Case Report

Anastomosing Hemangioma of the Ovary Clinically Masquerading as Epithelial Malignancy: A Rare Case Report

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Abstract

Anastomosing hemangioma (AH) is a rare vascular tumor, which has a predilection for the genitourinary system. Ovarian AH is rare, only few cases have been reported in literature so far. Here, we report a case of 50-year-old woman with right ovarian mass clinically diagnosed as ovarian epithelial malignancy. We received a specimen of hysterectomy with bilateral salpingo-oophorectomy. Grossly, the right ovary showed a well-demarcated solid and spongy lesion with congested areas which was continuous with a cystic lesion, the wall of which showed luteinization. Microscopy revealed a vaguely lobulated lesion composed of anastomosing capillaries with sinusoidal pattern lined by cytologically bland endothelial cells with hobnail appearance in an edematous and hyalinized stroma. Focal areas showed fibrin thrombi within the capillaries. Immunohistochemically, the endothelial cells were strongly positive for CD31 and CD34. The surrounding ovarian parenchyma showed stromal luteinization.

Keywords: *Anastomosing hemangioma, CD31, luteinization, ovary*

Introduction

Anastomosing hemangioma (AH) is a benign vascular tumor, considered to be a rare variant of capillary hemangioma. It was first described by Montgomery and Epstein in 2009 in kidney and testis.[1] Since then only a handful of cases have been reported in literature. Although AH has a predilection for the genitourinary tract, other rare sites include paravertebral region, adrenal, gastrointestinal tract, and liver.[2-7] Histologically, the tumor is characterized by anastomosing sinusoidal capillary sized blood vessel lined by bland endothelial cells with hobnail appearance, in a hyalinized and edematous stroma. The cells can exhibit minimal atypia which can be confused with low-grade angiosarcoma. Here, we report a case of AH arising from the ovary with stromal luteinization.

Case Report

A 50-year-old woman came to the hospital with chief complaints of abdominal discomfort and pain. Abdominal examination revealed mild ascites. Computed tomography showed a well-defined heterogeneous poorly enhancing mass measuring 5.5 cm × 4.5 cm × 4.3 cm in the right ovary and mild ascites. Serum CA125 levels were within normal limits. Clinically, a diagnosis of malignant ovarian epithelial malignancy was considered, following which total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed, and the specimen was sent for histopathological examination. Grossly, the right ovary showed a well-demarcated solid and spongy tan lesion with focal hemorrhagic specks measuring 3 cm × 2.5 cm × 1.5 cm. The residual ovary showed cystic change with yellowish discoloration [Figure 1a]. Microscopically, sections from the right ovary showed a vaguely lobular lesion composed of closely packed capillary sized blood vessels lined by bland endothelial cells with hobnail appearance, arranged in anastomosing or sinusoidal pattern in a hyalinized and edematous stroma. Few capillaries showed fibrin thrombi. Scattered mononuclear cells were seen within

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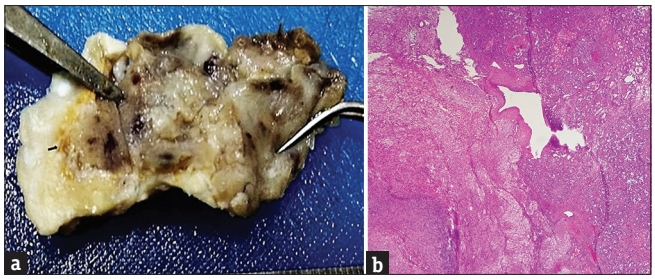

Figure 1: (a) Gross photograph of right ovary showing solid and spongy tan-colored lesion with hemorrhagic specks. The adjacent area showing luteinization (yellowish discoloration-arrow). (b) Microscopic examination scanner view showing vaguely lobulated lesion with adjacent ovarian parenchyma shows stromal luteinization (H and E, ×4)

