CASE STUDIES

Alveolar soft part sarcoma: A rare cytological impression

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Abstract

Alveolar soft part sarcoma (ASPS), a malignant soft tissue tumor with unknown etiology, predominantly occurs in the head and neck region, mainly the tongue and the orbit. The characteristic histopathological feature includes alveolar or pseudoglandular pattern, with PAS-positive, diastase-resistant intra-cytoplasmic inclusions. Primary therapy is total resection and adjuvant chemoradiation therapy. The present case study describes a rare case of ASPS of the anterior chest wall in a 15-year-old patient, diagnosed on fine-needle aspiration cytology. Total resection of the tumor with chemo-radiotherapy was performed and the follow-up after 2 years showed neither local recurrence nor distant metastases.

Keywords: alveolar soft part sarcoma, anterior chest wall, cytology

Introduction

Alveolar soft part sarcoma (ASPS) is an unusual malignant soft tissue sarcoma, accounting for 5% of the pediatric soft tissue sarcomas.[1] The exact nature of the tumor is uncertain, though some immunohistochemical studies indicate an origin from striated muscles.[2, 3]

ASPS occurs most commonly between 15-35 years of age, with female preponderance.[4] The tumor usually originates in the head and neck region, including the orbit and tongue, and the skeletal muscle of the extremities. Approximately 20% of the patients are diagnosed with metastasis to the lung, bone and central nervous system, on initial presentation. The tumor has an indolent course with prolonged disease-free intervals of 10 years.[3]

ASPS is a rare neoplasm of unknown histogenesis and poor prognosis.[5] Although several immunohistochemical stains are available to arrive at a diagnosis, the characteristic morphology of the tumor cells with periodic acid-Schiff (PAS) positive, diastase-resistant, intra-cytoplasmic inclusions, is considered the main diagnostic feature of alveolar soft part sarcoma.[6]

Case report

A 15-year-old male presented with a slow-growing mass on the anterior chest wall for 2 years, which rapidly increased in size during the last few weeks. There was no history of fever or trauma. A well-defined firm, non-tender mass measuring 8 × 6 cm with smooth surface was noted over the sternum. No palpable axillary lymph nodes were observed. On MRI, a well-demarcated heterogenous lesion was seen with no bony involvement. Fine-needle aspiration cytology (FNAC) from the swelling showed moderately cellular smears with scattered polyhedral tumor cells having abundant clear cytoplasm, central to eccentrically placed nucleus and prominent central nucleoli. Binucleate and multinucleate forms were also seen (Fig.1 and Fig. 2). A diagnosis of ASPS was considered, with differential diagnoses of metastatic renal (clear) cell carcinoma (RCC), clear cell sarcoma of tendon sheath, and granular cell tumor. Wide excision biopsy was performed. Histopathological examination showed tumor cells in well-defined nests in typical alveolar pattern, separated by fibrous tissue with PAS positivity (Fig. 3). The patient received cobalt-60 (Co-60) teletherapy 50 Gy and 6 cycles of adjuvant chemotherapy (VACA regimen), comprising of vincristine 1.0 mg/m^2, adriamycin 40 mg/m^2/day, cyclophosphamide 1000 mg/m^2 and actinomycin-D 0.5 mg/m^2. Follow-up after 2 years demonstrated that the patient was doing well with no evidence of recurrence or metastasis.

Fig. 1: Fine-needle aspiration cytology from the swelling showing moderately cellular smears with scattered polyhedral tumor cells having abundant clear cytoplasm with central to eccentrically placed nucleus. Binucleate and multinucleate forms were also seen. Hematoxylin and eosin x 10X.

