Epithelioid Sarcoma Presenting as Recurrent Thumb Ulcer: a Lesson to Learn

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Abstract

Epithelioid sarcoma first described by Enzinger (Cancer 26:1029–41, 1970) is a rare soft-tissue sarcoma typically presenting as a subcutaneous or deep dermal mass in distal portions of the extremities of adolescents and young adults. They are frequently mistaken for ulcers, abscesses, or infected warts that fail medical management. Patients often develop multiple local recurrences of long duration, with subsequent metastases in 30 to 50% of cases (Chase and Enzinger (Am J Surg Pathol 9:241–63, 1985)). We here report a case of left thumb epithelioid sarcoma that presented as an ulcer and subsequently metastasized to forearm, arm, axillary lymph nodes, and lungs.

Keywords Epithelioid sarcoma · Distal type · Hand sarcoma · Recurrent thumb ulcer

Introduction

Epithelioid sarcoma (ES) is a rare slow growing soft-tissue sarcoma typically involving the distal extremity of young adults, with a strong propensity for local recurrence and, ultimately, metastasis. Microscopically, most tumors are characterized by a granuloma-like pattern: nodules of spindled and epithelioid cells circumscribe areas of central hyalinization and necrosis. In 1997, Guillou et al. described a different type of epithelioid sarcoma, called proximal-type epithelioid sarcoma, which is found mostly in the pelvic and perineal regions and genital tracts of young to middle-aged adults, characterized by a proliferation of epithelioid-like cells with rhabdoid features and the absence of a granuloma-like pattern [1]. These have worse prognosis than those arising in distal locations.

Case Report

A 32-year-old male presented with complaints of swelling in his left thumb for 2-year duration. The swelling was non tender and *had progressively increased* in size to involve the distal portion of left thumb. The swelling underwent spontaneous rupture to form an ulcer 2 months prior to his hospital visit. *The ulcer persisted despite multiple debridements and in spite of administration of several antibiotics.* In November 2015, he underwent excision of left thumb.

Histopathological examination revealed a spindle cell neoplasm with epithelioid features. It also showed underlying bone and marrow infiltration by the tumor cells. Immunohistochemistry shows diffuse positivity for EMA and CD34 and focal positivity for CK and SMA. Neoplastic cells were negative for S100. Ki-67 was around 25%.

Patient had not received further treatment following amputation.

In February 2016, he presented with complains of multiple and hard mobile swellings in his left forearm and arm, with enlarged left axillary lymph nodes (Fig. 1) Excisional biopsy of one of the forearm nodule confirmed a recurrence with the same histology with positivity for same IHC markers as seen in left thumb amputated specimen (Fig. 2). CT thorax revealed multiple pulmonary metastases. Left axilla shows 4.2 × 4-cm-large lymph nodes.

Patient was started on chemotherapy consisting of doxorubicin and ifosfamide. Reassessment CT scan performed after

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Fig. 1 Clinical picture of the patient showing the partially amputated thumb and multiple cutaneous metastatic nodules over hand, forearm and arm (arrows)

three cycles of chemotherapy showed progressive disease. Second-line chemotherapy (gemcitabine and docetaxel) was started but patient did not tolerate well; hence, best supportive care was offered.

Discussion

Epithelioid sarcoma (ES) is a rare high-grade malignancy that represents the most common primary soft-tissue sarcoma of the hand. Because of its epithelial and mesenchymal differentiation, this tumor is *often mistaken* for chronic inflammatory processes, necrotizing granulomas, and various fibrohistiocytic tumors [2]. ES often presents in young men, but its significance is great because of an inherent propensity for local recurrence in addition to regional lymph node involvement and distant metastases [3]. Males are disproportionately affected with a ratio approaching 2:1. ES initially appears as a single nodule. However, at the time of diagnosis, multiple nodules representing local disease spread may be present [4], as seen in our patient. Periosteal bone invasion can occur [5].

Radical tumor excision is the primary treatment for patients with ES. Negative tumor margins must be obtained, but optimal function, particularly of the upper limb is also to be considered. Therapeutic lymph node dissection is indicated when lymph node metastases are present. A decreased risk of local recurrence has been reported with the addition of preoperative or postoperative local radiation therapy when combined with radical surgery [4]. Radiation can also be considered for patients with marginal primary resection, local regional recurrence, or palliative treatment.

Single-agent or combination chemotherapy is the treatment offered in patients with stage IV disease, as in our patient with lung metastasis. The most commonly administered chemotherapy regimens are single-agent anthracycline therapy or the combination of an anthracycline with ifosfamide [6]. A single group reported activity of a regimen combining gemcitabine with docetaxel, but the experience is limited to a small number of patients [7].

Despite the administration of palliative chemotherapy, patients with metastasis have a poor prognosis. The reported median survival is around 52 weeks and the 1- and 5-year survival rates are 46 and 0%, respectively [6]. Therefore, a

Fig. 2 **a** Tumor involving the reticular dermis and subcutaneous tissue (H&E $\times 10$). **b** Tumor cells are predominantly epithelioid and spindle shaped (H&E $\times 40$). **c** Immunohistochemistry, tumor cells are cytokeratin (CK) positive. **d** Tumor cells are strongly positive for Epithelial membrane antigen (EMA)

substantial unmet need exists to improve the medical management of ES patients by establishing novel systemic regimens and exploring novel targeted therapy.

Male gender, depth of invasion, and vascular invasion are associated with poor prognosis [8]. Anatomic location of ES appears to play a role in prognostication. The overall survival and metastases-free survival are worse in lesions proximal to the elbow or knee. Close, long-term follow-up is necessary because recurrence and metastases may occur long after definitive treatment.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no competing interests.

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