

Primary Intracranial Alveolar Soft-Part Sarcoma: Case report and a Review of the Literature



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Abstract

Alveolar soft part sarcoma (ASPS) is a rare, malignant form of soft tissue sarcoma affecting young adults. A 24 years old male patient presented with the complaints of progressively increasing occipital swelling. On imaging, the lesion was suspected to be an intra-diploic meningioma. The patient underwent craniotomy and excision of tumor. Histopathology revealed the tumor to be alveolar soft part sarcoma. He received radiotherapy for small residual tumor invading transverse sinus. Post-operative metastatic work up did not reveal any other lesions. At 14 months follow up, the patient is asymptomatic with stable residual on imaging. We report a rare case of primary occipital ASPS along with a comprehensive review of literature.

Keywords: Alveolar Soft Part Sarcoma; Sarcoma; Meningioma; Brain tumor

Abbreviations: Symp: Symptoms; NCCT: Non-Contrast Computed Tomography; RT: Radiotherapy; CT: Chemotherapy; Mets: Metastasis

Introduction

Alveolar soft part sarcoma (ASPS) is a rare, malignant form of soft tissue sarcoma affecting young adults with a high potential for systemic metastasis [1]. Intracranial manifestation occurs late in the disease due to metastasis from peripheral ASPS [2]. We report a rare case of primary occipital ASPS along with a comprehensive review of literature.

Case description

A 24 years old male patient presented with the complaints of progressively increasing occipital swelling noticed for last 5 years and headache. On examination, he had a hard, non tender, non palatial swelling approximately 7x6 cms over the occipital region. Neurological examination was unremarkable except for mild cerebellar signs. On-contrast computed tomography (NCCT) scan of head showed a midline occipital hyper dense mass causing expansion and erosion of bone (Figures 1a & 1b). Contrast enhanced magnetic resonance imaging (MRI) brain showed an T1 is intense, T2 hyperintense extra-axial occipital lesion eroding bone and compressing cerebellum. It was intensely enhancing on contrast administration with central necrotic cavity. MR angiography showed highly vascular tumor with large tortuous vessel supplying the tumor (Figures 2a-2f). On the basis on imaging, he was suspected to have an intra-diploic hemangioma. Intraoperative a highly vascular tumor was present in subcutaneous tissue, eroding

and filling the intradiploic space, invading the dura and extending into the posterior fossa extra axially.

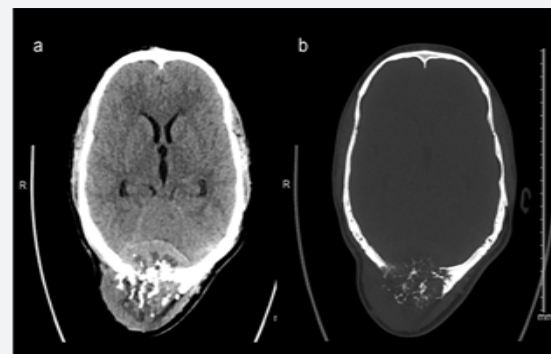


Figure 1: NCCT head:

- a.** Midline occipital hyper dense mass with intracranial and extra cranial components. Presence of bone destruction can be noted.
- b.** Bone windows showing evidence of bone erosion in occipital region.

Tumor invading transverse sinus was left in situ. Excision of the tumor was done with 2 cm of margin of dura and bone. Post operatively patient developed pseudo-meningocele which was managed conservatively. The histopathology evaluation was suggestive of an alveolar soft part sarcoma showing a nodular architecture of large cell nests separated by thin vascular setae

in a pseudo-alveolar pattern. Tumor cells were large, polygonal with abundant eosinophilic to clear cytoplasm, vesicular nuclei and prominent nucleolus. Infiltration of underlying bone was noted. Tumor cells were immunopositive for desman, negative for myogenin and cytokeratin. Mib-1 index was approximately 8% (Figure 3). Post-operative metastatic workup did not reveal

tumor anywhere in body. In view of small residual tumor and histopathological diagnosis of ASPS, he was advised radiation therapy for the residual lesion. At 14 months follow up, the patient was asymptomatic with no evidence of tumor growth or new lesions on follow up imaging (Figures 4a-4d).

