

Epithelioid Sarcoma: A Frequently Misdiagnosed Neoplasm

Sir,

Epithelioid sarcoma, originally described by Enzinger in 1970, is a rare, malignant, soft-tissue neoplasm (1). The frequency with which the tumour is mistaken for a benign process is chiefly a result of its deceptively harmless appearance during the initial stage of the disease.

CASE REPORT

A 19-year-old white man had noticed a slowly growing, slightly painful nodule on the medial side of the distal parts of his forearm over the past 2 years. During this time he had been treated with several external agents under different diagnoses. At presentation in our outpatient clinic, he had a solitary, skin-coloured, hyperkeratotic, firm tumour, 1.5 cm in diameter. On clinical examination no other skin lesions were noted. Wide excision was performed.

Histopathological evaluation by conventional haematoxylin & eosin staining revealed a solitary tumour composed of epithelioid-shaped cells with only slightly pleomorphic nuclei, with a vesicular and sometimes granular chromatin pattern (Fig. 1a).

Immunohistochemical staining revealed the epithelioid and spindle-shaped tumour cells to be uniformly immunoreactive for vimentin

(Fig. 1b), pan-cytokeratin (Fig. 1c), and in a more scattered pattern for epithelial membrane antigen. Immunohistochemical staining showed no reactivity of tumour cells for CD34, CD45, CD68, factor VIII-related antigen, desmin, CEA or S100 antigen.

No evidence for multinodular or metastatic spreading was found on computed tomographic scans of the abdomen and thorax, magnetic resonance imaging of the neurocranium, or ultrasonic examination of either the soft tissue surrounding the tumour or the locoregional lymph nodes.

DISCUSSION

Epithelioid sarcoma usually occurs in young adults between 20 and 29 years of age, predominantly in males. It is rare in children and older people, but no age group is completely excluded (2). In 58% of cases epithelioid sarcoma occurs at the distal parts of the upper extremities (3). Epithelioid sarcomas are characterized as slowly growing, often multinodular tumours which may become painful and centrally ulcerated (4). Because of their infiltrating growth along nerves, vessels or tendons, multiple recurrences are a characteristic feature of epithelioid sarcomas (3, 5). Distant metastasis occurs in 30–40% of patients, most commonly to the regional lymph nodes and the lungs (5).

As a rare soft-tissue neoplasm, epithelioid sarcoma is frequently misdiagnosed, most commonly as necrobiotic granuloma, chronic inflammation, squamous cell carcinoma or metastasis from occult epithelial tumours (1).

Adequate treatment requires wide or radical local excision, which may be combined with adjuvant radiotherapy or chemotherapy (5, 6). Elective regional lymph-node dissection has been suggested because lymph-node metastasis is a fairly common occurrence (4). Especially in cases of recurrent disease surgical treatment can be combined with local radiotherapy, systemic chemotherapy or, if localized at the limbs, with hyperthermic limb perfusion with high-dose tumour necrosis factor- α and melphalan as recommended for other sarcomas, or even amputation (5–7).

The case presented here should alert physicians to include epithelioid sarcoma in the clinical differential diagnosis of harmless-looking, sometimes painful nodules, especially when they are localized on the hand or forearm of young adults.

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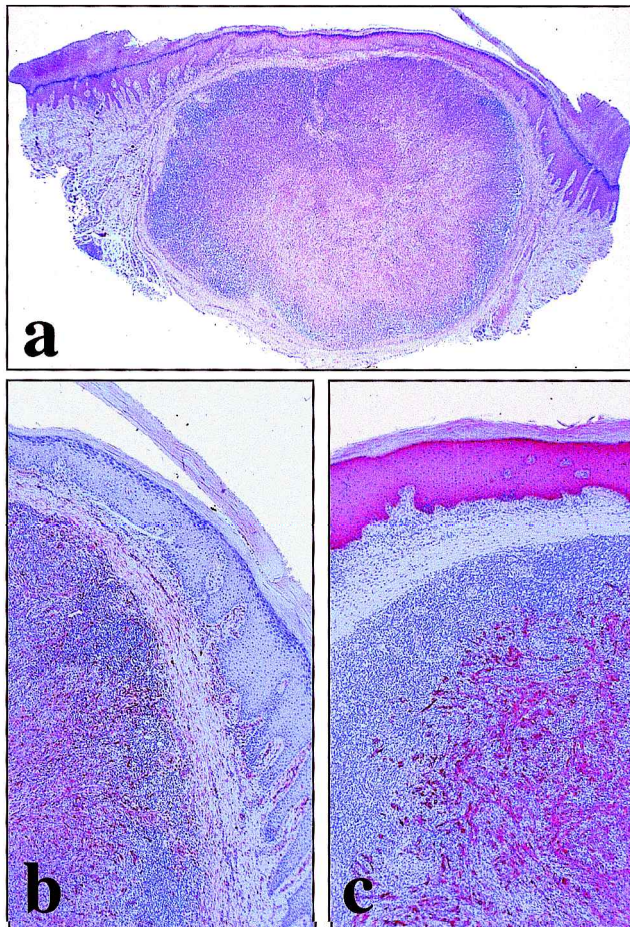


