Case Report

Recurrent Distal Epithelioid Sarcoma with Post-auricular Metastatic Nodule in a Young Adult Female

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Abstract

Epithelioid sarcoma (ES) is a rare slow-growing aggressive tumor of uncertain histogenesis that preferentially affects young adult males with the mean age of 27 years at presentation. A 19-year-old female represented to our hospital with 3 months history of slow-growing left posterior auricular nodule. She first presented 3 years earlier with 8 years history of a circumscribed regrowing painless right-hand mass located between the middle and ring fingers which had previously been excised twice at a private hospital without the histopathological diagnosis. The mass at presentation measured 8 cm × 8 cm in diameter and was covered by an intact skin with a healed surgical scar. A tru-cut biopsy was diagnosed ES-based on the cytomorphology of malignant epithelioid cells arrayed in nests, vague alveolar pattern with foci of pseudoangiomatous growth. Tumor was CD34 + focally. She had four courses of chemotherapy and tumor *en bloc* excision with the excision of the 3rd and 4th hand-digit with hand reconstruction. Tissue microscopy showed ES with the resection margins involvement and was CD34+, Vimentin +, epithelial membrane antigen +, S100−, and Desmin −. She did well for a year until the development of the posterior auricular nodule which was morphologically similar to the hand mass. Chest X-ray and hand radiograph revealed no lesions. ES is a distinct clinicopathologic tumor fraught with recurrences as seen in this female whose age of occurrence appears to be 9 years anecdotally. ES may be misdiagnosed due to nonspecific clinical symptoms, radiological, and pathological findings. Distant metastasis often involves lymph nodes, lungs, scalp, and bone, though the posterior auricular location was seen in this case. Large tumor size, recurrences, inadequate excision, and cutaneous metastasis are bad prognostic factors despite the young age and female sex of our patient.

Keywords: Adolescent, epithelioid sarcoma, female, postauricular cutaneous metastasis

Introduction

Epithelioid sarcoma (ES) is a rare slow-growing aggressive tumor of uncertain histogenesis with a poor prognosis outcome.[1,2] It accounts for <1% of all soft-tissue sarcomas and is more common in males.[1] The mean age at presentation is 27 years with a range of 10–39 years.[3] There are two clinico-morphological types and the more common distal (classic) type which was identified by Enzinger[4] in 1970 has predilection for the extremities, in particular, the hand, forearm, and feet in young adult males though any body part may be affected. This type is, however, rare in children and older individuals. The uncommon proximal (axial) type described by Guillou[5] *et al*. in 1997 is a more aggressive tumor fraught with early recurrences and often located in the trunk, pelvis, groin, head, and neck. In general, ES is associated with local recurrences, and metastasis to lymph nodes, lung, bone, brain, and scalp have been reported in the literature, while a young age at presentation and female gender confers favorable prognosis.[6,7] We present a young female with distal type EP of the left hand and a posterior auricular metastatic nodule.

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Case Report

A 19-year-old female represented to our hospital with 3 months history of slow-growing painless left posterior auricular nodule. The nodule was fixed, attached to the lower earlobe and measured 3 cm × 4 cm in diameter. The nodule was clinically considered as a lymph node, and there were no other palpable

