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Alveolar soft-part sarcoma (ASPS) of Right Iliac bone: A Case report and Review of literature

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ABSTRACT

Alveolar soft-part sarcoma (ASPS) is a rare type of malignant soft-tissue tumor. It is highly vascular tumor and affects primarily soft tissues in young adults. This tumor can affect any part of body, but is generally observed in the deeper tissues of the lower extremities. Bony involvement by this tumor is very rare. The present study reported a case of ASPS of the right iliac bone. The 18 year old male patient presented with a 1-year history of a mass on the right gluteal region which showed rapid growth in the last 4 months. Clinico-radiological examinations were done to rule out any other primary site. This tumor was excised and submitted for histopathological examination. The diagnosis was verified as Alveolar soft-part sarcoma (ASPS) by its characteristic clinical, radiological and histomorphological features however no immunohistochemical analysis was done. No distant metastases were noted

Keywords: Bone tumor, Gluteal mass, Sarcoma.

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a very rare tumor. It accounts for approximately 0.5%–0.9% of all soft-tissue sarcomas in adults. Out of these 0.8–1.8% of tumors found in children [1] and it is more commonly seen in young females [2]. It is a slow-growing tumor and affects mainly the lower extremities in adults and predominantely head and neck region in children [3]. Orbit, oral cavity and tongue are mainly involved organ in head and neck region. In adults involvement of upper extremities has been rarely observed. This tumor is very vascular and carries high risk for metastasis by the hematogenous route. The lungs (42%) are the most commonly involved organs by metastasis and sometimes it may be the only presenting feature. Other organs which can show metastasis are bones (19%), brain (15%), and lymph nodes respectively [4]. Breast, larynx and other organs are also reported to be the rarest site of this tumor. The differential diagnosis includes vascular malformations and highly vascularized soft tissue tumors. Fibrosarcoma, arteriovenous malformations, hemangioma, metastatic hypernephroma in a young patient. Alveolar soft part sarcoma (ASPS) is a malignancy with low incidence and poor prognosis. Children have a better overall survival as compared with adults. Surgical excision with tumor free margins is accepted as the treatment of choice for this tumor. The present study reports a case of ASPS of the right gluteal region involving bone which is evident

radiologically. Clinic-radiological examinations confirmed it to be a primary tumor arising in the the ilium and not from metastasis from other site. Final diagnosis was made on the basis of radiological and histopathological features. However this patient could not be followed up further.

Case presentation:-

An eighteen year old young male presented with mass on the right gluteal region that was present for last 2 years, but had rapidly grown over the last 3 months. The patient gave a history of associated pain and significant weight loss during this period. There was a history of fever since two months. No other significant history was present. On examination, a localized, well defined, tender mass was noted over right gluteal region. It was firm in consistency and fixed and had a size of approximately 4×3.5 cm. Sensory and motor examinations of the right lower limb were normal as well as other systems were also within normal limits.

On radiological examination X-ray Pelvis with both hips AP view showed soft tissue density in right iliac region. No evidence of calcification or bony destruction is noted (**Figure 1a**). X-ray Chest PA view is showing normal lung fields with no evidence of metastasis. (**Figure 1b**) MRI Post contrast T1FS coronal image showing well defined homogenous enhancing soft tissue mass lesion in right iliac fossa involving right iliac bone and iliac crest (**Figure 2a)**. STIR axial image showing well defined STIR hyperintense signal soft tissue mass lesion in right iliac fossa involving right iliac bone and iliac crest involving right iliacus muscle. No evidence of calcification is noted (Figure **2b)**. PET scan shows metabolically active mass lesion in right iliac fossa region. No evidence of metabolically active distant metastasis is noted in liver, chest, ribs and vertebrae (**Figure 3a)** . Excision biopsy from this mass was taken and sent for histopathological examination. On histopathological examination it showed charcterestic alveolar pattern comprising of malignant tumor cells with eosinophilic vacuolated cytoplasm, disposed in small clusters, sheets and nests. The tumor cells are having vesicular and hyperchromatic nuclei with prominent eosinophilic nucleoli. The tumor cell nests are separated by fibrous tissue . Rare mitotic figures with minimal pleomorphism is seen **(Figure 3b & Figure 4a, 4b, 4c)** .All these features are suggestive of ASPS. This patient could not be followed up further. Our case might contribute to an increased awareness of this rare disease by stressing the involvement of bone which is extremely rare kind of presentation of this disease.

