Case Report

Anastomosing Hemangioma of Ovary with Stromal Luteinization Masquerading as Sex Cord-Stromal Tumor on Intraoperative Consultation

Shivani Jha, Pragya Jain, Sonali Dixit, Sonal Sharma
Department of Pathology, UCMS and GTB Hospital, Delhi, India

Abstract

Anastomosing hemangiomas (AHs) are benign vascular tumor with rare occurrence in the ovary and the majority being asymptomatic. We report one such case of AH with stromal luteinization. A 35-year-old female had complaints of abdominal pain and heavy menstrual bleeding for 2 years. Her lactate dehydrogenase was markedly raised. Following the clinical suspicion of germ cell tumor, exploratory laparotomy and right salpingo-oophorectomy were done. On frozen section due to extensive stromal luteinization, diagnosis of sex cord-stromal tumor was suggested. However, the case was finally diagnosed with AH with extensive stromal luteinization. This case highlighted the potential mimics of AH due to coexistent raised biomarkers and secondary changes, thereby posing a diagnostic dilemma on intraoperative consultation.

Keywords: Anastomosing hemangioma, frozen section, ovary, stromal luteinization

Introduction

Hemangiomas are benign vascular tumors with rare occurrence in the ovary.[1,2] Only a few cases have been reported in the literature so far.[1] They have variable clinical presentation ranging from asymptomatic incidental findings during gynecological surgery to large mass presenting in emergency as torsion.[1,2] Sometimes, they present with extensive ovarian stromal luteinization due to hormonal stimulation (estrogen receptor, progesterone receptor, and androgen receptor) which can lead to erroneous diagnosis on intraoperative consultation.[3] Here, we present the case of a 35-year-old woman, P2 L2A1, who underwent hysterectomy and salpingo-oophorectomy for suspected ovarian malignancy and was finally diagnosed as anastomosing hemangioma (AH) on permanent sections.

Case Report

A 35-year-old female presented to the gynecological outpatient department with chief complaints of abdominal pain and heavy menstrual bleeding for 2 years. Informed and written consent obtained from the patient. Abdominal examination revealed a firm mobile mass of smooth surface measuring 10 cm × 10 cm in the right iliac fossa. The uterus was anteverted, and bilateral fornices were free. Tumor marker analysis revealed increased lactate dehydrogenase (LDH) (977 IU/ml), whereas other markers such as CA 19-9, CA-125, and carcinoembryonic antigen were normal. Mass was solid with minimal vascularity on transabdominal ultrasonography. The patient underwent exploratory laparotomy with right-sided salpingo-oophorectomy with clinical suspicion of germ cell tumor. Grossely ovarian mass was lobulated, solid and hemorrhagic measuring 11 cm × 8 cm × 4 cm. No cystic/papillary areas were seen. The specimen was received in histopathology for rapid diagnosis [Figure 1a]. Frozen histopathology sections revealed predominantly cellular ovarian stroma having scattered luteinized cells with clear to eosinophilic cytoplasm and occasional foci of

Address for correspondence: Dr. Sonal Sharma,
Department of Pathology, UCMS and GTB Hospital, Delhi, India.
E-mail: sonald76@gmail.com

Received: 27-11-2021 **Revised:** 07-05-2022
Accepted: 09-05-2022 **Published:** 04-08-2022

Access this article online

Quick Response Code:

Website: http://www.jmau.org/

DOI: 10.4103/jmau.jmau_92_21

How to cite this article: Jha S, Jain P, Dixit S, Sharma S. Anastomosing hemangioma of ovary with stromal luteinization masquerading as sex cord-stromal tumor on intraoperative consultation. J Microsc Ultrastruct 2022;10:208-10.

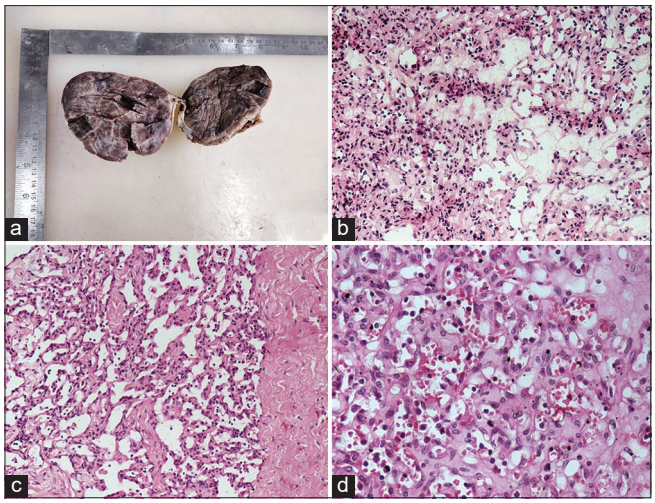

Figure 1: Photomicrograph shows (a) gross pictures of ovary showing solid tan-colored tumor with hemorrhagic specks, (b) frozen section (H and E, ×20) showing stromal luteinization, and (c and d) paraffin-embedded sections (H and E, ×20 and × 40) show numerous anastomotic vascular channels lined by endothelial cells with intervening hyalinized stroma

