Case Report

Proximal Epithelioid Sarcoma with Chest Wall Infiltration Presenting as Recurrent Ulcerative Lesion at the Back in a Young Female

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Abstract

Epithelioid sarcoma is a rare soft tissue tumour in young adults. These are broadly classified into proximal and distal tumours; proximal tumours being less common and more aggressive than the distal tumours. The tumour is more common in males. We present a rare clinical presentation of epithelioid sarcoma with involvement of chest wall and chest cavity. Surgical excision with wide margin excision is the treatment of choice followed by chemotherapy. However, regular follow-up is advised as these tumours have high rate of recurrence.
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Key words: Epithelioid sarcoma, Proximal, Chest wall, Surgery.

Introduction

Proximal epithelioid sarcoma is more likely to be confused initially with benign lesions due to their nature of presentation as small subcutaneous growth. Thus, delaying the diagnosis at the cost of tumour extension, as in our case. Early detection, histopathological confirmation followed by appropriate surgical excision is the primary goal of treatment. However, proximal epithelioid sarcoma are notorious for recurrence and usually have poor outcomes even after surgical treatment.

Case Report

A 26-year-old female developed a small nodular lesion (2cm × 2cm) over the interscapular region one year back. There was no itching or pain associated with it. She used over the counter available local application and hot fomentation for the same. However, over a period of one year, the swelling grew larger (8cm × 10cm).

Physical examination revealed a tachypenic patient with tachycardia. Examination of the back showed localised growth with erosion of overlying skin (Figure 1). According to the patient, the growth over the back started as a small lesion but enlarged rapidly in a matter of few weeks.

Chest radiograph showed an opaque left hemithorax (Figure 2) with a mediastinal shift. Intercostal chest tube was placed and fluid drained out. The fluid was exudative in nature and cytology was positive for malignant cells. Repeat chest radiograph showed only partial clearance. Computed tomography (CT) of the chest, done to assess the extension of the tumour and to evaluate lung status

Figure 1. Photograph of clinical examination of the back of the patient showing a large ulcerative lesion.

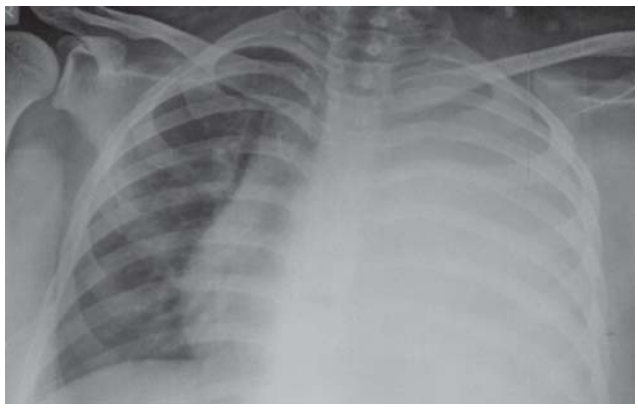

Figure 2. Chest radiograph (antero-posterior view) showing massive left-sided pleural effusion with a mediastinal shift.

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(Figures 3 and 4), demonstrated infiltration of the tumour into the chest cavity along with residual effusion. Wide excision surgery with clear margins was performed.

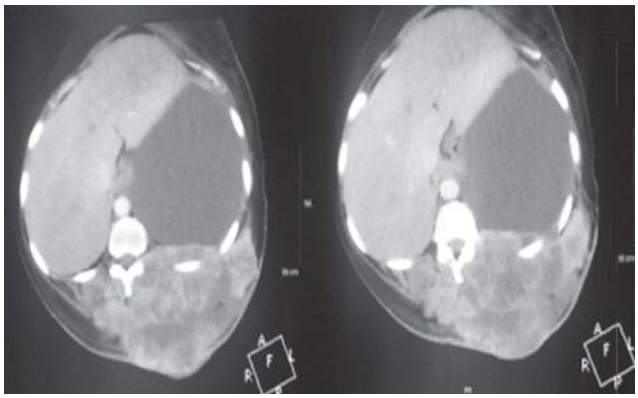

Figure 3. Computed tomography of chest showing tumour infiltrating into the posterior chest wall with significant fluid in pleural space.

