

CASE REPORT

EPITHELIOID SARCOMA OF WRIST IN 52 YEARS OLD FEMALE IN CHENNAI, INDIA

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ABSTRACT

Epithelioid sarcoma is an uncommon kind of sarcoma that often affects young individuals and has a preference for distal upper extremity; forearm and hand. This usually begins as slow-growing, painless tumor. It can also be seen as multiple tumours. Lymphatic channels are the primary routes of dissemination for the disease's recurrence and spread. Using polygonal cells, a biopsy is the preferred technique of diagnosis. Treatment primarily consists of wide resection.

Here, we present a case of epithelioid sarcoma involving the wrist of 52-years old female. It was initially diagnosed by imprint cytology. To make an accurate diagnosis in a case with a unique history, it is necessary to consider the clinical presentation, morphologic examination, and a broad panel of immunohistochemical staining.

KEY WORDS: Sarcoma; Epithelioid Sarcoma; Wrist; Adult; Female; Middle Aged.

Cite as: Kannan AM, Muddha SS, Gali V. Epithelioid sarcoma of wrist in 52 years old female in Chennai, India [case report]. *Gomal J Med Sci* 2022 Jan-Mar; 20(1):115-6. <https://doi.org/1046903/gjms/20.01.1135>

INTRODUCTION

It is estimated that about 1% of all soft tissue sarcomas are epithelioid sarcoma; a mesenchymal soft tissue sarcoma. Enzinger¹ defined it for the first time in 1970. Young people (median age of 23 years) are usually involved. Fingers, hands, forearms and pretibial regions are most frequently affected by this disease.¹ A proximal variant of the disease has also been identified; with occurrences more commonly in the upper extremities.² Other rare sites of presentation are pelvis, vulva, penis, and spine.²

CASE REPORT

A 52-years old female presented with swelling in right forearm for five years, with no history of trauma or pain. The patient was evaluated with radiology and surgical excision was suggested. The mass was excised. (Figure 1)



Figure 1: Epithelioid sarcoma of wrist in 52 years old female in Chennai, India

The history of upper limb swelling, which was hard in consistency, provoked a suspicion of sarcoma by senior consultants. Imprint cytology was done on table to confirm the diagnosis, which showed pleomorphic cells with few areas showing large round to polygonal cells with centrally placed nuclei with eosinophilic cytoplasm, confirming sarcoma. Surgery was extended to wide local excision to have clear margins.

Histopathology showed features of nodular arrangement of cells, large round to polygonal cells with centrally placed nuclei with eosinophilic cytoplasm and presence of necrosis in center of tumor nodules. Patient was followed from 2020 till 2022 date to rule out metastasis or recurrence.

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Date Submitted: 04-01-2022

Date Revised: 03-03-2022

Date Accepted: 22-03-2022

DISCUSSION

Sarcomas are tumors of bone, fat, cartilage, muscle or connective tissues; mesenchymal elements. Epithelioid sarcoma is a rare type of sarcoma which usually involves upper limb; mostly finger, forearm and hand. The patients usually presents with history of mass from a longer duration with no pain and having firm to hard consistency. Superficial mass tends to ulcerate leading to misdiagnosis and deeper mass leads to under diagnosis.

Cells of origin: primitive mesenchymal cells with histiocytic and fibroblastic differentiation. The slow growing tendency of tumor, its recurrence and spread via lymph node makes the choice of treatment as well as follow-up mandatory. Wide local excision or amputation is done in large mass specially in fingers and toes followed by radiation/chemotherapy. The common sites of metastases are lungs and bones.

Grossly the mass appears as glistening grey white or grey tan mottled surface with yellow or brown area due to focal necrosis or haemorrhage.

Histopathological features are polygonal cells with eosinophilic cytoplasm, nodular arrangement of pleomorphic cells with central necrosis and peripheral spindling of cells.

The immunohistochemistry express vimentin, EMA, cytokeratin and CD34 positive and negative to S100 and desmin.

The prognosis of tumor depends on age group; younger age group has a better prognosis when compared to older patients. Approximately 13% of patients will have multifocal tumours, and approximately 13% of patients will have metastatic illness.

For epithelioid sarcomas, inactivation of the SMARCB1 gene, also known as loss of INI-1, is the most prevalent

genetic mutation, which may be discovered in 80-90 percent of the cases. As a result of chromosomal 22q11.2 changes or deletions, epithelioid sarcoma is distinguished from other malignancies by the occurrence of 8q gains, specifically i8 (>q10). Cell division is controlled by SMARCB1 because it is a key component of the 15-subunit SWI/SNF (or BAF) complex, which regulates the nucleosome architecture of our genome and was demonstrated to be the strong tumor suppressor gene such as signals to over-replicate. In epithelioid sarcoma, this tumor suppressor is frequently inactivated, leading in uncontrolled cell proliferation and tumor development.

Treatment of choice is surgical resection of the tumor, followed by chemotherapy in a combination of ifosfamide and doxorubicin. The use of radiation therapy is also a possibility when a tumour is declared inoperable or when broad surgical margins are not obtainable after surgery.

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CONFLICT OF INTEREST

Authors declare no conflict of interest.

GRANT SUPPORT AND FINANCIAL DISCLOSURE

None declared.

AUTHORS' CONTRIBUTION

The following authors have made substantial contributions to the manuscript as under:

Conception or Design:	AMK, SSM
Acquisition, Analysis or Interpretation of Data:	AMK, SSM, VG
Manuscript Writing & Approval:	AMK, SSM, VG

All the authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.



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