

Alveolar Soft - Part Sarcoma- MR Imaging Features

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Abstract: Alveolar soft part sarcoma (ASPS) is an extremely rare malignant soft tissue tumor seen more commonly involving the extremities in young females . Though slow growing, the tumor often shows the presence of metastases at the initial presentation itself in many cases. We present the MRI findings in two histologically proven cases of ASPS involving the thigh and the other affecting the orbit.

Key Words: alveolar soft part sarcoma- thigh- orbit

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I. Introduction:

Also known as malignant myoblastoma, alveolar soft part sarcoma (ASPS) gets its name due to its typical arrangement of cells in the form of pseudoalveolar or organoid arrangement¹. It is a slow growing highly vascular of the soft tissues seen mostly in adolescents and young females. The most common site is the deep soft tissues of thigh. Rarely it can involve mediastinum, genital tract, breast, gastrointestinal tract. In children ASPS are seen mostly in orbit and tongue. These tumors are generally slow growing , but has a poor prognosis due to early development of metastases and high incidence of recurrence .

II. Case Details:

Case 1.)

A 16 year old boy presented with soft tissue mass from right lower conjunctiva with associated bleeding since two months. The vision was poor in the right eye . There was no previous history of trauma to eyeball . General condition of the patient was normal. MR imaging of the orbits showed a 7 x 14 mm sized enhancing soft tissue mass from right lower eye lid. There was another soft tissue intraconal retro orbital mass of size 27 x 27 x 23 mm (Fig 1A,B,C). The mass was slightly hyper intense on T1 weighted sequence and hyper intense on T2 weighted sequence with intense homogenous contrast enhancement. Multiple flow voids are seen within the lesion. The optic nerve is compressed and displaced laterally. The medial wall of the orbit was thinned out. The lesion produced mild axial proptosis. There was no intracranial extension. A provisional diagnosis of lid tumor with orbital metastases was made. Biopsy from the entire tumor showed grey brown mass with ulcerated surface with the stroma showing an infiltrate of large polygonal cells with pleomorphic nuclei and abundant vacuolated cytoplasm. Multinucleate tumor giant cells were also seen along with mitosis. PAS positive and diastase resistant crystals were seen in the cytoplasm of few cells. The nuclei were large with 1-3 prominent nucleoli. There are interspersed dilated small blood vessels in between the tumor cells. The above features were consistent with alveolar soft part sarcoma, possibly a solid variant. (Fig 2 A,B,C)

Case 2.

A 22 year old female presented with painless swelling of scalp of one month duration. There was no head ache , seizures or loss of consciousness. Twenty days later she noticed a painful swelling of right thigh which was increasing in size. There was no trauma or restriction of limb movements. Clinical examination revealed a 2 x 3 x 3 cm mass arising from the left parietal bone was also seen . The mass was slightly hyperintense on T2W and isointense on T1 W sequences with erosion of parietal bone.

MRI of the thigh revealed the presence of a well defined mass of size 8 x 5 x7 cms within the muscle planes of anterior compartment of right thigh. The mass was hypo intense on T1 W and hyper intense on both T2W and fat suppression sequences of MRI and showed heterogeneous enhancement with contrast. Multiple flow voids were also seen in the inferior aspect of the tumor (Fig 3 A,B) . FNAC and biopsy from the scalp swelling showed islands of tumor cells and adjoining fibrocollagenous tissue. Tumor cells were arranged in alveolar nests separated by thin walled fibrovascular septae. These cells were polygonal, lining the inner wall of the alveoli with moderate well defined granular eosinophilic cytoplasm. The nuclei show single prominent nucleoli. Few cells were seen lying singly scattered in the centre of the alveoli. Occasional binucleate cells were seen. Diagnosis of alveolar soft part sarcoma was given. (Fig 4 A,B,C D)

