A, calretinin, and synaptophysin; but it is important to keep in mind that it may also express TFE3. HCC is positive for CK and HepPar1. Pediatric age RCC with TFE3 gene fusion is a tumor which usually shows CK and TFE expression; but sometimes it may not be possible to show these expressions properly, so it is critical to learn clinically if the patient has a tumoral mass in the kidney. Malignant melanoma shows immunoreactivity with S100 protein, HMB45, and melan-A. Paraganglioma stains positively for S100 protein, synaptophysin, and chromogranin. Alveolar rhabdomyosarcoma shows desmin and myogenin positivity. PEComas stain positively for smooth muscle actin, melan-A, and HMB45. Granular cell tumor is positive for S100 protein and it is also important to remember that it may also show TFE3 expression. In the immunohistochemical view of ASPS, it is known that it expresses vimentin, nuclear TFE3, and in some cases S100 protein in the rate of 15–30%; but not the other markers that are mentioned above.

Although these tumors are claimed to primarily develop from bones, there are immunohistochemical studies which assert that they originate from muscles.<sup>20,30</sup> Molecular studies reveal specific chromosome translocation.<sup>31,32</sup> Malignant melanoma, granular cell tumor, paraganglioma, and



**Fig. 3** The lesion shows intense and homogeneous enhancement on postcontrast T<sub>1</sub>-weighted image (A) and gross-total removal of tumor (B).

alveolar rhabdomyosarcoma can also give an alveolar appearance on histology. When diagnosis is difficult, the observation of typical crystals on electron microscopy and the use of immunohistochemical studies designed for tumor typing can be useful. For the diagnosis of ASPS, although various epithelial, neural, and muscle-related immunohistochemical parameters are being investigated, no specific indicator has been reported.<sup>33)</sup>

ASPS relatively grows slowly, and rarely demonstrates local recurrence after total excision; however its metastasis rate is high.<sup>34)</sup> ASPS generally metastasizes to lungs, brain, and surrounding bones. The metastases to locations such as chest wall, retroperitoneum, liver, and spleen have been shown.<sup>8)</sup> It has been reported that 40% of patients have lung metastases at initial diagnosis.<sup>12)</sup> Brain metastasis of sarcomas are quite rare. Generally, intraparenchymal brain metastasis develops in 1–8% of sarcomas in various histologies.<sup>35–41)</sup> This ratio exceptionally stands at 33% in ASPS patients.<sup>12)</sup>

In the treatment of localized ASPS, the most important step is the excision of the tumoral tissue with microscopically clean surgical margins.<sup>8)</sup> Adjuvant radiotherapy and chemotherapy are alternatively used in providing local control and when metastases are present; however there is limited information regarding their efficacy.<sup>8,20,34)</sup>

Presence of metastasis, tumor larger than 5 cm, and presence of bone involvement on the side of the tumor at the time of diagnosis are the most important factors for poor prognosis.<sup>8,20)</sup> For tumors smaller than 5 cm in diameter, 5-, 10-, and 15-year survival rates are 72%, 65%, and 65% respectively, and for tumors larger than 5 cm in diameter these ratios have been reported as 46%, 9%, and 0%.<sup>20)</sup> In patients without metastases at the time of diagnosis, the 5-, 10-, and 15-year survival rates are 81%, for patients with metastasis 5-year survival rate is 46% and 10-year survival rate is 0%.<sup>20)</sup> Lieberman et al. reported a 2-year survival rate of 77%, and a 5-year survival rate of 60% in 91 cases.<sup>12)</sup> Age, gender, and location of the primary tumor have been reported to have no effect on prognosis.<sup>20)</sup> For the current patient, presence of a tumor smaller than 5 cm in diameter, absence of metastasis, and gross total resection makes a good prognosis plausible.

For soft tissue sarcomas in extremities, to provide local control, achievement of microscopically clean surgical margins is very important.<sup>42)</sup> Sherman et al. provided very good local control with radiotherapy in six patients who had ASPS, and reported that routine radiotherapy was required.<sup>43)</sup> Local recurrence of ASPS is approximately 20%.<sup>12)</sup> Ogose et al. reported that in patients having insufficient surgical margins, radiotherapy should be considered.<sup>20)</sup> According to the established view, radiotherapy and chemotherapy have no significant effects on prognosis.<sup>8,9,12,44,45)</sup>

Recent chromosomal studies have provided us with detailed information regarding the genetics of the disease. The unbalanced translocation, der(17)t(X;17)(p11.2; q25), is characteristic of ASPS and was shown that this translocation fuses the TFE3 transcription factor gene at Xp11.2 to alveolar soft part locus (ASPL), a novel gene at 17q25.<sup>46)</sup> In the light of this recent information, specific targeting of ASPL-TFE3

mutation is deemed vital in order to control the disease and prolong the lifetime of patients. Hence, future research on the subject is considered to be of great importance.

To the best of our knowledge, this is the fourth case of primary intracranial ASPS without a demonstrable lesion elsewhere. Even though a primary focus cannot be determined in scans including PET CT, any future detection of a tumor at other sites of the body during follow-up will let us consider the described tumor as a brain metastasis. Therefore, the patients must be followed up for a long period due to the tendency of delayed recurrence.

## Conflicts of Interest Disclosure

None

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