"Functional" surgery in epithelioid sarcoma of the hand – case report

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Abstract

Epithelioid sarcoma is a rare malignant soft tissue tumour that affects mainly young adults and shows a high risk of local recurrence. We present a 21-year-old male with epithelioid sarcoma developing in the hand which was histologically misdiagnosed as squamous cell carcinoma. The patient was treated with a wide excision without amputation and in a 5-year follow-up period was free of local recurrence and metastasis. The functional and aesthetic results were satisfactory. Our observation suggests that wide resection with free resection margins is an alternative to more aggressive treatment.

Key words: epithelioid sarcoma, surgical treatment.

Introduction

Epithelioid sarcoma (ES) is a rare high-grade malignant soft tissue tumour. It was originally described by Enzinger in 1970 [1]. The neoplasm affects mainly young adults (10-35 years) with male predominance (approximately twice as common). The typical localization of ES is the upper limb, especially the hand (30%) [2, 3]. Treatment requires a wide resection or amputation which often results in a poor functional effect.

We present a case of a 21-year-old male with ES who was treated surgically with a good oncological, functional and aesthetic result.

Case report

A 21-year-old, right-handed, healthy male presented for 3 years with a solitary nodule in the palm at the base of the fourth finger of the right hand (Fig. 1). The tumour was painless; it slowly enlarged and ultimately ulcerated. Hypoaesthesia of the distal part of fourth finger was also observed. A biopsy of the nodule revealed an epithelioid tumour that was interpreted as squamous cell carcinoma. The lesion was excised in a block with a 5 mm wide margin under local anaesthesia. Infiltration of the digital nerve and tendon was observed. The surgical defect was covered with a full-thickness skin graft. Histopathological examination of the excised tumour revealed epithelioid and spindle-shaped tumour cells centred within the dermis. Focal necrosis of the tumour nodules, perineural and perivascular infiltration were noticed. Immunohistochemical stains revealed expression of cytokeratin, vimentin, epithelial membrane antigen, CD34 and S-100 protein (Fig. 2). The excision was incomplete. A diagnosis of ES was rendered. The patient was referred to the Department of Hand Surgery where he was operated on under general anaesthesia. A wide *en bloc* excision including digital nerves and capsule of tendon of the fourth finger was performed (Fig. 3). Digital nerves were reconstructed with sural nerve grafts. The wound was covered by a local pedicled skin flap from the third digit and a skin

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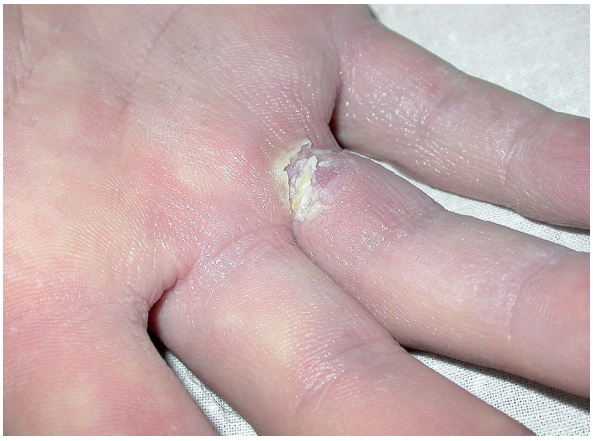

Fig. 1. Clinical features of ES, solitary nodule in the palm at the base of the fourth finger of the right hand

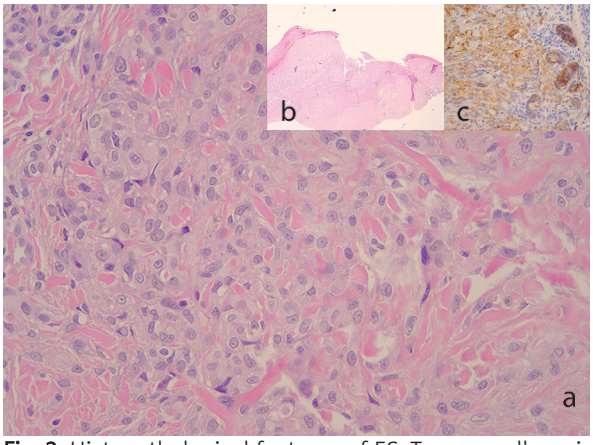

Fig. 2. Histopathological features of ES. Tumour cells, epithelioid in shape with bland nuclei, growing in irregular sheets (HE) (**a**). Tumour ulceration with the main bulk of the neoplasm growing in the mid-dermis (HE) (**b**). Expression of cytokeratin (AE1/AE3) in the tumour cells (**c**)