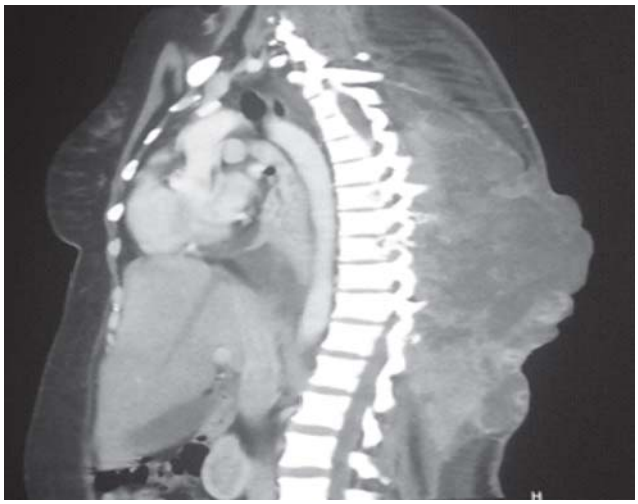

Figure 4. Computed tomography of chest (l midline saggital section) showing craniocaudal extent of the tumour.

Histopathological examination of the excised mass lesion revealed malignant cells with epitheloid to rhabdoid cells having prominent nucleoli and intra-cytoplasmic inclusions with large areas of tumour necrosis. Immunohistochemistry showed positivity for pancytokeratin, cytokeratin 7, cytokeratin 20, CEA and CDX-2. Tumour cells diffusely expressed AE1/AE3 and EMA in significant areas and focally positive for S100-P. Tumour cells were negative and controls positive for HMB45/INI-1/SMARCB 1. A diagnosis of proximal large cell type epitheloid sarcoma was made.

Patient was started on chemotherapy with adriamycin, ifosamide and holoxan. However after few cycles, she developed anaemia and thrombo-cytopaenia for which she received regular blood transfusions. Apart from low hemoglobin and platelet counts, all other routine investigations were normal. Her respiratory distress had markedly increased. Chemotherapy was planned but the patient developed respiratory distress and became unresponsive, she was intubated immediately. Her condition deteriorated rapidly thereafter and she died after a cardiac arrest.

Discussion

Epithelioid sarcoma is a rare soft tissue, slow growing tumour with high rate of recurrence and metastasis in young adults. The term was coined by Enzinger in 1970.[1] Depending on the location, the tumour can either be described as distal or proximal. Distal epitheloid sarcoma is more common variant than proximal and most frequently affects hands and forearms followed by distal lower extremities and proximal upper extremities.

The proximal variant, first described in 1997, is either subcutaneous or more commonly deep in the dermis. Various sites of its occurrence include perineum, thighs, inguinal, axilla, flanks and rarely over the back, as in our patient. The proximal variety is more aggressive with a high rate of recurrence and have a poor prognosis. No association has been noted between tumour size and metastasis.

The tumour usually presents as a firm to hard subcutaneous nodule that grows over time and may ulcerate. Pain is usually not present. Due to its rarity, mis-diagnosis with ulcerating wart or poorly healing traumatic wound is not uncommon. Approximately 13% patients present with multiple lesions and a similar percentage has metastasis as the clinical presentaion.[2] Depending upon the site, size and metastatic extent of tumour, TNM (tumour, nodes, metastasis) classification of soft tissue sarcoma can be used for staging.

Differential diagnosis of proximal type of epitheloid sarcoma includes extra-renal rhabdoid tumour, rhabdomyosarcoma, epithelioid nerve sheath tumour, melanoma and undifferentiated carcinomas. Involvement of chest wall is very rare in proximal epitheloid sarcoma. Hasegawa *et al*[3], in their study of 20 patients with epitheloid sarcoma observed chest wall involvement in only one case. Aizawa *et al*[4] described two cases with chest involvement.

Radiological investigations in the form of chest radiograph, computed tomography or magnetic resonance imaging are used to evaluate the extent of lesion and to plan surgical intervention. However, diagnosis can be confirmed on histopathology. Immunohistochemistry staining can differentiate this entity when cytokeratins, epithelial membrane antigen and vimentin are usually positive.[5] A common characteristic of epitheloid sarcoma observed in 80% of the patients is the loss of function of the SMARCB1 gene (also termed INI1).[6] The INI1 is a tumour

suppressor gene and loss of its function leads to unregulated growth.

Surgical excision of the tumour with wide negative margins is the treatment of choice. In advanced stage of the disease, inoperable and metastatic tumours, chemotherapy with ifosfamide and doxorubicin with or without radiotherapy is considered as the standard of care. However, the success rate remains low in advanced cases and recurrence is not uncommon. Proximal type epitheloid tumours carry high mortality rate. Hasegawa *et al*[3] reported that 65% of patients died during their study due to this disease.

In conclusion, proximal epitheloid tumours are rare, aggressive and difficult to treat. Chest wall involvement is extremely rare. Wide surgical excision followed by chemotherapy and/or radiotherapy is the treatment of choice. Recurrence is common with a high mortality rate.

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