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# Epithelioid Sarcoma: A Retroperitoneal Mass

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## ABSTRACT

Epithelioid sarcoma is a rare tumour, counting for less than 1% of all soft tissue sarcomas. It is a slow growing, high grade, soft tissue malignancy of mesenchymal origin. It has marked propensity for local recurrence, regional lymph node involvement and distant metastases. Clinical diagnosis is difficult and radiological findings are nonspecific as well as inconclusive. Therefore, histopathological examination along with immunohistochemical markers are required for the confirmation of diagnosis. Here, we report a case of a 65 years old female, with retroperitoneal mass.

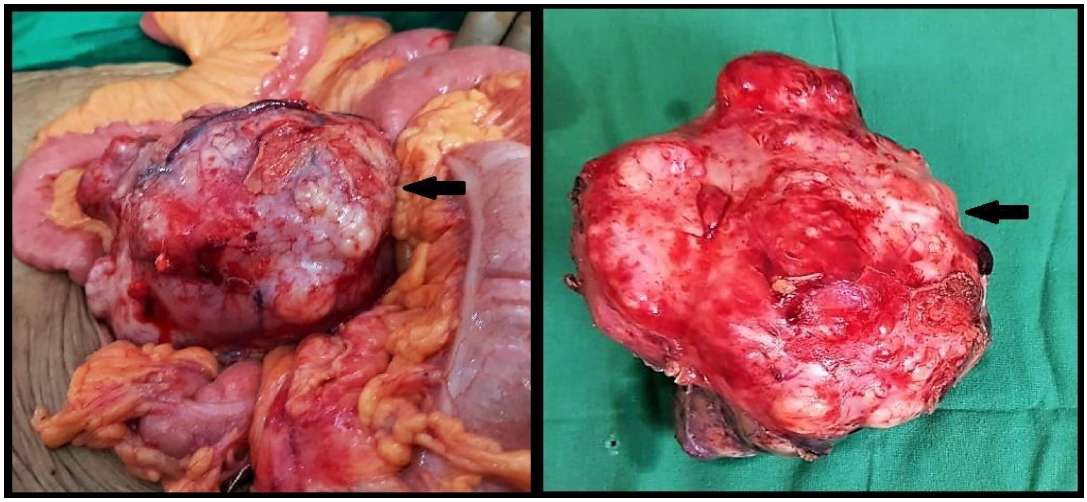
***Keywords:*** Epithelioid sarcoma, Retroperitoneal mass, Epithelioid cells, Spindle cells.

## INTRODUCTION

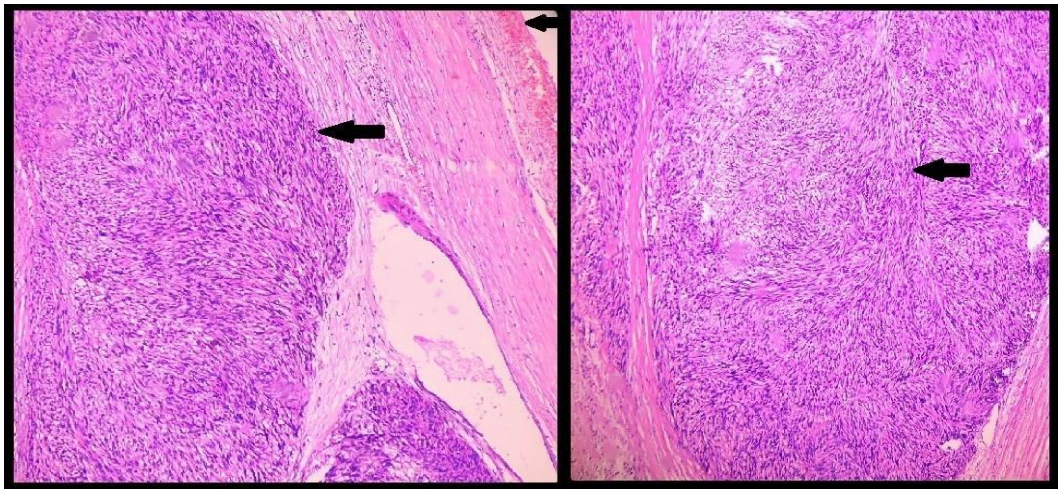
Epithelioid sarcoma (ES) is a rare tumour, counting for less than 1% of all soft tissue sarcomas. [1,2] It was first described by Laskowski in 1961 as "sarcoma aponeuroticum" because of its involvement of aponeuroses. [3] In 1970, Enzinger coined the term "epithelioid sarcoma" after recategorizing 62 previously misdiagnosed tumours. [3,4] Its peak incidence is seen among the males in their second or third decade of life. [2] The size varies from few millimeters to several centimeters. [5] There are two subtypes of ES namely the conventional/ classic/ distal type and the proximal type. [1] Microscopic appearance of ES ranges from spindle cells to large polygonal cells with an acidophilic cytoplasm [5] It shows reactivity for both epithelial and mesenchymal markers, such as cytokeratin (CK), epithelial membrane antigen (EMA), and vimentin. [5] Epithelioid sarcoma often presents innocuously and misdiagnosed as a benign lesion. [3] It is also associated with high recurrence index, metastasis and mortality. Hence, here we are reporting this unusual case to add more knowledge.