III. Discussion:

ASPS is an extremely rare tumor with an incidence of less than 1 % of all malignant soft tissue tumors. It is a slow growing tumor in the soft tissues of mostly young females. The most common sites of involvement being extremities but also reported in other rare sites occasionally² . In about 60% of patients the lesion presents initially along with metastases³ . The CT scan images are mostly nonspecific. On MR these tumors are well defined soft tissue masses with slight hyperintensity on T1 weighted and heterogeneously hyperintense on T2 weighted images and enhances profusely with contrast administration⁴. The lesion show numerous flow voids within the mass. Another differentiating point in these tumors is the presence of profound solid component unlike other vascular tumors. The common lesions that are considered in the differential diagnosis include highly vascular metastases, hemangiopericytoma, vascular soft tissue sarcomas and hemangioendotheliomas. The latter tumors are mostly infiltrative and show fluid –fluid levels on imaging due the presence of hemorrhage which is not usually seen in ASAP. M. Beth McCarville et al⁵ analysed the imaging features in 22 histologically proven cases of ASAP and concluded that a lobulated enhancing soft tissue tumor with thick enhancing peripheral rim around central necrotic area arising from extremities in children is suggestive of ASAP. On reviewing the clinic pathological and imaging features of 6 cases of ASAP, Peng –Fei QIAO and others⁶ found that though MRI findings are nonspecific but they can suggest the evidence of malignancy and aid in the preoperative treatment of the cases. Immunohistochemistry with CathepsinK ,desmin and muscle-specific actin are strongly positive in these tumors with IHC for TFE 3 being specific. Cytogenetics of alveolar soft part sarcoma is typically characterized by a specific chromosomal alteration del(17)t(X:17)(p11;q25) which results in transcription factor E3 fusion with alveolar soft part sarcoma region 1 at 17q25.⁷ The tumor often metastasizes to brain , lungs , bone and even after 30 years.⁸ . The most unusual findings in our two cases are that the primary tumor of the eyelid presented with retro orbital metastases at the initial presentation which is extremely rare. The other unusual finding in the other case was the development of calvarial deposit with intra-axial component within six months of the initial presentation of the thigh mass. The treatment of choice is radical resection of the primary tumor and also the metastatic deposits also. The prognosis is relatively poor. Conclusion: the imaging findings of these tumors are nonspecific and histological confirmation With immunohistochemistry and cytogenetics if available is very important for proper diagnosis .



Fig. 1 A)



Fig 1.B)



Fig 1 .C)

Fig 1 ABC) : Axial contrast MR images of orbit show enhancing mass from the right lower eye lid and lobulated retro orbital , intraconal mass compressing the optic nerve .