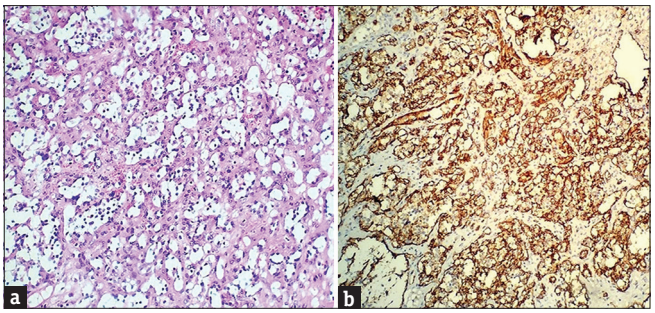

Figure 2: (a) Microscopic examination showing closely packed capillaries lined by bland hobnail endothelial cells. Mononuclear cells are seen within the lumen of capillaries and in the stroma (H and E, ×20). (b) Immunohistochemistry - CD31 showing intense membranous positivity for endothelial cells (×20)

the capillaries and stroma. The residual ovarian stroma showed stromal luteinization [Figures 1b and 2a]. Immunohistochemically, the endothelial cells showed strong positivity for CD31 [Figure 2b] and CD34 confirming the vascular origin of the lesion. The stromal cells were negative for these markers.

Discussion

Vascular tumors of the ovary are rare, with cavernous hemangioma being the most common, vascular tumors.[8] AH is a newly described variant of capillary hemangioma which has a predilection for genitourinary tract especially in the kidney. It was first described by Montgomery and Epstein in 2009 on a series of six cases in kidney and testis.[1] Since its first description, only few cases have been reported in the ovary.[2,5,6] Other reported sites include paraspinal region, retroperitoneum, liver, adrenal, and rarely in the gastrointestinal tract.[2-7] Most lesions are diagnosed incidentally, however, it can be associated with pseudo-Meig syndrome, endometrial hyperplasia, or carcinoma and clinically present as abdominal mass, ascites, or elevated CA125 levels.[2] In such cases, it can masquerade as malignant epithelial tumor both clinically as well as radiologically. Our case presented with abdominal pain and mild ascites.

Histologically, AH is characterized by a vaguely lobular mass composed of tightly packed capillaries arranged in sinusoidal or anastomosing pattern which resembles red pulp of the spleen. The lining endothelial cells are bland with hobnail appearance, lack atypia and shows hyaline globules in some cases. The hyaline globules represent secondary lysosomes or thanatosomes. Fibrin thrombi and extramedullary hematopoiesis has been described in few cases.[1-7] Our case had hobnail appearance and fibrin thrombi. Cases with lipomatous differentiation have also been described by few authors.[5,9] Stromal luteinization in the adjacent ovary is a common finding associated with ovarian hemangioma[5,6,9] which was concordant with our findings as well.

The pathogenesis of ovarian hemangioma is still controversial. Some authors believe that hyperestrogenism due to stromal luteinization has growth stimulatory effect on vasculature. Others propose that associated stromal luteinization is due to mass effect of hemangioma which acts like an enlarged follicle inducing pressure effect on the adjacent ovarian tissue leading to the development of theca-like luteinization.[10] Recently, Bean *et al*.[11] have shown recurrent GNAQ mutations in AH, identical to those found in capillary hemangioma implying its clonal nature.

The differential diagnosis of AH in ovary includes low-grade angiosarcoma, nonneoplastic vascular proliferation of ovarian hilus. Angiosarcoma is an infiltrative lesion, wherein the lining endothelial cells exhibit nuclear atypia, mitosis, solid proliferation of endothelial cells and necrosis. The presence of sinusoidal or retiform pattern, bland nuclei, hobnail cells, hyalinized, and edematous stroma along with stromal luteinization like in our case initially prompted us to consider the possibility of microcystic stromal tumor. However, the presence of red blood cells, fibrin thrombi within the lumen of capillaries, and immunohistochemically CD31 and CD34 positivity and negative CD10-positive tumor cells rules out the diagnosis of microcystic stromal tumor. Another differential diagnosis of ovarian AH includes yolk sac tumor. The latter is histologically characterized by primitive cells with hyperchromatic nuclei with brisk mitosis lining the anastomosing spaces and small cysts.

Conclusion

AH is a rare benign vascular lesion, which can clinically present with symptoms similar to ovarian epithelial malignancies. Awareness of this benign entity and correct diagnosis is essential, as it determines patient management. In addition, the pathologist should be aware of the association with stromal luteinization, to

avoid misinterpretation as steroid cell tumor with stromal vascularization or mixed stromal–vascular tumors.

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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