Fig. 1. Solitary, well-marked, slightly expansive and hyperkeratotic tumour located in the dermis. Note tumour cells surrounded by a dense mononuclear cell infiltrate (a). Immunohistochemical staining revealed the tumour cells to be uniformly immunoreactive for vimentin (b) and pan-cytokeratin (c).

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Infantile Acropustulosis Successfully Controlled with Topical Corticosteroids under Damp Tubular Retention Bandages

Sir,

Infantile acropustulosis (IA) is a relatively rare pustular disease, affecting children in the first few months of life. It is clinically characterized by chronic recurrent crops of sterile and pruritic papulovesicular or pustular lesions distributed mainly on the hands and feet (1, 2). The lesions usually last for up to 14 days, clear spontaneously and relapse within the next few weeks. This process resolves on its own after an evolution of a few years without residual problems. So far, it has been unaffected by any treatment except for sulfones, which demonstrate a moribistic effect. Since sulfones may have many side-effects, their use seems to be indicated only rarely in this benign disease, making the treatment of IA a formidable challenge. We report a typical case of IA, in which interval treatment with topical corticosteroids under damp conditions using wet tubular retention bandages was successful in disease control.

CASE REPORT

A 9-month-old male infant presented with a 6-month history of recurrent episodes of pruritic pustules on both soles appearing every 2–3 weeks. Severe pustulation was observed usually during a period of 5–7 days followed by an interval with only occasional eruption of single pustules. The initial clinical examination revealed several pustules, dried papulopustules and crusted lesions bilaterally on both soles, measuring about 1–3 mm in diameter (Fig. 1). Since a scabies infection was presumed clinically, although mites could never be detected, repeated antiscabietic treatment regimens using permethrin or crotamitex-containing ointments were carried out, all of which resulted in only temporary improvement.

A diagnosis of IA was made based on the clinical picture and the characteristic disease pattern, and oral glucocorticoid therapy was initiated, starting with 1 mg/kg body weight of betamethasone for 3 days. During steroid therapy a complete resolution of all skin lesions was seen. However, within 5 days of steroid withdrawal new lesions developed. Thereupon, therapy was changed to topical glucocorticosteroids (mometasone fuoroate) which were applied with the wet-wrap dressing technique according to Oranje et al. (3). The glucocorticosteroid ointment was applied to the involved extremities. The first layer comprised appropriate sizes of wet tubular retention bandages (Tubifast®; Seton Healthcare Group, Oldman, UK), followed by a second dry layer of Tubifast. All skin lesions completely resolved within 3 days with twice-daily applications of wet-wrap dressing for 3–4 h. In the following months the disease was well controlled with immediate application of these steroid dressings as soon as new pustules developed, and a treatment period of just 2 days was sufficient. Disease activity continuously diminished over the following months. During the past 6 months no disease activation has been noticed, only postinflammatory hyperpigmentation without atrophic residuals.

DISCUSSION

IA was first described in 1979 simultaneously by Kahn & Rywlin and Jarrat & Ramsdell (4, 5). It is extremely difficult



Fig. 1. Sole of a 9-month-old infant with disseminated pustular lesions indicative of infantile acropustulosis.

to treat and despite being self-limited the pruritus, in particular, can be distressing for the children. Unfortunately, apart from the moribistic effect of sulfones, other treatment options such as antihistamines, local or systemic antibiotics, local antiseptics or tar-containing ointments are of no or only minimal temporary help (1, 2, 6, 7). The effectiveness of corticosteroids remains controversial. Systemic treatment seems to have no adequate effect, while responses to topical corticosteroids are inconsistent, with results ranging from no change, through a temporary slight to a marked improvement (1, 6, 7). This considerable variability in efficacy is difficult to explain, but could be due to the different potency of the corticosteroids used. In the study by Mancini et al. (7), only high-potency topical corticosteroids were found to be helpful, whereas