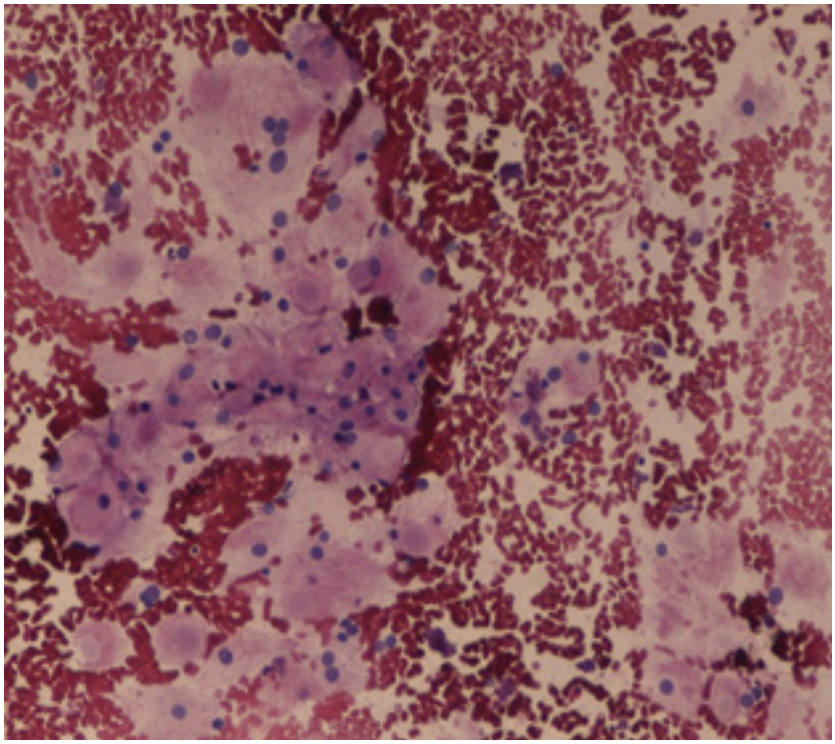


Fig. 2: Aspirate smears showing scattered polyhedral tumor cells with central to eccentrically placed nucleus and crystalloid cytoplasmic inclusions, with many binucleate and multinucleate forms. Hematoxylin and eosin x 40X.

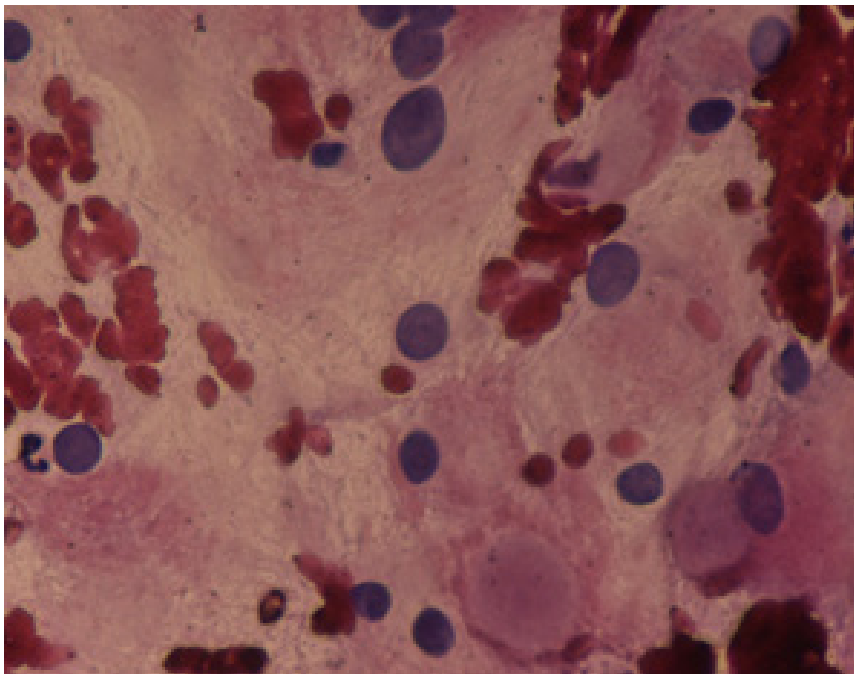
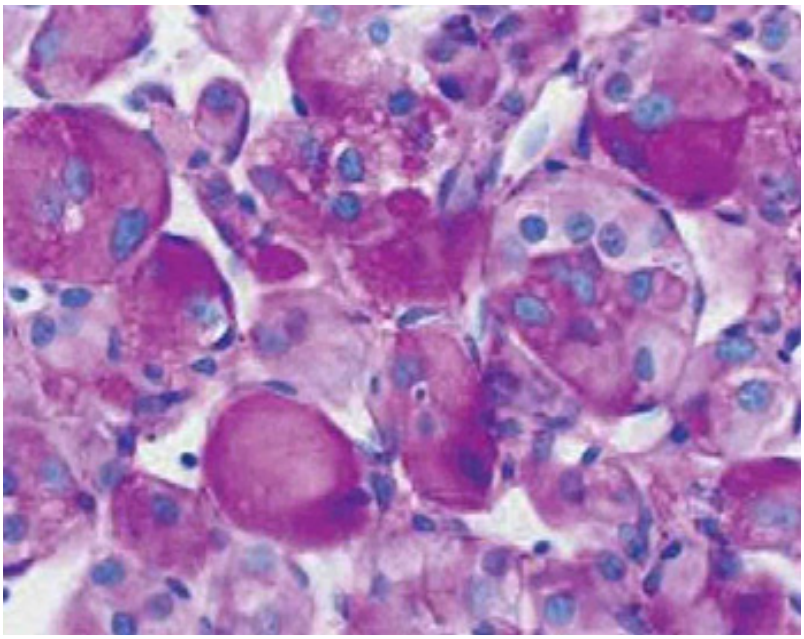