Discussion:-

ASPS are one of the rarest soft tissue sarcomas and primary tumor arising from bone is further very rare. [5] Its histogenesis is still unknown . It is generally believed to account for <1% of all soft-tissue tumors [6]. This neoplasm usually arises in the soft tissues of the lower limbs in adults and in the head and neck region in children. The mean age at diagnosis is about 22 years in females and 27 years in males, but the tumor can occur in children as young as 2 year old. It presents primarily as a slowly growing mass or as metastatic disease . The most common site of origin is the lower limb, followed by the trunk and the upper limbs. Few cases are also seen at other locations such as arm, chest and retroperitoneal tissues. In children few cases occurring in the head and neck primarily involving the orbit and tongue have been reported. Despite the slow growth rate of the primary tumor, metastases are common. They are detected in about 20-25% of patients at the time of diagnosis. Metastases are found most often in the lungs, followed by bone and brain [7]. It typically present as a very vascular tumor on imaging studies [8]. Histological features show characterestic alveolar pattern separated by thin walled vascular channels. Cells are polygonal with vesicular nuclei with prominent nucleolei and eosinophilic granular cytoplasm which contains PAS positive material. [9] It is characterized by a specific chromosomal alteration, der(17)t(X:17)(p11:q25), resulting in fusion of the transcription factor E3 (TFE3) with alveolar soft part sarcoma critical region 1 (ASPSCR1) at 17q25 . This translocation is diagnostically useful [10]. This tumor has high metastasis rate with relatively indolent clinical course and poor prognosis. The prognosis of five patients with primary ASPS of bone has reported to be poor.[11]. Treatment is mainly surgical along with adjuvant chemotherapy. [12]

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Volume 2, Issue 5; September-October 2019; Page No.121-125

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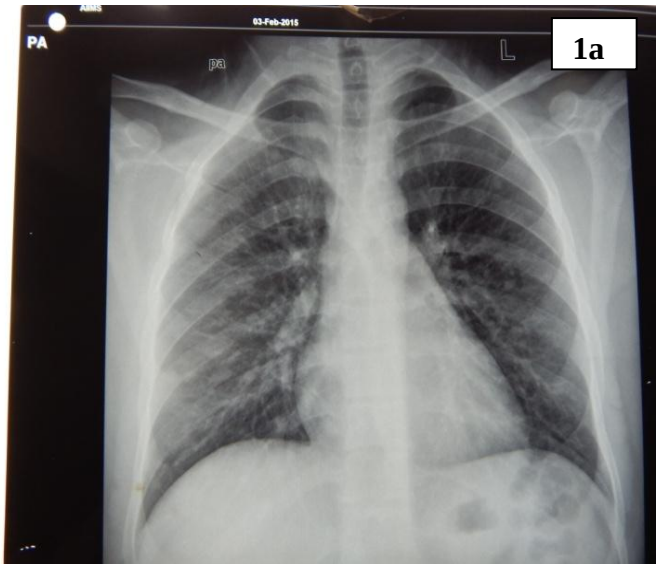


