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Letter to Editor

Alveolar Soft-part Sarcoma on the Abdominal Wall in an Adult: A Rare Presentation with Cytological and Histological Correlation

Sir,

We report a case of a male patient who presented with a painless swelling in the abdomen noticed 6 months back, with sudden increase in size over the past 3 months. On examination, there was a firm swelling of size 8 cm × 6 cm in the left hypochondrium, in the intramuscular plane. Contrast-enhanced computed tomography revealed a well-defined solid mass lesion with central hypodensity and heterogeneously intense postcontrast enhancement within the rectus abdominis muscle in the supraumbilical region. Imaging features suggested the possibility of a neurogenic tumor. Fine-needle aspiration revealed loose aggregate and dispersed large cells with eccentric nuclei, prominent nucleoli, and abundant granular cytoplasm [Figure 1a and b]. Many bare nuclei were seen. With the differential diagnosis of alveolar soft-part sarcoma (ASPS) and granular cell tumor, a wide excision was done to remove the tumor.

The specimen showed a circumscribed soft reddish-brown tumor of size 6.2 cm × 5.7 cm × 3.3 cm. Histology revealed large polygonal cells in nests and organoid pattern with delicate septa. The cells had abundant eosinophilic granular cytoplasm, eccentric round nuclei, and prominent nucleoli [Figure 1c]. Nuclear atypia, tumor giant cells, multinucleation, and extensive lymphovascular emboli were present. Periodic acid–Schiff (PAS) and PAS with diastase revealed tumor cells with intracellular crystalline material [Figure 1d]. The tumor sections were subjected to a panel of immunohistochemical markers including vimentin, desmin, pancytokeratin, chromogranin, CD10, myogenin, NSE, S-100, and Ki-67. The tumor cells were positive to vimentin [Figure 2a] and desmin [Figure 2b]. The other immunohistochemical markers were negative. These morphological and immunohistochemical features favored the diagnosis of ASPS.

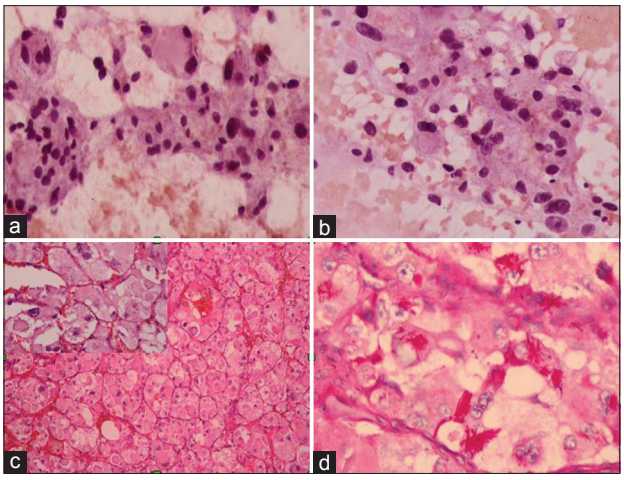

Figure 1: (a) Smear showing loose clusters of large cells with abundant granular cytoplasm (H and E, ×10). (b) High-power image showing frayed cytoplasm and large nuclei with nucleoli (H and E, ×40). (c) histology showing large cells in nests with delicate septa (H and E, ×10); inset showing granular cytoplasm, large nuclei, and prominent nucleoli (H and E, ×40). (d) Intracytoplasmic rod-like crystals (Periodic acid–Schiff with diastase, ×40)

Discussion

ASPS is a slow-growing malignant soft-tissue tumor of uncertain origin and usually occurs in young adults. It accounts for 1% of all soft-tissue sarcomas[1,2] with female preponderance. It is seen in the deep soft tissues of thigh or buttock in adults and the head and neck region, especially tongue and orbit, in children.[1,2] Jia *et al*. described ASPS on the abdominal wall in a 2-year-old child.[3]