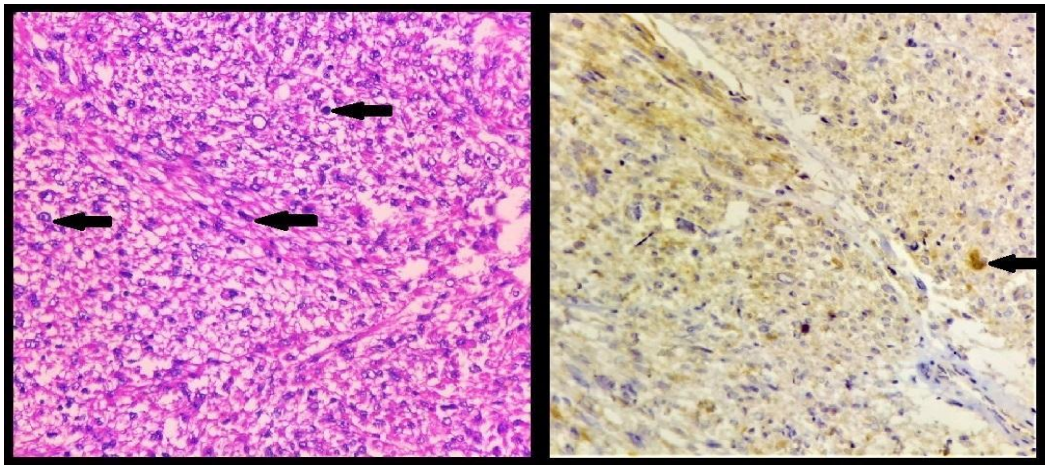
## CASE REPORT

A 65 years old female, presented to the surgery outpatient department with complaints of lump. It was associated with dull-aching pain in the abdomen since a month and a half. No other relevant history was present. On examining the patient, a palpable firm to hard, immobile, non-tender lump, roughly measuring of a size of a fist was noted in the hypogastric region of the abdomen. Routine laboratory investigations and systemic examination showed no abnormality. Computed tomography (CT) of the abdomen and pelvis showed a large well defined heterogeneously enhancing soft tissue lesion measuring 7x8x10 cm arising from the retroperitoneum, involving mid ureter. The patient was taken up for laparotomy and the tumour was excised following which it was sent for histopathological examination. Grossly, the specimen received was a single, globular, firm mass measuring 6.5x7x10 cm in size. External surface was glistening grey-white with bosselations and showed areas of haemorrhage along with congested blood vessels. Cut surface showed grey-white to grey-brown areas with foci of necrosis.