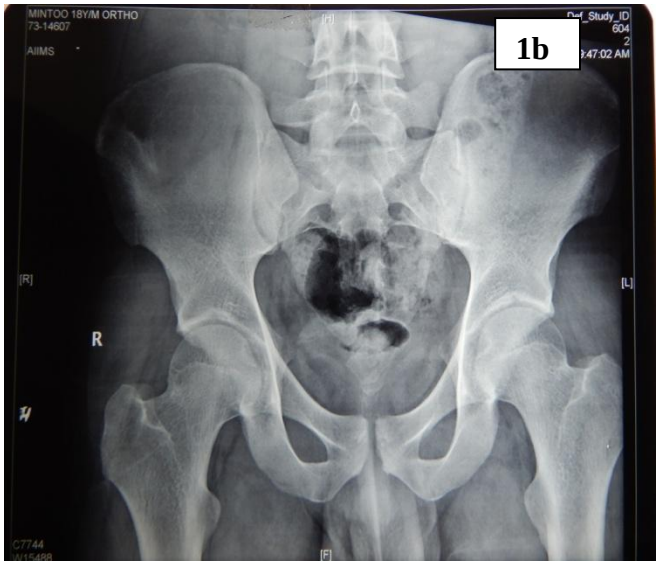
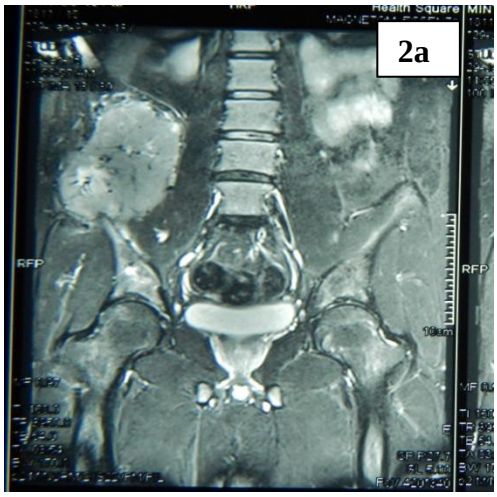


Figure 1a X-ray Chest PA view showing normal lung fields. **1b** X-ray Pelvis with both hip AP view showing soft tissue density in right iliac region. No evidence of calcification or bony destruction is noted

Volume 2, Issue 5; September-October 2019; Page No.121-125



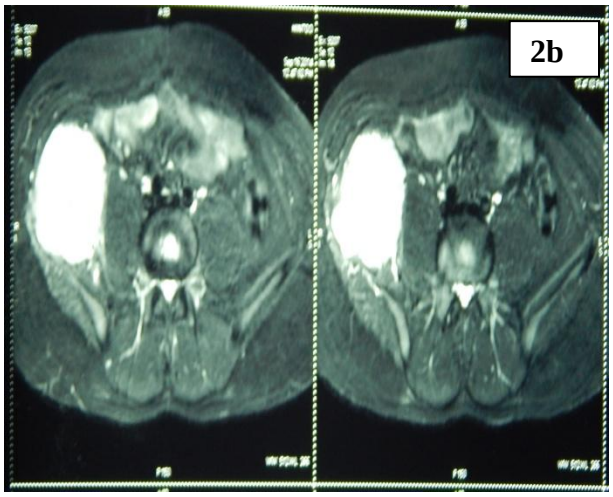
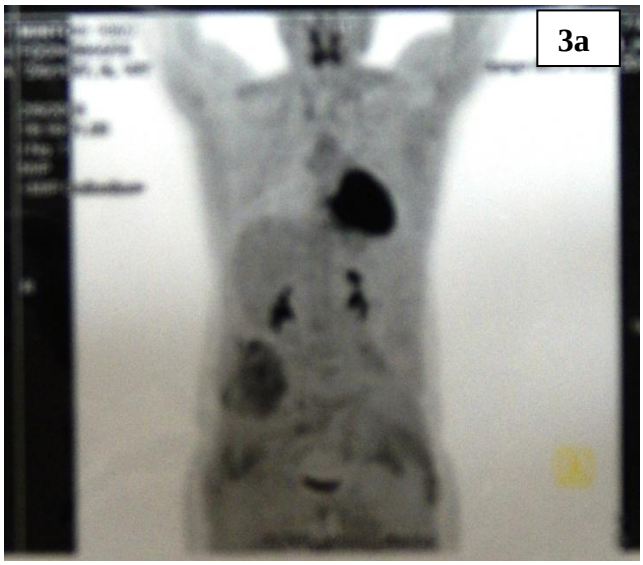