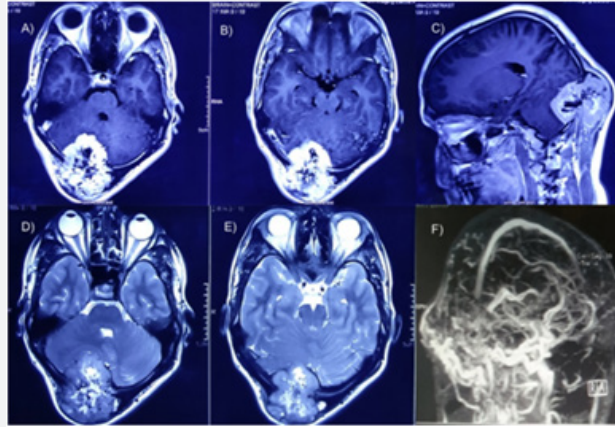


Figure 2: MRI Brain

A-C Contrast enhanced images showing intensely enhancing mass in occipital region with central necrotic areas.

D, E) T2W images showing a hyperintense mass with multiple flow voids and central hypointense area.

F) MR angiography images showing multiple tortuous vessels in the tumor and invasion of transverse sinus.

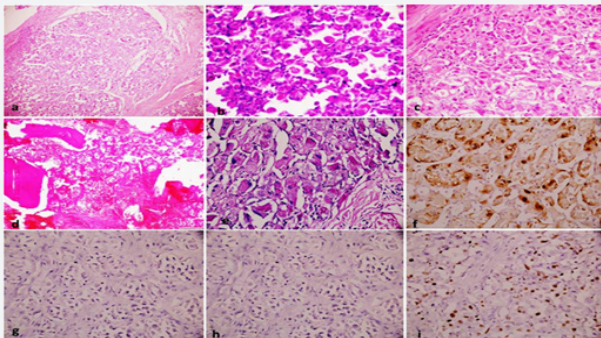


Figure 3: Photomicrograph showing a tumor displaying nodular architecture comprising of large sized nests separated by thin vascular setae

a. Pseudo-alveolar pattern resulting from loss of cellular cohesion and necrosis of the centrally located cells in the nest

b. Tumor cells are large, polygonal with abundant eosinophilic to clear cytoplasm, vesicular nuclei and prominent nucleolus. Multinucleated forms and occasional mitotic figures noted

c. Tumor infiltrating underlying bone

d. PAS stain demonstrates variable amount of glycogen and diastase-resistant granules in the cytoplasm

e. Tumor cells immunopositive for desman

f. Immunonegative for myogenin and cytokeratin

g. Mib-1 index of approximately 8 % (i, x400).

Discussion

Alveolar soft part sarcoma account for 0.5 to 1 % of all soft tissue sarcomas [3-9]. ASPS was first discovered by Christopher

son et al. [3]. They are slow growing indolent tumors with a peak incidence in the third decade [3]. Their histopathogenesis has been a source of considerable debate. Although early evidence supported a neural origin, more recent evidence points toward an origin from muscular tissue. Christopher son et al has suggested that ASPS may originate from the muscle spindle, which contains both intramural muscle fibers and nerve tissue [3]. Welsh et al. [4] hypothesized that ASPS arise from displaced paraganglionic mesoderm and have close homology with paragangliomas of carotid body. Primary intracranial ASPS are very rare and only eight other cases have been reported to date in English literature [5-11] (Table 1). Whether intracranial ASPS are primary or a manifestation of hidden primary somewhere in periphery is a matter of debate and long term follow up of some patients have shown it to be a metastasis from primary elsewhere [12]. Bony erosion is commonly seen.

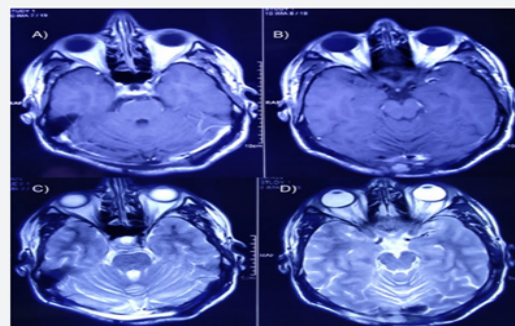


Figure 4: MRI Brain.

A, B: Post-operative contrast enhanced images showing small residual tumor along transverse sinus.

C, D: MRI T2 axial images post-operative.

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These tumors are usually hypo to is-intense on T1 and hyper intense on T2 weighted images with intense post contrast enhancement, multiple flow voids. Contrast enhancement is heterogeneous and central necrosis is usually present [1]. Lesions with similar findings on MRI like hemangioma, metastatic melanoma, liposarcoma, other soft tissue tumors with bleed or clear

cell sarcoma should be considered [13]. The intracranial lesions present as a hypervascular lobulated mass. These tumors may be dural based or may be found intraparenchymal in few cases (Table 1). The presumptive diagnosis on imaging is usually a meningioma and the diagnosis of ASPS comes as a surprise on his to-pathological examination as in our case.