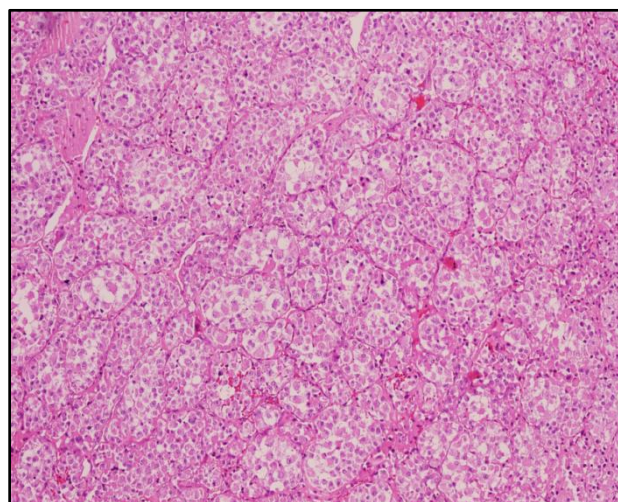


Fig 2 A)10x

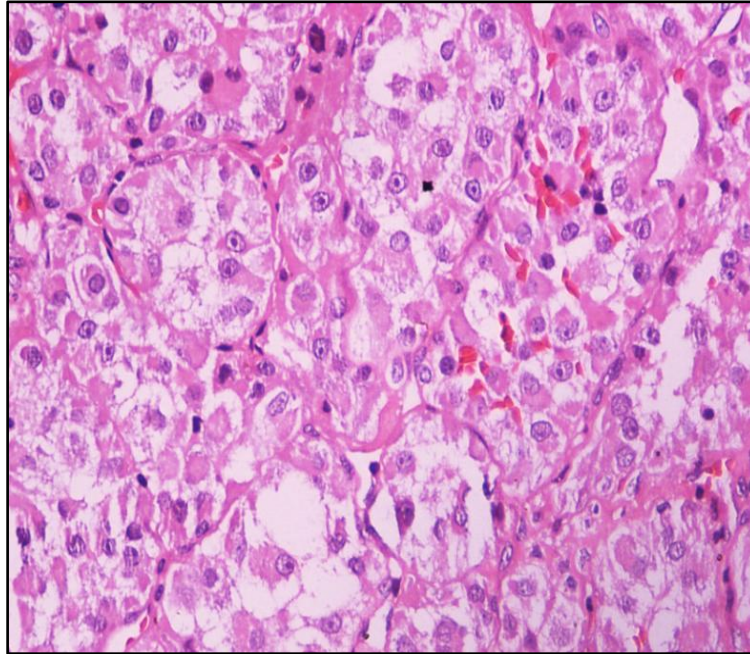


Fig2b.H&E 40x

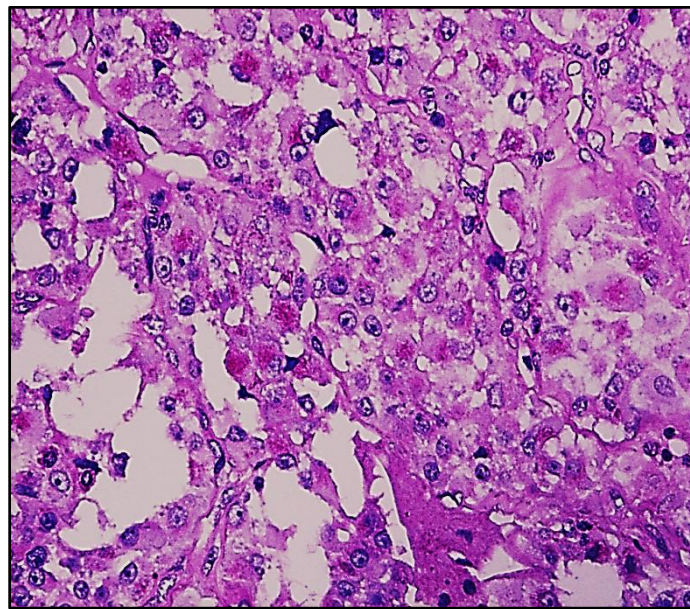


Fig 2 C .PASD 40x

- Fig2a. 10X magnification of H & E section showing alveolar pattern.
- Fig2b. 40X magnification of H&E section showing cells with abundant cytoplasm and vesicular nuclei with prominent nucleoli.
- Fig2c. 40X magnification showing PAS positive diastase resistant crystals and granules in cytoplasm



Fig 3. A)



Fig 3.B)

Fig3 A) coronal MR image of thigh shows an enhancing large soft tissue mass with in the muscle planes with multiple flow voids in the lower portion.

Fig 3.B) sagittal MR image of brain shows soft tissue mass in the parietal region with bone destruction.

