Case Report

Primary epithelioid sarcoma of scalp

ABSTRACT

Epithelioid sarcoma (ES) is a rare mesenchymal tumor of unknown histogenesis which displays multidirectional differentiation, predominantly epithelial. They have no normal cellular counterpart and differ from both synovial sarcoma and other carcinomas. It mainly affects young adults. It has two variants, classic type and proximal type. The more common classic type presents as a slowly growing painless nodule or plaque on the distal extremities. It is rare in children and older individuals. There is male predominance. The size varies from few millimeters to several centimeters. Central deeply seated lesions in pelvis and genital tract are termed as proximal ES. It has a multinodular growth pattern and usually occurs in older patients. These are comparatively more aggressive and metastasize early. On histopathological examination, these lesions need to be distinguished from other tumors showing epithelioid morphology. Primary ES of scalp is an exceedingly rare tumor. We present a case of nodular tumor on the scalp with cervical lymph node metastasis.

Keywords: Epithelioid sarcoma, classic type, proximal variant, scalp

INTRODUCTION

Epithelioid sarcoma (ES) is a soft-tissue tumor, composed of large, and polygonal cells resembling carcinoma. It was first characterized as a distinct clinicopathological entity by Enzinger in 1970. It is relatively rare, accounting for <1% of all soft-tissue sarcomas. The histogenesis is unknown. Usually, it is slow-growing for years, with a seemingly benign morphologic appearance. Therefore, often misdiagnosed on first encounter.^[1] It shows reactivity for both epithelial and mesenchymal markers, such as cytokeratin (CK), epithelial membrane antigen (EMA), and vimentin.^[2,3] Unlike most soft tissue sarcomas, it characteristically spreads via lymphatics to regional lymph nodes. Furthermore, metastasis to skin, deep soft tissue, fascia, bone, lung, and central nervous system are common.^[3-5]

We have come across a case of primary epithelioid sarcoma of scalp.

CASE REPORT

An 18-year-old male presented with a firm, nontender swelling in the occipital region. Fine-needle aspiration cytology done

outside was reported as positive for malignant cells. We received a specimen of wide local excision of occipital lesion with posterolateral neck dissection. On gross examination, it was a skin covered elliptical soft tissue mass measuring 6.5 cm × 3.5 cm × 2.6 cm. The skin was unremarkable. On sectioning, a partially circumscribed, gray-white tumor of size 2.7 cm × 2 cm × 1.5 cm was noted in the deep dermis and subcutaneous tissue with areas of hemorrhage and necrosis. Totally, 26 lymph nodes were dissected out.

The histopathological examination showed a partially circumscribed tumor exhibiting nodular pattern of growth with areas of central necrosis [Figure 1a and b]. The tumor cells had epithelioid morphology with vesicular nuclei, prominent nucleoli, and moderate to abundant densely

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Figure 1: The photomicrograph showing; (a) Tumor in deep dermis. (b) Tumor cells showing epithelioid morphology. (c) Perineural invasion (H and E, $\times 40$ and 400). (d) Pan-CK diffuse positivity. (e) S-100 focal positive. (f) INI 1-loss in tumor cells (IHC, $\times 100$ and 400)

eosinophilic cytoplasm. Perineural and vascular invasion was present. Four out of 26 lymph nodes [Figure 1c] showed metastatic deposits.

The differentials considered on light microscopy were the tumors with epithelioid morphology such as epithelioid sarcoma, epithelioid angiosarcoma, malignant melanoma, malignant peripheral nerve sheath tumor, and myoepithelial carcinoma.

On immunohistochemistry (IHC), the tumor cells were diffusely positive for Pan-CK [Figure 1d] and EMA with INI1/SMARCB1 loss [Figure 1f], S-100 was focally positive [Figure 1e]. CD 34, HMB 45, and p63 were negative.

Thus, the final diagnosis of Epithelioid Sarcoma was offered.

DISCUSSION

Epithelioid sarcoma is a rare, mesenchymal malignancy. It has two principle types; classic type (far more common) and proximal type. The proximal variant (with larger cells, prominent nucleoli, and rhabdoid changes) is clinically more aggressive.^[6] The classic type is usually superficially located and sometimes rendered in reticular dermis. Others are found in subcutis or deeper soft tissue. Histologically, ES is characterized by irregular nodules composed of a proliferation of relatively uniform, polygonal or epithelioid cells, often with loss of cohesion, which merge peripherally into spindle cells embedded in desmoplastic fibrous stroma without a clear demarcation. The cells usually have relatively abundant, deeply eosinophilic cytoplasm, and minimal pleomorphic nuclei [Figure 1b] with occasional mitoses.^[7]

Epithelioid sarcoma has many histopathologic mimics including benign and malignant conditions such as granuloma annulare, necrobiosis lipoidica, chronic granulomatous inflammation (especially rheumatoid nodules), fibrous histiocytoma, amelanotic melanoma, epithelioid malignant peripheral nerve sheath cell tumor, metastatic squamous cell carcinoma, synovial sarcoma, epithelioid hemangioendothelioma, leiomyosarcoma, and malignant extrarenal rhabdoid tumors of the soft tissue.^[8]

Positivity for Pan-CK, EMA along with the loss of expression of INI 1/SMARCB 1 is diagnostic.^[9,10] Although CD 34 positivity is documented in 70% of cases, in our case, it was negative. S-100 is usually negative; however, in our case, it was focally positive.

ES is an aggressive neoplasm, and local recurrence is the rule. It recurs persistently, often with successive lesions appearing more proximally, and eventually metastasizes.^[1-3,6] Wide, total surgical excision with clear margins (amputation or wide *en bloc* excision) and high-dose chemoradiotherapy represent optimum treatment and achieve low rates of local recurrence. However, even this regimen is associated with a recurrence rate of between 34% and 77% (depending on the adequacy of initial excision). Metastases in about 40% of the patients primarily involve the lungs, regional nodes, scalp, bone, and brain. In our case, lymph node metastasis was present. Median overall survival of about 88 months for patients without distant metastases and just 8 months for those with distant metastases has been documented.^[1-3,6]

Tocco *et al.* have reported a case of primary epithelioid sarcoma of the scalp complicated by hypocalcemia of

malignancy.^[11] According to them, the osteolytic process of bone invasion and several factors generated by disseminated tumor (such as parathyroid hormone-related peptide) can result in an increase in serum calcium concentration which is known as humoral hypercalcemia of malignancy.

Factors associated with poor prognosis are size >5 cm, deep subcutaneous location, focal necrosis, hemorrhage, vascular invasion, and proximal or axial tumor site.^[6,9]

Tocco *et al.* have reported a case of primary epithelioid sarcoma of the scalp complicated by hypercalcemia of malignancy.^[11] According to them, the osteolytic process of bone invasion and several factors generated by disseminated tumor (such as parathyroid hormone-related peptide) can result in an increase in serum calcium concentration which is known as humoral hypercalcemia of malignancy.

CONCLUSION

Epithelioid sarcoma is an aggressive neoplasm with a propensity for local recurrence and metastasis. Careful light microscopic analysis and judicious use of IHC panel will aid in diagnosis. Wide local excision followed by chemotherapy and radiotherapy is the treatment advocated.^[2,11] One needs to differentiate it from other mimickers as the treatment protocol differs.

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Conflicts of interest

There are no conflicts of interest.

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