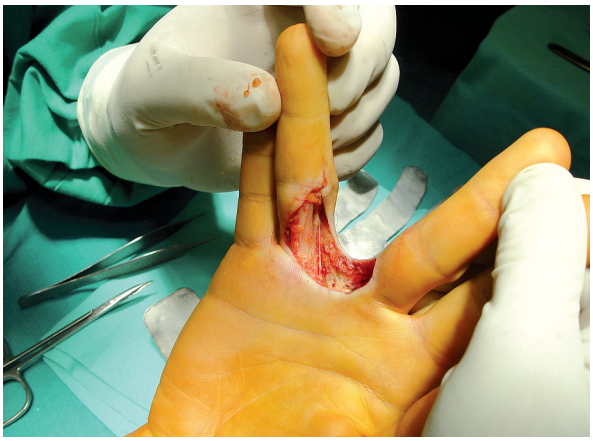

Fig. 3. Surgical defect after wide excision

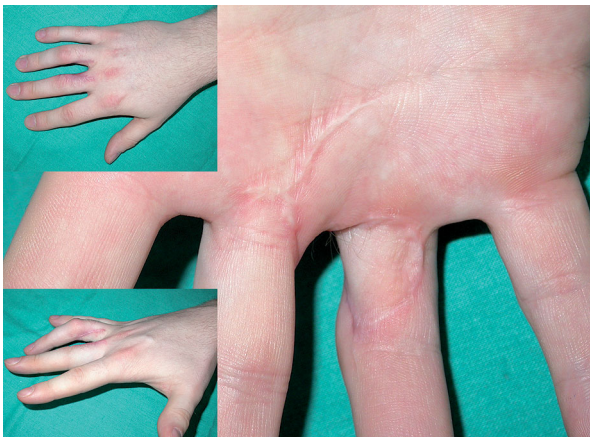

Fig. 4. Good functional and aesthetic result

graft from the wrist. The healing was not complicated. The hand function is satisfactory but hypoaesthesia of the distal part of the third and the fourth fingers and a moderate degree contracture of the fourth finger was observed (Fig. 4). Recurrence has not been established for 5 years since the original surgical treatment.

Discussion

Epithelioid sarcoma represents only 1% of soft tissue tumours but is one of the most common sarcomas involving the hand as it makes up more than 15% of malignancies occurring at that site [4]. Its clinical features depend on the location of the tumour bulk. Only a small portion of the neoplasm is confined to the skin and subcutis. Some authors believe that this is a superficial variant of ES [5]. Epithelioid sarcoma situated in the dermis are often elevated and ulcerated. A slow growing and painless nodule deeply infiltrating along the fascial planes and nerve sheaths is a typical appearance of ES [2]. Aetiology is unknown, although in around 20% of patients an antecedent trauma has been reported [3]. Due to bland morphology the tumour may be erroneously taken for a benign condition (granulomatous inflammation) or other malignant process (squamous cell carcinoma, amelanotic melanoma or synovial carcinoma of epithelioid haemangioendothelioma) [2].

Histogenesis of ES is still unclear and the most recent WHO classification included this entity among tumours of uncertain differentiation. Synovial origin has been proposed and some authors suggested derivation from primitive mesenchymal cells with fibroblastic and histiocytic differentiation [6-9].

Epithelioid sarcoma is characterized by a high risk of local recurrence (87%) and metastasis (30%). Metastatic dissemination usually involves the lungs (51%) and region-

al lymph nodes (34%) [1, 2] and the clinical course of ES is usually unpredictable. Adverse prognostic factors include male gender, advanced age, large tumour size, local recurrence, high mitotic index, haemorrhage, necrosis, vascular invasion and "proximal type" of ES. The 5-year survival rate was more favourable in females (78-90%) than in males (40-65%) [3, 10, 11]. Likewise, the superficial location also confers better prognosis in ES. Comparison of superficial vs. deep tumours revealed distant metastasis free survival rates of 87% and 56% and overall survival rates of 100% and 61%, respectively. The better prognosis in superficial tumours is probably dependant on the earlier manifestations of the tumour [12].

There is still no consensus as to the treatment of hand ES. Some authors recommend aggressive treatment with middle or distal amputation [2, 3]. Analysing 28 cases of hand ES Herr *et al.* [11] noted similar survival in patients treated with a wide excision and those that underwent forearm or hand amputation. A tendency for less aggressive surgery prevails in the contemporary reports, despite not very clear efficacy of adjuvant therapy and radiotherapy [13]. Sentinel node biopsy is advocated by some as a useful treatment and diagnostic (staging) procedure [11, 14].

In conclusion, the presented case suggests that a wide excision with a free margin may be an alternative to amputation in cases of hand ES. In our opinion treatment must be appropriate to malignancy staging but "functional" surgery should be considered in less advanced tumours, especially in superficial ES. However, long-term follow-up is required for that patient, as ES may present a protracted course and may recur even 10 years after the original surgery.

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