

Malignant eccrine poroma

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ABSTRACT

Benign eccrine poroma arises from the intraepidermal portion of the eccrine gland duct. Malignant transformation is rare and should be suspected when these lesions present with pain, bleeding or itching. We report a 44-year-old male patient who presented primarily with a lesion diagnostic of benign eccrine poroma of the right foot sole with no clear evidence of malignancy, which was incompletely excised, followed 5 months later by local recurrence, ulceration, occasional bleeding and right inguinal lymphadenopathy. Incomplete excision of the primary tumor as well as excision of a skin lesion on the right knee joint revealed malignant eccrine poroma with aggressive histology, lymphovascular and perineural invasion. Investigations revealed no evidence of distant metastasis. This tumor might be malignant at the first presentation, which was not confirmed histopathologically considering the short duration of only 5 months for malignant transformation. The patient received 3 cycles of Docetaxel (Taxotere), Cisplatin combination chemotherapy with partial response. The management of metastatic malignant eccrine poroma is difficult. It has proven resistant to many chemotherapeutic agents and radiotherapy.

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Malignant eccrine poroma is a rare cutaneous neoplasm, which originates from the intraepidermal ductal portion of the eccrine gland. The first case was reported by Pinkus et al¹ in 1956, followed by more than 200 cases reported in the literature to date.² It commonly presents as verrucous plaques or polypoid growth, which occasionally bleeds with minor trauma. It usually metastasizes to regional lymph nodes. Distant metastasis to lungs or other organs are less common. Patients usually present between the fifth and eighth decades of life.³ More than 50% of malignant eccrine poromas are found in the lower limbs, 20% on the trunk, 15% on the head and 10% in upper limbs.³ De Novo malignant eccrine poroma of the nose as a location may occur rarely.⁴ This paper reports a young patient with malignant eccrine poroma at a younger age than that described in the literature for awareness of our doctors in the field of oncology, dermatology and surgery. Literature review was carried out to look for any effective

systemic chemotherapy or any other treatment modality.

Case Report. We present a case of a 44-year-old male patient with an ulcer of the right foot sole for 2 years duration, swelling, pain and difficulty in walking. Incomplete surgical excision was performed 5 months prior to presentation. Histopathology revealed dermal tumor composed of cord and broad columns of basaloid cells extending into the dermis from under the surface of the epidermis. Ducts are seen within the tumor columns. The stroma is vascular, with no clear evidence of mitosis or atypia. Final diagnosis was benign adnexal tumor consistent with eccrine poroma with no clear evidence of malignancy (**Figure 1**). He presented recently with right inguinal lymphadenopathy of 3 x 4 cms in size and recurrent enlarging right foot sole ulcer with occasional bleeding. Excision of the recurrent tumor and right

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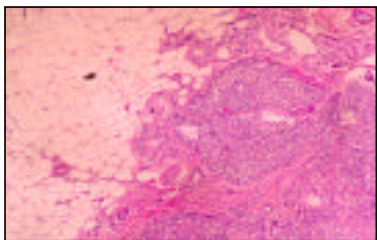


Figure 1 - Benign adnexal tumor consistent with eccrine poroma with no clear evidence of malignancy. Hematoxylin and Eosin stained X 200.

knee skin lesion revealed malignant eccrine poroma with aggressive histology. Excision was incomplete with positive resection margin as well as lymphovascular (Figure 2a) and perineural invasion (Figure 2b). Investigations revealed no evidence of distant metastasis. The patient received 3 cycles of Taxotere, Cisplatin combination chemotherapy with partial response. He was then referred for palliative radiotherapy.

DISCUSSION. Primary eccrine gland carcinomas may arise from preexisting, less mature eccrine gland tumors or may develop de novo from eccrine glands within the skin. Primary eccrine gland carcinomas include syringoid eccrine carcinoma, mucinous eccrine carcinoma, clear cell carcinoma, microcystic eccrine carcinoma, adenoid cystic carcinoma, aggressive digital papillary adenocarcinoma and eccrine adenocarcinoma. Other varieties were displayed including squamous, spindle cell differentiation and colonization by melanocytes.³ Mitosis, the presence of lymphovascular invasion, and tumor depth >7 mm were associated with a poorer prognosis. Tumors presenting an "infiltrative" advancing margin are particularly prone to local recurrence and require wide excision.³ It commonly presents as verrucous plaques or polypoid growth, which occasionally bleeds with minor trauma. It usually metastasizes to regional lymph nodes, and distant metastasis to lungs or other organs are less common.

The management of patients with metastatic malignant eccrine poroma is difficult. It has proven resistant to many chemotherapeutic agents and radiotherapy has not generally been effective.³ However, there are several cases reported that have responded to different chemotherapy regimens. A complete response was obtained with melphalan, intraarterial infusions of 5-fluorouracil (5-FU), and hyperthermia.⁶ A partial response was reported with

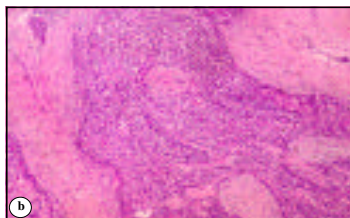
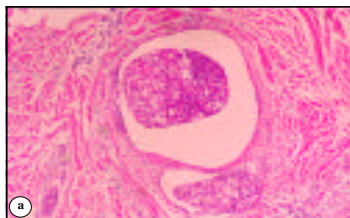


Figure 2 - Malignant eccrine poroma with aggressive histology showing (a) lymphovascular invasion identified. Hematoxylin and Eosin stained X 400, and (b) Perineural invasion identified. Hematoxylin and Eosin stained X 200.

radiotherapy combined with 5-FU. A prolonged, complete remission was achieved with doxorubicin, mitomycin C, vincristine, and 5-FU alternating with cisplatin and bleomycin.⁷ In addition, some response was seen with isotretinoin and a remission lasting several months was obtained with docetaxel.⁸ Some local benefit has resulted from peri-lesional injection with interferon-alpha and interleukin-2.³ Gutermonth et al⁹ reported a long term stable disease and minor remission with less aggressive treatment using a combination of Interferon-alpha 9 million units subcutaneous 3 times per week and Paclitaxel 100mg/m² weekly intravenous, in a 67-year-old male patient with local recurrence and regional lymph node metastasis after resection of eccrine poroma.⁹ Gonzalez-Lopez et al¹⁰ reported a patient treated with prophylactic lymphadenectomy, radiotherapy, and oral isotretinoin, subsequently substituted by tegafur with subsequently no evidence of distant metastasis after a 5.6 year follow-up.¹⁰ Our patient received 3 cycles of systemic chemotherapy using Taxotere (Docetaxel) and Cisplatin, which resulted in partial response. The patient was then referred for palliative locoregional radiotherapy.

The development of multiple cutaneous deposits with a lymphangitic pattern and microscopic epidermotropic spread is a rare pattern of metastasis in malignant eccrine poromas.¹ There are only 7 reported cases of similar modes of spread in the literature.^{8,11} The tumor cells reportedly travel along dermal lymphatics and establish themselves within the epidermis. This histological picture simulates Paget's disease. In our case, lymphatic drainage followed the typical pattern of anatomical drainage of the lower limbs. The presence of cutaneous lesion at the knee joint suggests lymphangitic pattern and epidermotropic spread.

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