Erythematous Painful Nodules on the Scalp and Ulcerartive Nodules on the Back of a 49-year-old Woman

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CASE REPORT

A 49-year-old woman had noticed multiple asymptomatic nodular lesions growing slowly on the distal part of her right forearm for about 18 months. She went to a medical cancer center 6 months ago, a biopsy was taken from one of these nodules and the diagnosis of epithelioid sarcoma was made. The patient had undergone aggressive radical excision as amputation of the right upper arm and post-operative adjuvant radiation. Unfortunately, lung metastasis was noted during her regular follow-up.

Four months ago, she suffered from a few painful, easy-bleeding, erythematous, and firm nodules on the scalp while washing her hair. In addition, some firm ulcerative nodules on the back were also noted. The lesions on the back slowly enlarged and intermittently drained clear to turbid fluid. She came to our clinic for evaluation and management.

Cutaneous examination revealed multiple erythematous, indurated, and non-movable nodular lesions with ill-defined border on the scalp, along with some erythematous indurated nodulo-ulcerative lesions on the back (Fig. 1).

Two biopsies were taken from her lesions located on the scalp and the back respectively. Histopathology and immunohistochemical stains were shown as follows (Fig. 2, 3).

![Fig. 1](image1)
(A) An erythematous, indurated, non-movable nodular lesion on the scalp
(B) Erythematous indurated nodulo-ulcerative lesions on the back

![Fig. 3](image2)
The immunohistochemical stains for tumor cells showed positive for (A) cytokeratin (B) vimentin (original magnification x200)

(A) Tumor cells infiltrate in the middle to lower dermis (H&E, original magnification x40)
(B) Granulomatous histocytes infiltration with transition from epitheloid to spindle cells and interdigitating collagen bundles (H&E, original magnification x100)
(C) Ovoid, polygonal epitheloid tumor cells with varied amount of eosinophilic cytoplasm, pleomorphic hyperchromatic nuclei, and few mitotic figures merging with plump spindle tumor cells. (H&E, original magnification x400)
DIAGNOSIS: Cutaneous Metastasis of Epithelioid Sarcoma

DISCUSSION

Epithelioid sarcoma is a rare mesenchymal neoplasm first described in 1970, and typically presents as a painless, slow growing, firm to hard, 3–6 cm nodule localized in the deep and superficial fascia, tendon sheaths, subcutis, or rarely in the dermis. The primary site of involvement is on the distal upper extremities of adults in their third decade, usually hands and fingers, with a 2 : 1 male predominance.

Epithelioid sarcoma has high rates of recurrence (75%), involvement of regional lymph nodes (47%), and distant metastasis (51% to the lungs, 34% to the lymph nodes, and 22% to the scalp). The possible route of metastasis in our case may be vascular invasion resulting in spreading to the lung and the skin.

Histopathology of epithelioid sarcoma is composed of irregular nodules of ovoid, polygonal epithelioid cells with varied amount of eosinophilic cytoplasm, and pleomorphic nuclei, merging with spindle cells. Mitoses are present in varied frequency; vascular invasion is a common feature. Besides, ulceration usually follows epidermal involvement. At low power, a characteristic finding in approximately 40 to 50 percent of cases is tumor cells arranged in a palisade around areas of necrosis, a pseudogranulomatous appearance; hence the histological differential diagnosis of epithelioid sarcoma includes granulomatous processes such as granuloma annulare, necrobiosis lipodica, rheumatoid nodule, and granulomatous infection. In immunohistochemical study, epithelioid sarcoma is immunoreactive for vimentin, cytokeratin and epithelial membrane antigen. Up to 60% of cases are positive for CD34.

The treatment of choice for the primary epithelioid sarcoma is radical excision with radiotherapy. Chemotherapy is limited to metastatic disease.

In our case, histopathological examination of both specimens revealed typical features of epithelioid sarcoma showing tumor cells infiltrate in the middle to lower dermis. The bulk of tumors was comprised of plump, spindle-shaped cells and oval, polygonal epithelioid cells with eosinophilic cytoplasm, prominent pleomorphic nuclei and few mitotic Figs. However, the characteristic pseudogranulomatous appearance has not been found. In immunohistochemical staining, both specimens showed intensive positive for cytokeratin, vimentin, epithelial membrane antigen and CD34; negative for S-100, HMB-45, and smooth muscle actin. It is very useful in the diagnosis of cutaneous metastasis of epithelioid sarcoma by the help of immunohistochemical analysis, especially in the absence of the characteristic histopathologic findings.

REFERENCES