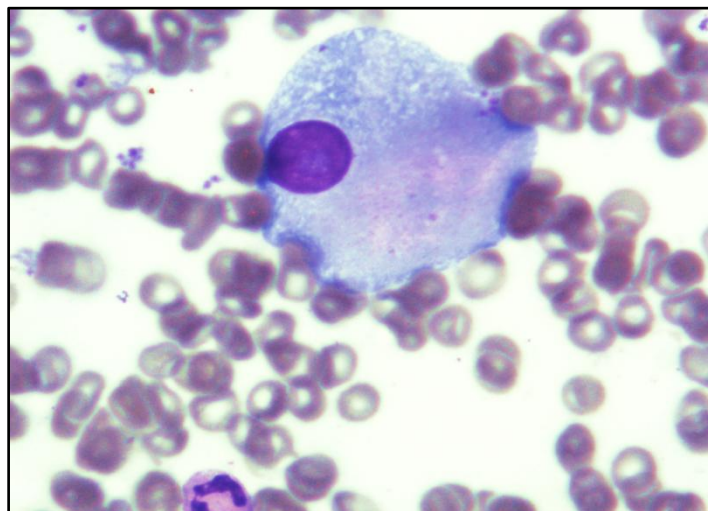


Fig.4 A). FNAC Scalp

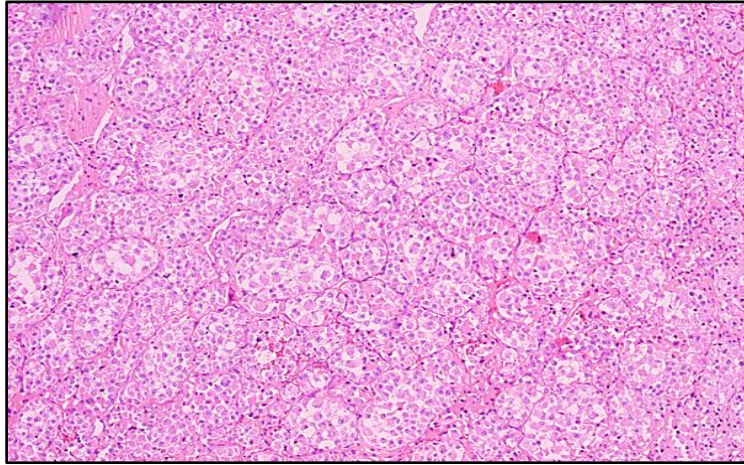


Fig.4.B) H&E, 10x

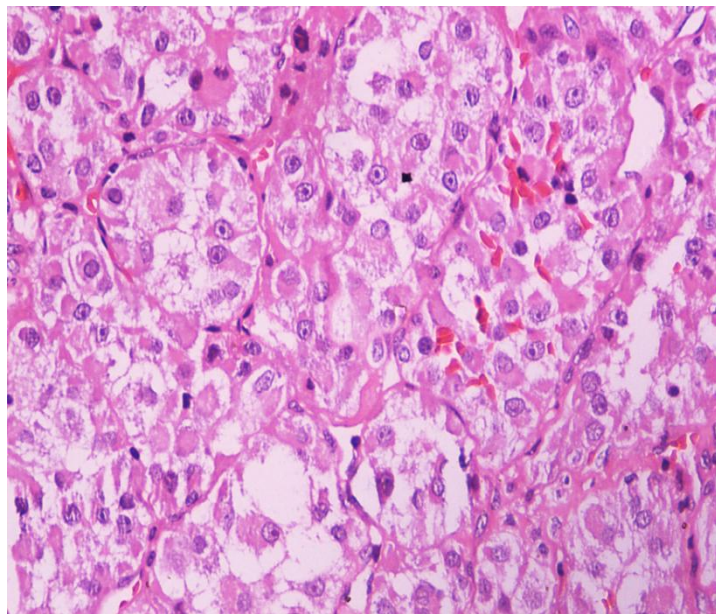


Fig.4 c.) H&E, 40x

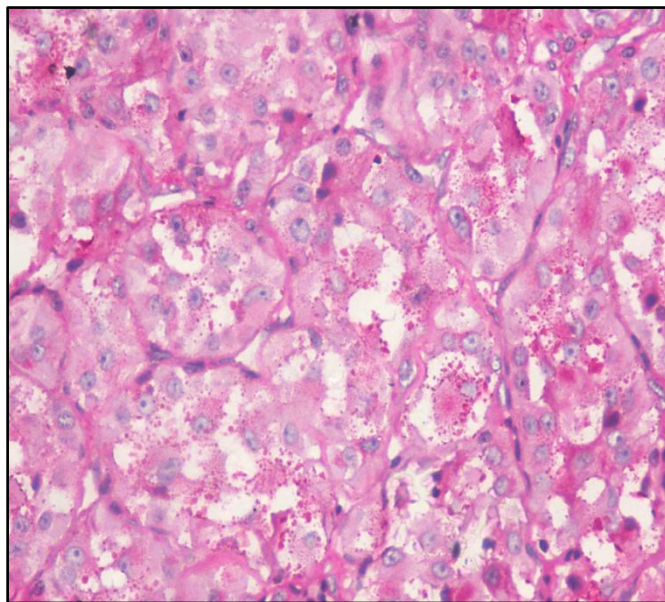


Fig.4.D) PASD

- Fig 4.A). Giemsa stained smear of FNAC scalp showing large cells with abundant cytoplasm and vesicular nuclei, 100x magnification.
- Fig 4.B) H & E section of scalp biopsy at 10x magnification showing alveolar pattern.
- Fig 4.C) H & E section of scalp at 40x, magnification showing nests lined by large polygonal cells with abundant granular cytoplasm.
- Fig 4 D) Section showing PAS positive diastase resistant cytoplasmic granules in cytoplasm

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