Figure 2a MRI Post contrast T1FS coronal image showing well defined homogenous enhancing soft tissue mass lesion in right iliac fossa involving right iliac bone and iliac crest with multiple small internal flow voids. **2b** STIR axial image showing well defined STIR hyperintense signal soft tissue mass lesion in right iliac fossa involving right iliac bone and iliac crest involving right iliacus muscle. No evidence of calcification is noted



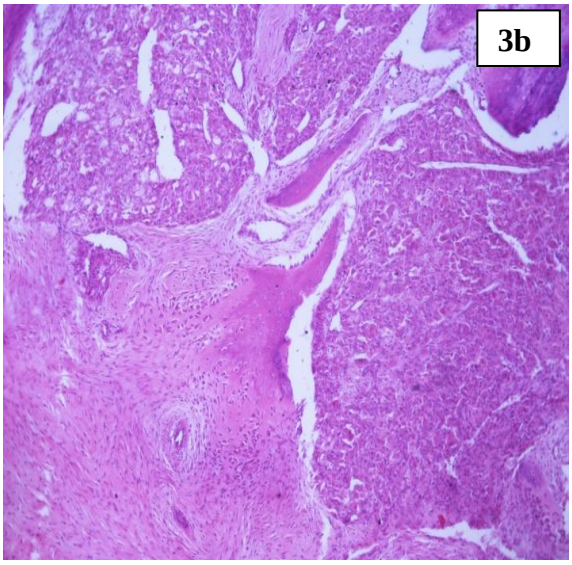


Figure 3a PET scan image showing metabolically active mass lesion in right iliac fossa region. No evidence of metabolically active distant metastasis is noted in liver, chest, ribs and vertebra. **3b** Photomicrograph showing malignant tumor disposed in sheets and alongwith adjacent fibro-osseous tissue and necrosis. (H&E; 10X).

Volume 2, Issue 5; September-October 2019; Page No.121-125

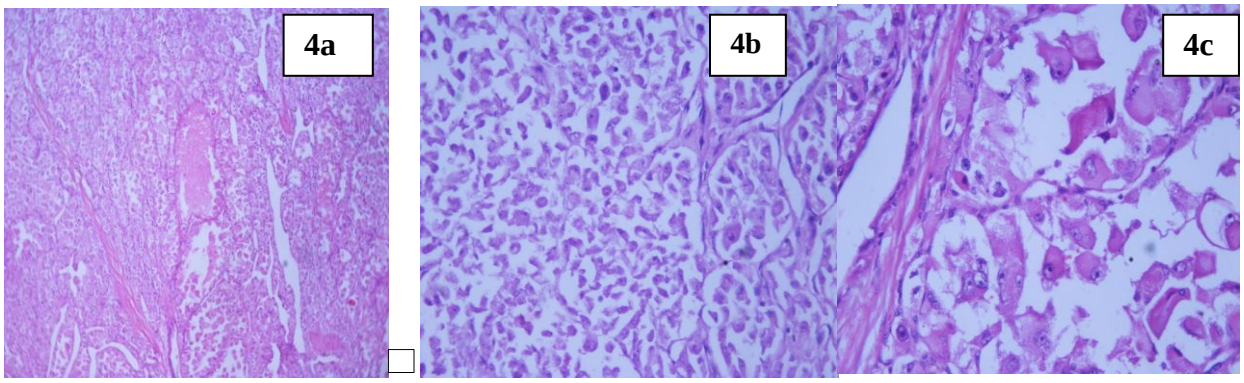


Figure 4a &4b Photomicrograph showing alveolar pattern comprising of malignant tumor cells with eosinophilic vacuolated cytoplasm, disposed in small clusters, sheets and separated by fibrous tissue. Rare mitotic figures with minimal pleomorphism is seen (H&E; 10X and H&E; 20X). **4C** Photomicrograph showing tumor cells having vesicular and hyperchromatic nuclei with prominent eosinophilic nucleoli . (H&E; 40X)

Volume 2, Issue 5; September-October 2019; Page No.121-125