Fig. 3: Histopathological examination showing tumor cells in well-defined nests in typical alveolar pattern, separated by fibrous tissue with PAS positivity. Hematoxylin and eosin x 40X.



Discussion

ASPS occurs more commonly in young women, especially in the proximal extremities.[3] The tumor has an indolent and slow growth with late metastases. ASPS has a typical organoid alveolar grouping of cells, with abundance of capillaries and PAS-positive, crystalloid cytoplasmic inclusions.[6]

A lymphogenic metastasis of the alveolar sarcoma has been predominantly observed.[7] Histogenetically, the alveolar soft part sarcoma is assumed to be closely related to striated muscles. Microscopically two types of tumor cells namely dark and light are seen, while PAS-positive substances are noted only in the cytoplasm of the light cells.[8]

The differential diagnosis of ASPS includes granular cell tumor, renal cell carcinoma, malignant melanoma, hepatocellular carcinoma and adrenal cortical carcinoma.[9] Malignant melanoma and primary or metastatic renal cell carcinoma closely resemble ASPS microscopically; but they can be differentiated from ASPS by the absence of the characteristic PAS-positive crystalloid cytoplasmic inclusions and the HMB-45 and epithelial membrane antigen positivity respectively, in these tumors. Tumor cells of hepatocellular carcinoma have granular nuclear chromatin and nucleoli with intranuclear pseudoinclusions and presence of bile. Unlike ASPS, adrenal cortical carcinoma typically exhibits nuclear hyperchromasia and mitotic activity with immunohistochemical positivity for inhibin, calretinin, synaptophysin, and Melan-A. Glycogen is present in both ASPS and renal cell carcinoma, but absent in granular cell tumor. Furthermore, the cells of granular cell tumor have a distinct granular cytoplasm and are strongly positive for S-100 protein. Independent prognostic factors of ASPS include high-grade tumor, size ≥10 cm and deep-seated lesions. Tumor grade is a poor prognostic factor.[10]

Complete surgical excision is the mainstay of therapy and the principal predictor of outcome.[11] The use of adjuvant chemo-irradiation therapy is debatable. Patients are generally managed with a multi-modality therapeutic approach including surgery, radiotherapy and chemotherapy.[12] If conservative complete excision is unfeasible at the time of diagnosis, neoadjuvant chemotherapy is employed to reduce the tumor mass and improve resectability. Radiotherapy is administered to patients at high risk of local relapse due to incomplete resection. External beam irradiation is usually administered with fractionated doses of 50 Gy of Co-60. Chemotherapy is recommended in the presence of microscopic or macroscopic residual disease after surgery.

Competing interests

The authors declare that they have no competing interests.

Citation

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