Aspiration smears will show large dyscohesive monotonous cells, having abundant fine vacuolated to granular cytoplasm and round nuclei with prominent nucleoli. Stripped nuclei and fraying of the cytoplasmic margins are quite common. ASPS shows distinctive histomorphology of organoid or nesting pattern at low magnification, separated by delicate vasculature. Loss of cellular cohesion of the centrally located cells in nests results in pseudoalveolar pattern.[1,4] The neoplastic cells show distinct cell borders, granular cytoplasm, and central nuclei with prominent nucleoli. Multinucleation and atypia can be seen. Faint intracytoplasmic rod-shaped inclusions and granules seen in hematoxylin and eosin stain; highlighted by PAS stain after diastase digestion as rod like crystals is very characteristic.[1,2,4] The differentials to be considered are granular cell tumor, metastatic renal cell carcinoma, and clear-cell sarcoma of soft parts [Table 1].[5,6]

ASPS demonstrates consistent strong nuclear staining to Transcription factor binding to IGHM enhancer3 (TFE3), in addition to focal desmin and S-100 positivity. Electron microscopy and cytogenetics further helps confirm the

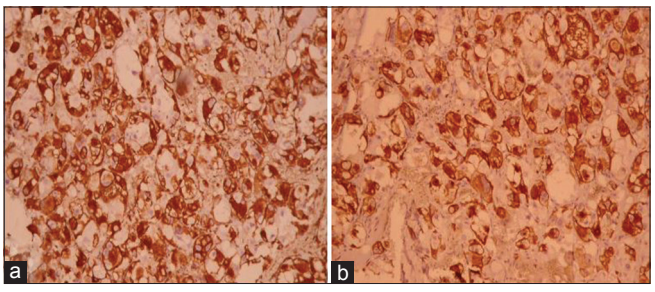

Figure 2: (a) Tumor cells showing positivity to vimentin (×40). (b) Tumor cells with desmin positivity (×40)

Table 1: Diagnostic criteria to differentiate alveolar soft-part sarcoma and its mimics

	Alveolar soft-part sarcoma	Clear cell sarcoma	Renal cell carcinoma	Granular cell tumor
Histology	Pseudo-alveolar pattern	Alveolar pattern	Alveolar pattern	Sheets and nests
	Granular cytoplasm	Clear cytoplasm	Clear cytoplasm	Dense granular cytoplasm
Periodic acid-Schiff stain	Intracytoplasmic rod-like crystals	Positive due to cytoplasmic glycogen	Positive due to cytoplasmic glycogen	Positive due to cytoplasmic phagolysosomes
Electron microscopy	Membrane bound rhomboid crystals	Melanosomes with barrel stave internal structure	Long microvilli and abundant cell junctions	Autophagic vacuoles
IHC				
Vimentin	++	++	++	++
Desmin	+	-	-	-
TFE3	++	-	-	-
S-100	+/-	++	+/-	++
HMB-45	-	++	-	-
CD68	-	-	-	++
EMA	-	-	++	-
CD10	-	-	++	-
Molecular studies	der(17)t(X; 17)(p11;25) ASPSCR1-TFE3 fusion transcript	t(12;22)(q13;q12)	VHL gene mutation	t(12;22)(q13;q12)

IHC – Immunohistochemistry; + – Positive; - – Negative; TFE3 – Transcription factor binding to IGHM enhancer3; HMB-45 – Human melanoma black; EMA – Epithelial membrane antigen

diagnosis, with distinctive membrane-bound rhomboid or rectangular crystals, composed of a periodical lattice work of rigid fibrils and ASPSCR1-TFE3 fusion transcript due to specific translocation der(17)t(X; 17)(p11;25).[1]

Surgical resection with clear margins is the appropriate treatment plan for ASPS. Chemotherapy and radiotherapy are under consideration to prevent local recurrence or metastasis. Age at presentation, tumor size, and the presence of metastasis influence the prognosis.[1,4]

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Conflicts of interest

There are no conflicts of interest.

Rajeshwari K Muthusamy, Sangita S Mehta

Department of Pathology, Kovai Medical Center and Hospital, Coimbatore, Tamil Nadu, India

Address for correspondence:
Dr. Rajeshwari K Muthusamy, Department of Pathology, Kovai Medical Center and Hospital, Coimbatore - 641 014, Tamil Nadu, India.
E-mail: drrajipranav@gmail.com

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