Diagnostic dilemma in skin adnexa malignancy: a rare case report with review of literature

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INTRODUCTION

Apocrine malignancies are rare to occur. Axilla and the anogenital skin are the most common sites of primary tumour. Apocrine malignancy carries high risk of lymphatic and vascular spreading to lung, liver and bone. A 40 years old painter, presents with diffuse swelling and pain in the left lower limb up to left inguinal region since 5 to 6 months. With generalised lymphadenopathy. On evaluation, revealed adenocarcinoma of apocrine, skin adnexa in the extremities of the limb with extensive distant metastasis. Apart from Dermatologists other board specialty consultant should be aware of the possibility of apocrine adenocarcinoma while differentiating cutaneous tumor located in the extremities. Apocrine malignancy should be treated aggressively with wide local excision or amputation the involved part for improving the life expectancy. Since prevalence of apocrine malignancy are rare, and it is associated with increased mortality, it provoke us the field of research for discovering other modalities of treatment.

Keywords: Apocrine gland, Apocrine malignancy, Chemotherapy, Skin adnexa, Wide local excision

CASE REPORT

A 40 years old painter by occupation admitted to our hospital with c/o swelling below the left ankle joint for 2 years, progressively patient developed diffuse swelling and pain in the left lower limb up to left inguinal region, throbbing in nature, with painful movement at both ankle and knee joint since 5 to 6 months.

It is associated with loss of appetite, with significant loss of weight. Swelling is not associated with fever/chills/rigors. No h/o abdomen pain/ abdominal distension/cough/breathlessness. Patient underwent surgery for swelling below the left ankle joint 8 months back at private hospital, 2 months later patient developed swelling at the operated site. Patient is alcoholic for 20 years and left 3 months back.

On examination, patient is moderately built, poorly nourished. Patient was limping due to pain. He was anemic. generalized lymphadenopathy present, involving b/l inguinal lymph node, central group of right axillary.
right and left supra clavicular lymphnode and right sub mandibular lymphnode. Pitting edema of the left lower limb. Right sided hydrocele (Figure 1 and 2).

Figure 1: Medial aspect of the left foot with lesion situated near the heel.

A solitary, ill defined, 5 x 6 cm of size of swelling with globular surface at medial aspect of the left foot just below the ankle joint, with overlying linear scar mark of 6cm in length. Overlying skin is stretched with pitting edema and tenderness present over the swelling. Per abdomen examination s/o hepatomegaly.

Figure 2: Lateral aspect of the left foot with pedal edema.

Excision biopsy was performed histopathological examination was revealed that cutaneous adnexal tumor-tubular Apocrine adenocarcinoma. Fine needle aspiration cytology from inguinal lymphnode was done and histopathological examination revealed that malignant epithelial lesion adenocarcinoma, malignant adnexal tumour) metastasis to lymphnode. CECT thorax and upper abdomen s/o metastatic malignant lesions involving b/l lung parenchyma (Figure 3), mediastinal/ hilar lymph nodes and liver, spleen, periportal and retroperitoneal lymph nodes.

USG scrotum s/o dilated lymphatic channels within the scrotum B/L with foci of curvilinear echogenic structures with minimal right hydrocele. No e/o deep vein thrombosis venous Doppler of left lower limb. X- Ray of the diseased part of the limb does not show involvement of bone (Figure 4). Patient is been treated with palliative chemotherapy and has expired within 2 weeks of initiation of treatment.

Figure 3: X ray foot does not show bony involvement.

Figure 4: X ray chest showing metastasis canon ball appearance

DISCUSSION

There are two types of sweat glands: eccrine glands and apocrine glands. Eccrine glands do not show cytological changes during secretion and apocrine glands, characterised by decapitation secretion, in which part of the cell is pinched off and released into the lumen. Eccrine glands play a major role in thermoregulation and electrolyte balance. They are present everywhere in the human skin and are composed of a secretory portion, an intradermal duct and an upper intraepidermal part, called acrosyringium.

The acrosyringium has a unique symmetrical and helicoidal course, which length is correlated to the thickness of the epidermis. Apocrine glands are located only on genital, axillary and mammary areas, where they are always connected to a hair follicle. Their exact role in humans is unknown. A third type of intermediate sweat glands, the apoecocrine glands, was recently described in axillary areas. Sweat glands can be involved in various inflammatory processes and can lead to a large range of both benign and malignant tumors. Sweat gland malignancies are the rare. The first report was published in 1944. There are two subtypes mainly eccrine and
apocrine gland adenocarcinoma, in world literature it accounts of approximately 200 and 38 cases respectively. Apocrine tumours are much less common than eccrine glands. Apocrine adenocarcinoma also been observed to arise in association with other benign tumours such as apocrine adenoma and Apocrine hyperplasia.\(^6\,\,7\)It is often clinically diagnosed incorrectly. Mainly diagnosed on histopathological examination of resected specimen, which is again an incidental finding. These neoplasms are both complex and cumbersome. Tumour spread through lymphatic and haematogenous route, most commonly spread to the regional lymph nodes, liver and lungs. At the time of presentation, about one third of the patient presents with involvement of regional lymph node.\(^7\)

Microscopically, eccrine and apocrine both have the appearance of adenocarcinoma with PAS (Periodic acid Schiff) because of the presence of glycogen granules. Both can be differentiated on basis of diastase digestion property.\(^7\) Apocrine tumour cells are diastase resistant cells as apocrine tumour cell digest diastase and they retain PAS stain. An eccrine tumour cell is diastase sensitive and does not retain the PAS stain.\(^7\,9,10\) Treatment choice is wide local excision with adequate tumour free margins.\(^7\,8\)

The prognosis of apocrine adenocarcinoma is very poor and its prognosis all depends on size of the tumors, histological type, lymph node involvement and distant metastasis. The disease-free survival rate for 10 years in the absence of metastasis to the lymph nodes is reported to be 56%. This percentage, however, drops to 9% if lymph node metastasis is involved.\(^9,10\)

Various case reports mentioned that transient response to various chemotherapy agents. Bellman et al., described excellent response to systemic 5-fluorouracil chemotherapy by patient with widely metastatic apocrine gland. Two patients with sweat-gland carcinoma well responded to combination chemotherapy with doxorubicin, cyclophosphamide, vincristine, and bleomycin was described by Mezger et al., In one of these patients has achieved complete remission of tumour for 2 years duration; the other patient had a partial remission of tumour for 4 months duration.\(^11,12\)

CONCLUSION

Apart from Dermatologists other board specialty consultant should be aware of the possibility of apocrine adenocarcinoma while differentiating cutaneous tumor located in the extremities of the limb.

Apocrine malignancy should be treated aggressively with wide local excision or amputation the involved part for improving the life expectancy. Since prevalence of apocrine malignancy are rare, and it is associated with increased mortality, it provokes us the field of research for discovering other modalities of treatment.

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