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lymph nodes or swellings on physical examination. She first presented to our hospital 3 years earlier with 8 years history of a circumscribed regrowing sometimes painful right-hand dorsal mass located between the middle and ring fingers which had previously been excised twice at a private hospital without histological diagnosis and also treated with traditional herbal medication without improvement. The first excision was 2 years after the onset of the hand growth which started as a seedling and rapidly increased to the size of her thumb. This first excision was followed by rapid regrowth of the mass. The mass was re-excised for the second time 9 months later. She had no other complaints, no history of trauma, or similar swellings and had no constitutional symptoms or significant contributory medical or family history. Clinical examination at presentation was of a well-looking, calm young female with a right firm multilobulated circumscribed dorsal hand mass attached to the underlying tissue and measuring 8 cm × 8 cm in diameter [Figure 1]. The overlying skin was intact with a healed surgical scar. A clinical diagnosis of fibroma was made. The left hand was unremarkable and normal. Radiological X-ray of the right hand revealed a soft-tissue mass with no bony involvement, and a tru-cut biopsy was sent for the histopathological diagnosis. Microscopy of the lesion revealed a tumor composed of epithelioid cells with moderate cytoplasm arranged in nests, diffuse sheets and pseudoangiomatous pattern in a minimal fibrocollagenous stroma [Figure 2]. An occasional mitotic figure was seen. There was no vascular invasion by the infiltrating tumor. Tumor cells demonstrated foci of epithelial membrane antigen (EMA), vimentin, and CD34 positivity. It was diagnosed ES. She was counseled about the tumor and treatment options. She subsequently had four courses of chemotherapy (vincristine, adriamycin, MESNA, and ifosfamide) and had *en bloc* tumor excision with amputation of the 3rd and 4th right-hand-digits and hand reconstruction [Figure 3]. The specimen was fixed in 10% formalin and sent to the pathology laboratory. It measured 9 cm × 8 cm × 8 cm and weighed 172 g. Grossly, three firm nodular gray-white nodules averaging 3 cm × 5 cm were situated between the 3rd and ring fingers. Tissue histological sections were stained with hematoxylin and eosin and immunohistochemical antibodies. Microscopy showed ES with the resection margins involvement, and tumor was CD34+, EMA +, Vimentin +, Desmin −, and S100−. She could not afford further chemotherapy due to financial constraints throughout a 1-year follow-up in the outpatient clinic.

The posterior auricular nodule [Figure 4] started insidiously 15 months postreconstruction surgery. Fine-needle aspiration of the nodule was reported as suspicious for malignancy and tru-cut biopsy showed metastatic ES. There was no lymphoid tissue in the posterior auricular tru-cut biopsy. Chest and right-hand radiological examinations and abdominal and pelvic scan were all unremarkable. She is been encouraged to source for funds to recommence chemotherapy.

Discussion

ES is an aggressive mesenchymal tumor of young adult males that is often misdiagnosed and fraught with recurrences,[1] though this index patient is female. The definitive tissue of origin is uncertain because the tumor may develop in the skeletal and connective tissues and can affect any body part.[1] The uncertainty surrounding the tissue of origin and the absence of pathognomonic clinicopathologic features further compound the frequency of misdiagnosis with other neoplastic mesenchymal tumors such as fibroma, fibrosarcoma, angiosarcoma, rhabdomyosarcoma, leiomyosarcoma, and melanoma, and even inflammatory lesions such as necrobiotic and infectious granulomas due to the presence of lymphocytic infiltrates in ES histologically.[1,8] This patient was diagnosed with fibroma on clinical examination only.

ES affects any age. However, the peak age of presentation is in young adults in the age range of 10–39 years with a mean age of 27 years, though a median age of 40 years and range of 13–80 years have been documented for the proximal type ES.[3,9] Distal ES is uncommon in children, however, anecdotally, our patient's age of occurrence appears to be 9 years, although initial lesions had no definitive histopathological diagnosis. Al-Salam and Al Ashari also

Figure 1: Dorsal hand mass

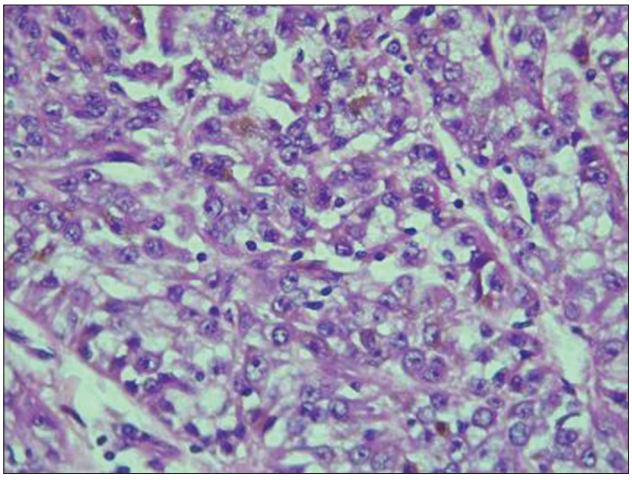

Figure 2: Sheets of epithelioid cells (H and E, ×40)

