Case 24

A 69 year-old Thai male from Bangkok

Chief complaint: Indurated plaque on scalp for 1 year

Present illness:

1 year previously, the patient gradually developed asymptomatic red to violaceous plaque on scalp. There was no bleeding or ulcer. There was no history of radiation at this site. He denied fever or body weight loss.

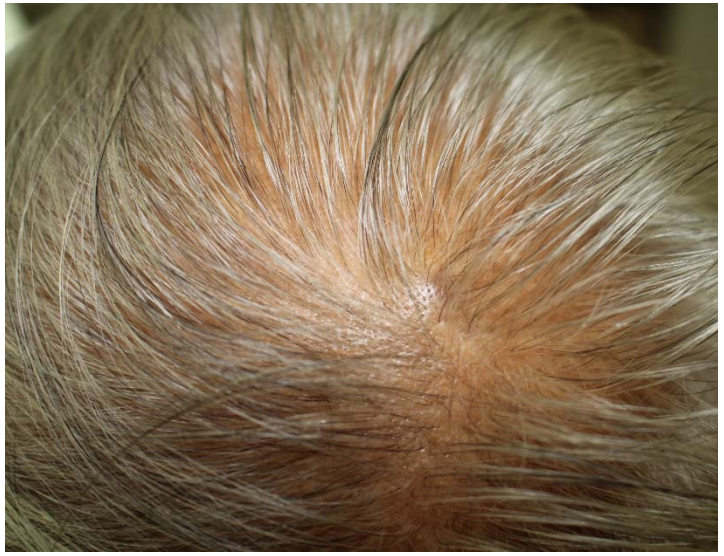
Past history:

- Underlying disease: DM type2, hypertension, dyslipidemia, left renal calculies.
  Current medications: ASA(81) 1x1 pc, HCTZ(25) 1x1 pc, sitagliptin(100) 1x1 pc, metformin(850) 1x2 pc, atorvastatin(40) 1x1 hs, $CaCO_3$(600) 1x1 pc
- Previous history of angiosarcoma on left cheek status post wide excision with local flap 10 years ago and regular follow up.

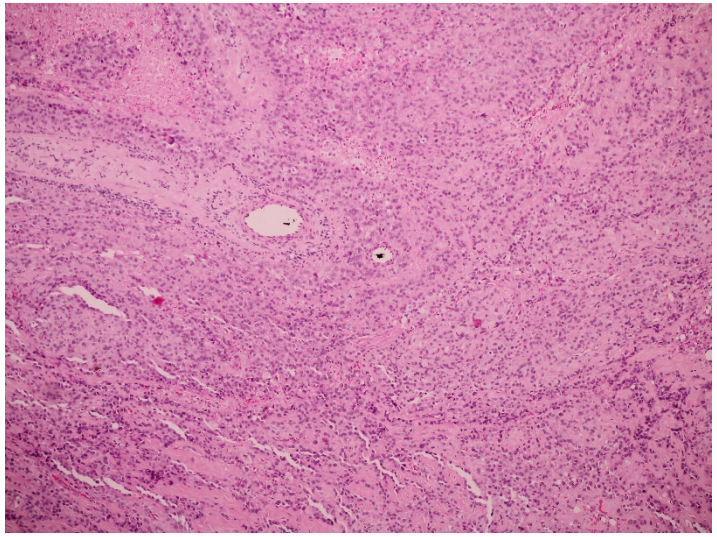
Physical examination:
HEENT: not pale conjunctivae, anicteric sclerae, no lymphadenopathy
CVS&RS: normal
Abdomen: soft, not tender, no hepatosplenomegaly
Extremities: no pitting edema

Skin examination:

- Solitary asymptomatic red to violaceous indurated plaque on scalp.



Histopathology: (S16-21071A, scalp)



- Multiple sheets and diffuse aggregations of atypical epithelioid cells with pleomorphic nuclei, atypical mitotic figures and pale abundant cytoplasm
- Some tumor cells lined along abdominal vascular channels

Immunohistochemistry: tumor cells, markedly positive EGR, Fil-1, focally positive CD31, CD34, negative S100, AE1/AE3

Diagnosis: Recurrent cutaneous epithelioid angiosarcoma

Investigation:

CBC: WBC 7,300/cumm (N 67%, L 20%, Mo 5%, Ba 1%, Eo 7%), Hb 13.2 gm/dl, Hct 40%, Plt 243,000 /cumm
FBS: 210 mg/dl, HbA1C: 7.8%, BUN/Cr: 22/1.09 mg/dl
LFT: TB 1U/L, DB 0.6U/L, AST 26U/L, ALT 20U/L, ALP 147U/L, Alb 30mg/L
CXR: normal
MRI brain: few old lacunar infarction at right cerebellar hemisphere and small cortical infarction at frontal lobe

Treatment: Wide excision with rotational scalp flap.

