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Respiratory symptoms as first manifestation in an occult alveolar soft part sarcoma

Sir,

A 23-years-old male presented in chest diseases out-door patient department with complaints of cough and occasional breathlessness for the last 4 weeks. X-ray chest showed nodular opacities in lower halves of both lungs [Figure 1]. All his hematological investigations including erythrocyte sedimentation rate and sputum examination were normal. CT scan of chest showed multiple nodules in both lungs with possibility of the diagnosis of metastasis [Figure 2]. After suspicion of the multiple metastases in lungs, CT scan of the abdomen and pelvis was performed to find out the possible primary site and/or the metastasis in the abdominal organs. During this investigation, a soft tissue mass in the left gluteal region was detected. There was an ill-defined heterogeneous mass involving the left gluteal medius and minimus muscle [Figure 3]. MRI of this region showed the heterogeneous mass of size 6.5 × 5.5 × 5.0 cm with poor capsulation and increased vascularity [Figure 4]. Tc-99 methylenediphosphonate (MDP) bone scan showed no skeletal metastasis. Fine needle aspiration cytology showed dyscohesive cells with an eccentrically located nucleus, nuclear pleomorphism, and variable nuclear chromatin. These neoplastic cells showed a single cell population of epithelioid to plasmacytoid morphology [Figure 5]. The core needle biopsy confirmed the diagnosis of alveolar soft part sarcoma and showed a characteristic morphology with large tumoral cells in nests separated by delicate fibrovascular septae creating pseudo-alveolar pattern confirming the diagnosis of alveolar soft part sarcoma [Figure 6].

The tumor was widely resected and measured 6 × 5 × 5 cm in dimensions. This was poorly circumscribed. The cut surface was grey-white to grey-yellow and friable with areas of hemorrhage and necrosis. There were large tortuous vessels in the surrounding normal tissue [Figure 7]. The patient was treated with chemotherapy using doxorubicin and ifosfamide. However, the patient died within 18 months of treatment due to extensive metastasis and bilateral malignant pleural effusion.

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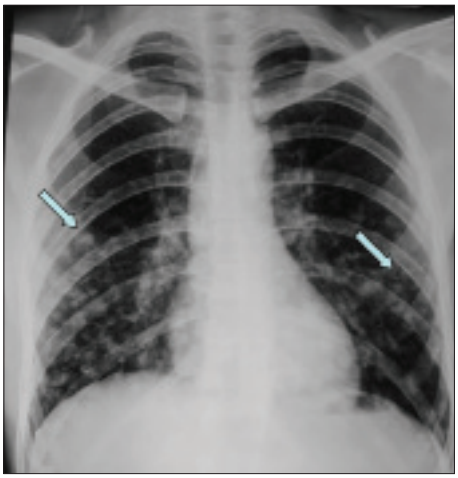

Figure 1: X-ray chest showing nodular opacities in both lungs

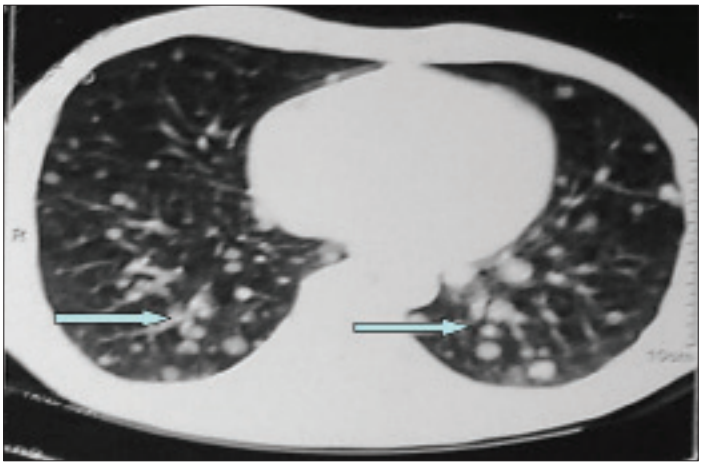

Figure 2: Computed tomography scan thorax confirming multiple nodular opacities in both lungs

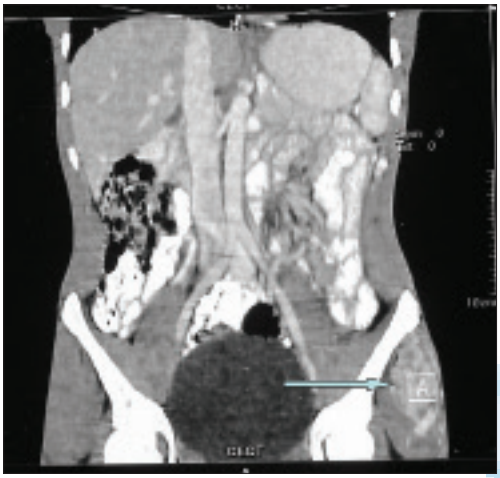

Figure 3: Computed tomography scan abdomen and pelvis detecting mass in left gluteal region

