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Epithelioid Angiosarcoma of Perineum with Lung and Bone Metastasis; A Case Report and Review of Literature

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Abstract
Epithelioid angiosarcoma (EA) is an extremely rare subtype of angiosarcoma, which is characterized by large cells with an epithelioid morphology. Epithelioid angiosarcoma is a rare malignancy of lymphatic or vascular endothelium[1]. That can arises from any part of body like vulva, peritoneum, ovary, uterus, spine, or bones, cutaneous etc. A 47 yrs old man presenting with complain of right inguinal swelling since one year, swelling was progressively increases in size. CEMRI(6/10/2018) Whole abdomen showed Evidence of multiloculated solid focal lesion of size 3.5x3.2 cm at perineum with right predominance with 2.0x2.0 nodular discrete lesion involving upper most right thigh. Initially biopsy was done on 23/1/2018, biopsy was suggestive of round cell soft tissue tumour probably Alveolar rhabdomyosarcoma showed vimentin, CD34, pan-CK, fli-1 positive. IHC confirmed diagnosis of epithelioid angiosarcoma of perineal regions. Contrast enhance computed tomography thorax was suggestive of lung and bony metastasis. This case highlights the difficulty of diagnosing EA, which requires careful pathological examination and immunophenotype labelling. At present, CD31 and CD34 are the most sensitive marker for detecting EA
Keywords- *Epithelioid angiosarcoma,immunohistochemistry, perineum, endothelium.*

Introduction

Angiosarcoma is a rare malignancy of lymphatic or vascular endothelium that can arise anywhere in the body[1]. Angiosarcomas make up approximately 2% of soft tissue and 5.4% of cutaneous sarcomas. Chronic lymphedema and radiation treatment are each known risk factors for development of angiosarcoma, which has been well described primarily in patients treated for breast cancer[4].

Lymphangiosarcoma occurring secondary to chronic lymphedema in patients who had undergone mastectomy was first reported in 1948 and is now known as Stewart-Treves syndrome[4]. Epithelioid angiosarcoma of perineal region is rare one. We present a case of perineal epithelioid angiosarcoma with lung and bone metastasis.

Case Report

A 47 years old man presenting with right inguinal swelling since one year, swelling was initially small in size and painless. Swelling progressively increases and became painful. Contrast enhance MRI whole abdomen showed Evidence of multiloculated solid focal lesion of size 3.5x3.2 cm at perineum with right predominance with 2.0x2.0 nodular discrete lesion involving upper most right thigh Biopsy was suggestive of malignant round cell tumour with possibility of alveolar rhabdomyosarcoma, IHC was advised for further confirmation of diagnosis. True cut biopsy and IHC vimentin positive, CD34 positive, pan-CK positive, FLI-1 positive CK7negative, P-40negative, S-100 negative, Melan-A negative, CD30 negative, CD138 negative, confirmed diagnosis as Epithelioid

angiosarcoma. Contrast enhance computed tomography of thorax 06/03/2019 suggestive of few soft tissue attenuation nodules are noted in posterior segment of right upper lobe, anterior, medial basal segment of right lower lobe, superior segment of left lower lobe and posterior basal segments of left lower lobe. Enlarged necrotic lymph node/deposits in azygo-esophaseal recess measuring 16x12mm.21x17mm lytic soft tissue intensity lesion involving 7th posterior costochondral joint on left side-? Metastatic Contrast enhance computed tomography whole abdomen (07/03/2019) showed,in pancreas few hypoenhancing lesions are noted ,two in uncinated process and one in distal body region ,measuring 7-8 mm Kidney-few small hypoenhancing lesions are scattered In bilateral(left>right) renal cortex measuring size up to25mm. Heterogenous left external iliac lymphadenopathy .Few small lytic lesions involving iliac aspect of both sacroiliac joints. Patient received two cycle of methotrexate and vinblastine weekly on 1st and 8th may 2019 at outside of our hospital.In view of right inguinal fungative mass patient currently undergoing palliative radiation.

Discussion

Angiosarcomas are rare malignant tumors derived from endothelial cells that line the blood vessels. They constitute only 1%–2% of all soft tissue sarcomas while sarcomas themselves account for <1% of all malignancies.[3] They can be either cutaneous or visceral involving the breast, spleen, and liver.[3]

Predisposing factors for angiosarcoma include trauma, chronic lymphedema, irradiation, and age. Due to its wide range of plausible etiologies, it has been categorized into lymphedema-associated, radiation-induced, primary breast angiosarcoma, sporadic cutaneous angiosarcoma (age related), commonly located on the head and neck region,[13] and angiosarcoma of the soft tissue.[17] Although chronic lymphedema is the most widely recognized predisposing factor in angiosarcoma of the skin and soft tissue, other common predisposing factors may be trauma, irradiation, and age *per se*.[17]

Angiosarcoma has a tendency for aggressive local as well as distant metastasis through lymphatic and hematogenous routes. The rate of regional nodal involvement is said to be 20%–30%, which is higher than most other sarcomas. The most common site for metastasis is lungs. Thin-walled, cystic pulmonary lesions and pneumothorax are characteristic findings.[2],[3]

Pathologic diagnosis of angiosarcoma often requires thorough staining, particularly in poorly differentiated tumors. The subtypical epithelioid angiosarcomas are generally positive for CD31 and variably positive for CD34.[6] In contrast, a carcinoma would be positive for CK, but negative for CD31 and CD34.[6] In our case, the neoplastic cells were positive for CK, CD31, and CD34

Due to the rarity of angiosarcoma, information on prognosis is limited. Epithelioid angiosarcomas tend to metastasize early both to solid organs (especially lungs, bone, soft tissue, skin) and lymph nodes{5}. The overall survival rate for angiosarcomas at 5 years is approximately 35%. The epithelioid subtype is rare enough that no data

is available to compare survival to other subtypes. High proliferative index (MIB-1 at least 10%), older age, and larger tumor size are poor prognostic factors{5}. MRI or CT (for visceral and retroperitoneal tumors) can be used to characterize the primary tumor[12]. Chest CT or CXR is indicated for all soft tissue sarcomas, and imaging of the brain is indicated for angiosarcomas. Evidence-based treatments of angiosarcomas are scarce.[8] According to Seo et al,[8] radical resection is considered for radiation-induced angiosarcomas, and systemic chemotherapy is for inoperable, locally advanced or metastatic angiosarcomas.

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