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Discussion:

Angiosarcoma is a malignant tumor derived from endothelium that occurs in a variety of anatomic sites including the skin and account for less than 1% of all sarcomas. In contrast to other sarcomas, they have a predilection for skin and superficial soft tissue, especially in the head and face region.[1,2] Epithelioid angiosarcoma (EAS) is a rare variant of cutaneous angiosarcoma accounting for about 12% of cutaneous angiosarcoma. EAS mainly occured in deep soft tissues, but visceral and primary cutaneous lesions are documented. Conventional angiosarcoma frequently occurs on the scalp of elderly patients.[3] However, EAS lesions may occured in younger patients including children. There is a male predominance. Clinically, lesions of cutaneous epithelioid angiosarcoma are indistinguishable from conventional angiosarcoma. Patient usually present with solitary, or rarely multiple blue to violaceous nodules, ranging in size from 8 to 80 mm. The most common site for EAS is lower extremity. However, lesions on the scalp and face have also been described. The possible causes of this neoplasm include radiation exposure and reaction to a foreign body.[4,5,6] Cutaneous metastases of angiosarcoma are less than 1% of all soft tissue sarcomas. Therefore, cutaneous metastases of ESA are rare. ESA has been reported to metastasize to lung, bone, soft tissue, lymph nodes, brain, intestine, and skin.[10]

Histopathology of EAS mimics an epithelial neoplasm. Focal epithelioid change is frequently seen in angiosarcomas, but EAS is tumor in which greater than 80% to 90% of tumor cells are epithelioid. Histopathology of EAS characterized by a poorly circumscribed proliferation of atypical epithelioid cells arranged as single units and as nests and irregular aggregates with enlarged pleomorphic nuclei and prominent nucleoli with pale eosinophilic to amphophilic cytoplasm. There were typical and atypical mitotic figures. In some areas, epithelioid cells exhibit cytoplasmic vacuoles as an expression of primitive luminal differentiation.[5] Immunohisto-chemistry contributes to the demonstration of factor VIII-related antigen, ulex europaeus I lectin (UEA-1), ERG, CD31, and CD34. Most cases of ESA are positive for CD31, friend leukaemia integration-1 (FLI-1) and INI-1. EAS may be negative for vascular markers such as CD34 and factor VIII related antigen. However, these tumors also consistently express cytokeratins 25%,

attributable to the abundance of intracytoplasmic intermediate filaments in neoplastic cells, and this phenomenon may result in misinterpretation of this angiosarcoma as a carcinoma.[7,8,13] A small number of EAS with aberrant S100 protein expression are reported.[9]

Staging investigations in EAS are required to exclude extension of angiosarcoma from deep soft tissues. Cutaneous metastases from primary cardiac and mediastinal EAS have been described. In cases with multiple lesions involving skin, lymph nodes, and internal organs, it may be impossible to decide whether the lesion is primary cutaneous or metastatic.[10,11]

Currently there was no standard therapeutic protocol for cutaneous epithelioid angiosarcoma. The choices of treatment include surgery, radiotherapy, and chemotherapy, singly or in combination. The small number of reported cases to date precludes determination of the optimum treatment regimen at this stage, although where possible, wide excision is recommended. The need for adjuvant therapy is determined on an individual basis.[12,13]

The difference in the prognosis between epithelioid angiosar-coma and ordinary angiosarcoma is still controversial. Some reports suggest that epithelioid angiosarcoma is a lower grade malignancy than ordinary angiosarcoma,[4] while other reports suggest that epithelioid angiosarcoma has a high risk of recurrence and metastasis.[8,14]

In summary, we present a case of elderly man who had medical history of angiosarcoma on left cheek treated with wide excision and local flap 10 years ago, he presented with solitary asymptomatic indurated plaque on scalp for 1 year. The clinical, histopathological and immunohistochemistry findings suggested the diagnosis of cutaneous epithelioid angiosarcoma. Treatment is wide excision with rotational scalp flap.

References

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