Microscopically, the sections studied showed a circumscribed lesion comprising focal nodular arrangement of tumour cells composed of polygonal to plump spindle cells and epithelioid cells with dispersed collagen bundles. Individual cells showed abundant eosinophilic cytoplasm with nuclei showing minimal pleomorphism. Also noted were focal areas of necrosis, haemorrhage along with plenty of congested blood vessels. Abundant mitotic figures were also seen. At places, infiltration by chronic inflammatory cells comprising predominantly of lymphocytes were also noted. Vascular invasion was identified. Immunohistochemistry showed positivity for Epithelial membrane antigen while, CK 8 and CK19 were negative.





## DISCUSSION

Epithelioid sarcoma is a rare mesenchymal malignancy displaying multidirectional differentiation, predominantly epithelial. [5] It is a slow-growing tumour of unknown histogenesis associated with a high incidence of local recurrence, regional lymph node involvement and metastasis. [2,3,5-7] It mainly affects adult male population and it has two principle variants, the conventional/ classic/ distal-type (far more common) and the proximal-type. [1,5,8] It typically occurs in the distal extremities. Distal-type ES is histologically characterised by tumour nodules with areas of central necrosis surrounded by large polygonal to spindle cells. It includes angiomatoid variant, fibroma-like variant and myxoid variant. [9]

In 1997, a more aggressive, proximal or large cell type arising as deep-seated soft tissue mass at proximal body sites such as pelvis, perineum, genital tract and proximal extremities was described. [4,6] Histologically it is described as a tumour with multinodular growth pattern and large polygonal to spindle cells along with epithelioid cells arranged in sheets with abundant eosinophilic cytoplasm demonstrating moderate cytologic atypia, vesicular nuclei with prominent nucleoli. [9,10] It may focally or predominantly show characteristic large cells with rhabdoid morphology. [11] Lesions of epithelioid sarcoma can also be described as superficial (also referred as "hard woody knot") or deep (areas of firmness or localized pain). [12] ES has an unfavourable prognosis with reported 77% local recurrence and 45% distant metastasis rates, usually to regional lymph nodes, lungs, skin, scalp, bone, brain and other soft tissue parts. [5,6,13] Poor prognostic factors include proximal location of the tumour mass, amount of necrosis present, vascular invasion, delay in diagnosis and inadequate excision. [6]

A specific marker has not yet been identified in ES. Immunohistochemistry (IHC) shows positivity for epithelial markers- cytokeratin (CK) and epithelial membrane antigen (EMA) and most cases co-express vimentin. The marker CD34 is expressed in 60–70% of cases. [7,9] Epithelioid sarcoma has many histopathologic mimics and hence, IHC has an important role to play. The differential diagnosis includes few benign lesions such as fibrous histiocytoma, nodular fasciitis, fibromatosis, sclerosing epithelioid fibrosarcoma, necrobiosis lipoidica, granuloma annulare, chronic granulomatous inflammation and giant cell tumour of tendon sheath while malignant lesions include amelanotic melanoma, epithelioid malignant peripheral nerve sheath cell tumour, metastatic squamous cell carcinoma, synovial sarcoma, epithelioid haemangioendothelioma, leiomyosarcoma and malignant extrarenal rhabdoid tumours of the soft tissue. [5,9] Imaging studies are not much helpful as CT or MRI findings are nonspecific. [1] The diagnostic dilemma lies in the fact that ES shows deceptively harmless appearance during the initial period of the disease and a presumed benign diagnosis is frequently pursued causing delay. [12] Careful histopathological examination along with immunohistochemical markers are crucial for diagnosing Epithelioid Sarcoma. Treatment includes wide, total surgical excision with clear margins and high-dose chemoradiotherapy and is associated with low rates of local recurrence.

## CONCLUSION

Epithelioid Sarcoma is a rare malignant soft tissue tumour with marked propensity towards local recurrence and distant metastasis. As it mimics other neoplasms specifically benign, it frequently leads to difficult and missed diagnosis. This leads to delayed and improper treatment which adversely affects the prognosis. Hence, epithelioid sarcoma should also be considered a differential among the patients presenting with a retroperitoneal mass.

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