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Case Report

Primary epithelioid angiosarcoma of the adrenal gland: Report of a case and review of the literature

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ABSTRACT

Primary mesenchymal neoplasms of the adrenal gland are extremely rare. The most common primary sarcoma is adrenal angiosarcoma. We report the case of a 51-year-old patient who presented with left flank pain. After ultrasound investigations, the patient underwent surgical removal of an adrenal tumor. The gross adrenal specimen showed extensive cystic changes with old hemorrhage and necrosis. Histologically, irregular branching vascular channels were seen intermixed with solid areas of epithelioid cells. Immunohistochemical staining of tumor cells was positive with antibodies to cytokeratin and CD31. After a 12 months follow-up, the patient is still well with no sign of a relapse. Because of the epithelioid appearance and frequent expression of epithelial immunohistochemical markers, primary epithelioid angiosarcoma of the adrenal gland can be confounded with adrenal carcinomatous metastases. We report our findings with a brief literature review and discussion of differential diagnosis.

KEY WORDS: Adrenal gland, CD31, cytokeratin, epithelioid angiosarcoma, surgical treatment

INTRODUCTION

Angiosarcoma is a rare malignant tumor and it accounts <1% of sarcomas. It is mainly localized in the skin and superficial soft tissue. Adrenal localization is extremely rare and can easily be misdiagnosed, both by the clinician and the pathologist. We report a new case of primary epithelioid angiosarcoma (EA) of the adrenal gland in a 51-year-old man. A brief literature review and differential diagnosis were made.

CASE REPORT

A 51-year-old man was admitted for left flank pain with abdominal distention of 2 days duration. The patient did not have a history of hypertension or diabetes nor have exposition to chemicals. A physical examination revealed a left flank mass without any lymph nodes.

Computed tomography (CT) of the abdomen revealed a sharply delimited space-occupying mass extending from the spleen and tail of the pancreas to the upper pole of the kidney. This mass was cystic with calcified walls and measured 15 cm at its greatest diameter. No evidence of local tissue invasion or metastatic spread was seen.

The patient underwent laparoscopic total adrenalectomy, with no post-operative complications.

In the surgical exploration, severe adhesions of the adrenal mass to the left kidney forced us to perform a concurrent left nephrectomy in the operating room.

The gross adrenal specimen showed extensive cystic changes with old hemorrhage and necrosis. After serial sectioning, a focus of solid lesion was identified that contained yellow central necrosis [Figure 1].

On histological examination, irregular branching vascular channels were seen intermixed with the solid areas and dissociating residual cortex. They were lined by plump epithelioid endothelial cells with a moderate degree of pleomorphism, a variable amount of eosinophilic cytoplasm, and a few mitotic figures. Intracytoplasmic lumina were seen, some of which contained erythrocytes [Figure 2a].

On the periphery, a thinned residual cortex surrounded this proliferation. Large areas of hemorrhage and necrosis were present [Figure 2b].

Immunohistochemical staining of tumor cells was positive with cytokeratin AE1/AE3 [Figure 3a] and CD31 [Figure 3b] and negative for S-100, chromogranin, HMB 45, Melan-A, and synaptophysin. Therefore, we made the diagnosis of EA.

After surgical removal of the tumor, adjunctive chemotherapy or radiotherapy was not considered because it was believed that the entire tumor had been removed. To date (1-year later), the patient is well and free of tumor.

Figure 1: Gross cut section of the hemorrhagic and cystic tumor of the adrenal gland. A focus of solid lesion that contained yellow central necrosis is present

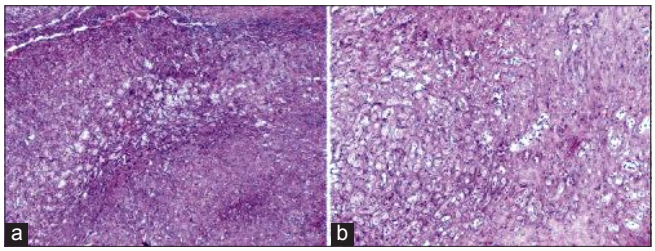

Figure 2: Histological features of the tumor. (a) Low power view of the tumor. The tumor shows residual cortex dissociated by irregular branching vascular channels with areas of necrosis (H and E, ×100) (b) Tumoral proliferation of cells arranged in tubules with red blood cells inside or in solid foci (H and E, ×200)

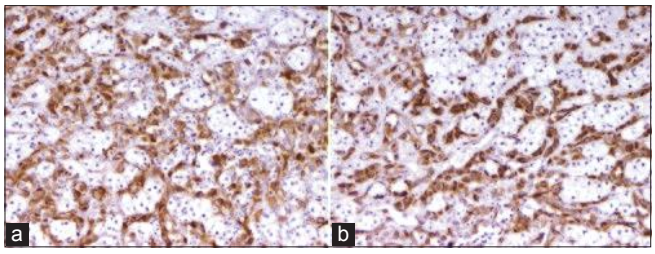

Figure 3: Immunohistochemical features of the tumor. (a) Cytokeratin stain highlights the vascular endothelial cells. Residual cells are free of this antigen (H and E, ×400). (b) The same tumor cells are strongly positive for CD31 (H and E, ×400)

DISCUSSION

Angiosarcoma is a rare neoplasm that accounts for less than 1% of sarcomas. EA is an extremely rare subtype of angiosarcoma, which is characterized by large cells with an epithelioid morphology. Adrenal localization is very rare. Only several cases of adrenal angiosarcoma have been reported since the first case described in 1988 [1].

Direct exposure to arsenic has been suggested to play a role in the pathogenesis, but the etiology of the EA remains unknown [2]. Patients were on average 60 years (45-85 years). Men are the most frequently affected [3].

The most commonly reported symptoms have been pain and abdominal mass. Other complaints have been weight loss, fever, and weakness [3,4]. The radiological features of EA are non-specific [1].

Grossly, the neoplasms varied from solid to cystic in appearance ranging in size from 5 to 10 cm [4]; although our case was an almost exclusively cystic mass that measured 15 cm at its greatest diameter, we find a focus of solid lesion. So that detailed attention to the gross specimen is necessary to identify the solid angiosarcoma component.

The diagnosis is supported by histopathology and immunohistochemistry. EA is characterized by large cells with an epithelioid morphology. The cells had variable amount of eosinophilic cytoplasm and vesicular nuclei, many with prominent eosinophilic nucleoli.

This pseudo-carcinomatous appearance is misleading. Furthermore, immunophenotype makes confusion with an expression of epithelial markers. Indeed, reactivity for cytokeratin is typical of epithelioid morphology [3,5,6]. Thus, EA should not be diagnostically confusing with metastatic carcinoma especially as EMA is in contrast negative, which is characteristic of angiosarcoma [4,5]. Immunohistochemical staining of tumor cells was positive with endothelial markers including CD34 and CD31 [4,5].

The treatment of choice is surgical excision. Radiotherapy and chemotherapy are rather reserved for cases of local recurrence after surgical recovery or metastasis [1,4,5].

CONCLUSION

Angiosarcoma is a rare adrenal tumor, sometimes difficult to diagnose especially in its epithelioid form. Careful screening for vascular differentiation and immunohistochemistry with endothelial markers are essential for the diagnosis. Surgery represents the only possible curative treatment procedure known so far.

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