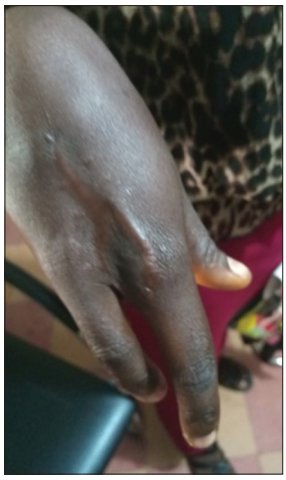

Figure 3: Postsurgical reconstruction of the right hand

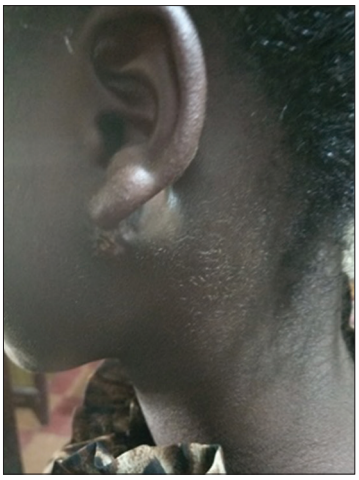

Figure 4: Posterior auricular nodule

reported a 9-year-old female with submandibular ES having distal type histological features in a proximal site.[10] There is no racial or geographic predisposition, though upper extremity sarcomas are more common in Caucasians, and the female gender confers a better prognostic outcome for this aggressive tumor.[6] Furthermore, distal type ES has better prognostic outcome compared with the proximal variant. However, tumor size >5 cm, inadequate excision, recurrence, and metastasis as in this case are poor prognostic factors.[6,7] The two early excisions were probably incomplete and were fraught with rapid regrowth, and progression rather than a recurrence, while the surgical *en bloc* tumor excision in our hospital had tumor in the resection margins. Although the tumor in this case did not exceed the initial local site on the dorsum of the hand, the tumor attained progressive larger sizes. ES has propensity for lymphatic spread and early metastasis to bone, lung, and scalp.[6,7] The metastatic focus, in this case, was a posterior auricular nodule which had no lymphoid tissue, thus eliminating a likely nodal metastasis.

ES is a distinct clinicohistopathological entity characterized by the epithelioid and spindle cells having moderate-to-abundant eosinophilic cytoplasm dispersed in the sheets and pseudo angiomatous patterns; hence, its mimicry of vascular lesions, fibro-myoepithelial tumors, rhabdomyosarcoma, and even melanoma. The stroma in ES is minimal and may show areas of lymphocytic infiltrates, hemorrhage and necrosis. Of recent, fibroma-like, fibrous histiocytoma-like, angiomatoid, and angiosarcoma-like variants have been documented.[1,8] Definitive diagnosis is based on tissue biopsy which should be supported with ancillary use of immunohistochemical antibodies such as CD34, EMA, desmin, vimentin, and S100 in particular.[1,8] Approximately, 50% of cases of ES show CD34 and EMA positivity as demonstrated in this case. Desmin and S100 negativity eliminates rhabdomyosarcoma and melanoma as possible differential diagnoses, while CD31, Factor VIII, and FL-1 are useful in vascular tumors. There is no documented specific radiological diagnostic finding, though tumor extent and metastatic foci can be demonstrated.

The best treatment outcome is achieved by surgical excision with wide-tumor free margins. Adjuvant chemotherapy has been advocated in the treatment of ES, though there is no significant difference in survival with patients who did not receive chemotherapy.[11] Our patient had surgery and adjuvant chemotherapy, though the surgical excision margin showed residual tumor and she also defaulted with chemotherapy regimen due to financial constraints, she is alive 10 years following initial presentation at a private clinic.

Conclusion

Definitive diagnosis of ES is based on clinical-pathologic correlation with supportive immunohistochemical analysis as seen in this case. Large tumor size, multilobulation, tumor regrowth, recurrences, inadequate excision, and cutaneous metastasis are bad prognostic factors despite the young age and female sex of our patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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