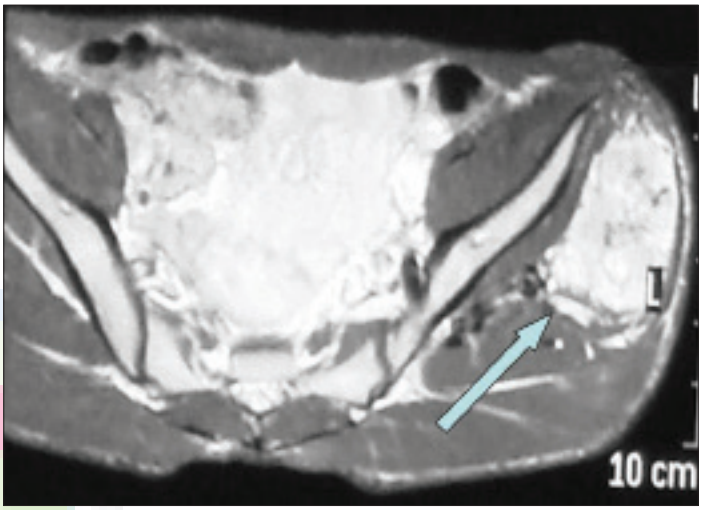

Figure 4: Magnetic resonance imaging T-2 weighted image showing the tumor in left gluteal muscles

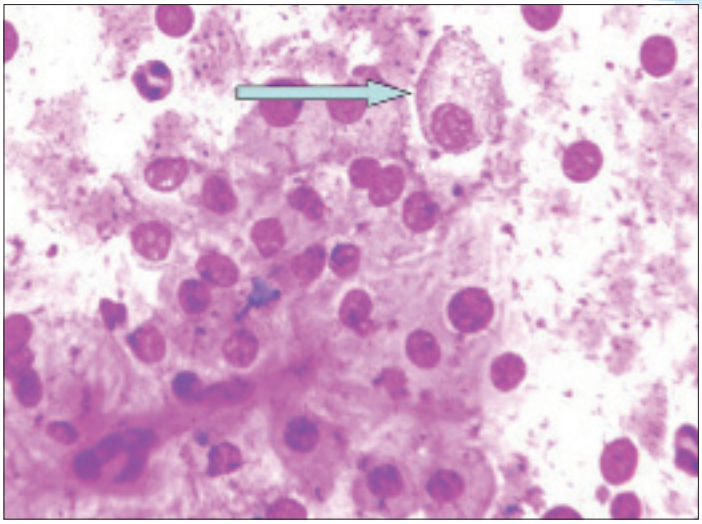

Figure 5: Fine needle aspiration cytology showing round to polygonal cells

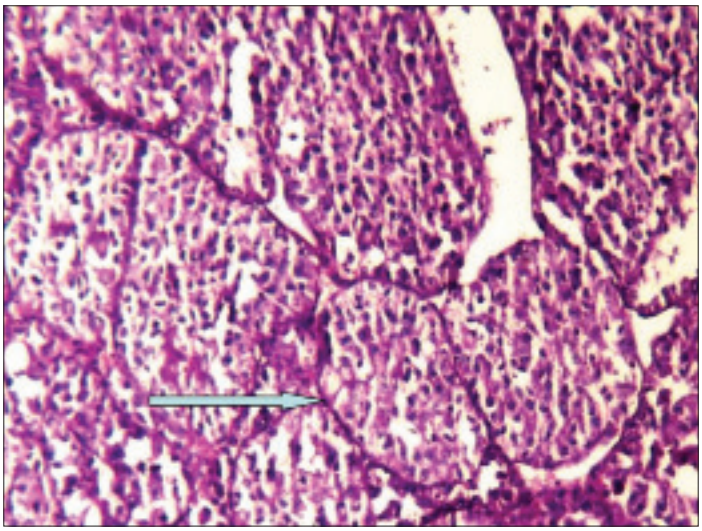

Figure 6: Histopathology showing pseudo-alveolar pattern

ASPS is a rare soft tissue tumor commonly involving the extremities of young adults and less commonly the trunk, head and neck, retroperitoneum, lung, mediastinum, stomach, and the female genital tract. This accounts for less than 1% of all soft tissue sarcomas.[1] This usually presents as a slowly-growing, painless mass, which can remain undetected due to lack of symptoms. Early metastasis is a characteristic feature of this tumor and, in a good number of cases, metastasis to the lungs or brain is the first presentation of the disease.[1-4] This young male patient being reported also first developed symptoms related to respiratory system, and the primary lesion remained asymptomatic, which could only be detected during the screening CT scan of abdomen and pelvis. During this imaging, the lesion in the gluteal region was detected and proved to be alveolar soft part sarcoma on histopathology.

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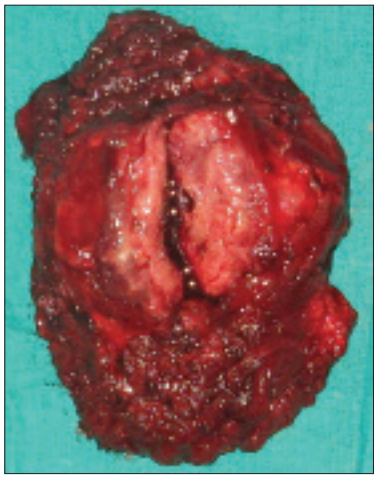

Figure 7: The widely excised specimen

We conclude that ASPS can remain undetected for a long period and can be easily overlooked due to relative lack of symptoms of the primary tumor. Early metastasis is a characteristic feature of this poor prognostic tumor and unfortunately, in large number of patients, it present with lung or brain metastasis as the first manifestation of the disease. In the cases of occult primary with metastasis to the lungs, ASPS should also be suspected as one of the possibility.

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