Table 1: Review of Literature.

Author	Age / Sex	Symp.	Location	NCCT	MRI T1W	MRI T2W	Contrast enhancement	Extent of resection	RT	CT	Mets	Follow Up	Outcome	Pre-op Diagnosis	Remarks
Perry JR et al. [11]	28/ M	Partial seizures	Left Frontal				Homogenous	GTE	WBRT	CT	Lung	18 m	Alive 18 m	Meningioma	
Sujit Kr et al. [12]	28/ M	Headache, diplopia, GTCS, 7th nerve palsy	Left frontal, Basal ganglia, Rt. Parietal				CE +nt	STE (Intraparenchymal)	WBRT		Lung	18 m	Alive 18 m	Abscess/ Metastasis	
Bodiet al. [4]	39/ M	Seizures	Left temporal meningial		Isointense	Hyperintense	Homogenous	GTE			CXR ?TB	10 m	Alive 10 m	Tuberculoma à Meningioma	ASPS TFE3+ve
Das et al. [7]	17/ F	Mass	Bifrontal	Hyperdense mass; -Bone erosion				GTE	RT		Nil	4 m	Alive 4m		
Ahn et al. [1]	9/ F	Tinnitus, Headache, Ataxia	CP-angle		Hypointense	Hyperintense with -flow -voids	Homogenous	Embolisation à STE + RT à NTE +GKRS + CT (ICE)	RT/GK twice	CT	Nil	29 m	Recurrence à GK à New lesions	Meningioma	Refused further treatment. ASPS TFE3+ve
Mandal et al. [10]	32/ F	Headache, Diplopia	Rt. Parietal meningial		Hypointense	Hyperintense					Nil	NA	NA	Meningioma	
Tao et al. [13]	28/ F	Mass	Frontal	Bone-erosion	Isointense	Isointense	Moderate	GTE	RT		Nil	27 m	Alive 27 m	Invasive Meningioma/ Haemangioma	ASPS TFE3 +ve
	13/ M	Tinnitus, Proptosis	MCF	Bone-erosion	Isointense	Isointense	Moderate	STE	RT		Nil	24 m	Died at 2 yrs	Invasive Meningioma	ASPS TFE3 +ve
Our patient	24/ M	Mass	Occipital	Bone-erosion	Isointense	Hyperintense with-multiple flow -voids	Intense	NTE (Residual in transverse sinus)	RT	-	Nil	14 m	Alive at 14 m with stable residual	Meningioma	

Histo-pathologically they display spindle cells arranged in pseudo-alveolar appearance with abundance of eosinophilic granules in cytoplasm. Immuno-histochemically they are negative for epithelial markers like EMA and CK, synaptophysin, S-100, HMB-45, Melan-A and chromogranin. Desmin is positive in 50% and vimentin in 30 to 50%. Nuclear TF3 is positive and can help in diagnosis [14].

They exhibit cytoplasmic crystals which are PAS positive and diastase resistant. These crystals can be seen on electron microscopy and are complexes of protein mono-carboxylate transporter 1 and CD147.8) FISH shows characteristic unbalanced translocation der(17)t(X;17)(p11.2;q25) in ASPS. This translocation fuses TFE3 transcription factor gene at Xp11.2 to alveolar soft part locus (ASPL), a novel gene at 17q25 [14]. Due to the rarity of primary intracranial ASPS, treatment recommendations are difficult to arrive at. In a review of the nine cases of primary ASPS reported so far, patients with gross total excision of tumor seem to have fared better than those with subtotal excision. Most (7/9) of the patients underwent radiotherapy while chemotherapy was given only to two patients. One patient in the above reported cases received GKRS but continued to show tumor growth and developed new lesions on follow up (Table 1). In view of the indolent course of the disease, long term survival in ASPS is possible. The aggressive removal of all accessible brain lesions is recommended in patients with ASPS who are not terminally ill and can result in a particularly favorable prognosis [15]. It is important to keep the patients under long term clinico-radiological follow-up.

Conclusion

Primary intracranial alveolar soft part sarcoma is a rare entity. The diagnosis usually comes to light only after histopathological examination with most of them suspected to be meningiomas. Complete excision of the tumor appears to be associated with better prognosis. Radiotherapy or gamma knife radiosurgery can be given as adjuvant therapies. Patients should be kept under long clinico-radiological follow up as primary lesion in the trunk / limbs have been discovered at long follow up.

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