vascular channels [Figures 1b and 2a]. The possibility of sex cord-stromal tumor was suggested. However, on permanent sections later on (taken after overnight fixation of specimen in 10% phosphate-buffered formalin), well-encapsulated tumor was seen, consisting of variable-sized, thin-walled, blood-filled vascular spaces anastomosing each other. These vascular spaces were lined by bland single-layered endothelial cells. The surrounding ovarian stroma showed extensive luteinization with abundant theca cells having clear vacuolated to eosinophilic granular cytoplasm. In few areas, stroma was hyalinized and edematous at places [Figures 1c, d, and 2b]. This anastomosing pattern of vascular channels was highlighted after immunohistochemical staining with CD34 [Figure 2c]. Stromal cells were negative for CD34. No nuclear atypia/mitosis was seen. There was no evidence of epithelial malignancy or teratomatous areas in the multiple sections. The final diagnosis of AH was made based on characteristic histopathology.

Discussion

Ovarian hemangiomas are rare benign vascular tumors arising due to failure in canalizing process.[1] The first case of ovarian hemangioma was described by Payne *et al*. in 1869.[2] Cavernous hemangioma is the most common subtype of hemangioma occurring in the female genital tract, whereas only a few cases of other subtypes such as capillary or anastomosing are reported in the literature.[4] AH is the rare type of capillary hemangioma having a capillary or sinusoidal pattern, firstly described by Montgomery and Epstein in 2009 in the genitourinary system.[5] Only a handful cases of ovarian AH are being reported in the literature.[6] Here, we are presenting one such rare case of AH with extensive stromal luteinization.

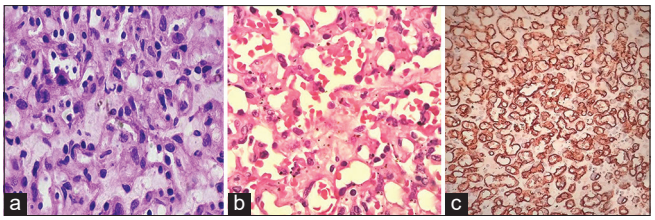

Figure 2: Photomicrograph shows (a) frozen section (H and E, ×100, oil immersion objective) shows scattered luteinized cells with clear to eosinophilic cytoplasm and occasional foci of vascular channels, (b) paraffin-embedded section, taken after specimen fixation (H and E, ×100, oil immersion objective) showing vascular channels lined by endothelial cells with intraluminal red blood cells, and (c) (IHC, ×40) anastomosing vascular channel highlighted by CD34

Majority of the hemangioma cases are nonfunctional lesions of the ovary; however, few cases are associated with stromal luteinization or endometrial hyperplasia, suggesting some hormonal stimulation.[4] Etiopathogenesis of the association between ovarian hemangioma and stromal luteinization is still debatable. Various hypotheses have been proposed to justify this association. Some authors proposed that endothelial cells of hemangiomatous component secrete stroma-stimulating substances leading to luteinization which, in turn, secrete hormones such as estrogen, progesterone, or androgen for sustainable growth of hemangiomatous component. Some authors suggest that enlarging hemangioma causes pressure effect on adjacent ovarian tissue leading to theca and luteinization.[7] In the present case, the heavy menstrual bleeding could be attributed to the luteinized stroma producing excess hormones leading to endometrial hyperplasia.

Sometimes, ovarian hemangiomas are associated with raised tumor markers such as CA-125 where they masquerade as surface epithelial on preoperative imaging.[1,4] The present case was suspected as a germ cell tumor clinically due to raised LDH.

Grossly hemangiomas have a smooth glistening outer surface and spongy blood-filled multiloculated cystic cut surface. The ovarian hilum and medulla are the most common location of ovarian hemangioma. The area itself is highly vascular having large abundant vascular channels, and in postmenopausal females, these vessels proliferate and become closely packed, thereby masquerading hemangioma. However, these normal vascular alterations are often small, more diffuse, and admixed with lymphatic and nerve fibers, whereas vascular proliferations of hemangioma are large and often lead to distinct mass formation.[4] In our case, the mass was large with no normal ovarian tissue visible grossly.

The various differential diagnoses for ovarian hemangiomas are sclerosing stromal tumor, microcystic stromal tumor, lymphangioma, and hemangioma as a part of teratoma.[2,8] Sclerosing sex cord-stromal tumor has pseudolobular architecture due to hypocellular and hypercellular areas. They are mainly composed of round–to-oval luteinized cells and spindle cells with interspersed hemangiopericytomatous vessels.

Immunohistochemically, these cells are positive for vimentin, inhibin, and alpha-smooth muscle actin.[9] Microcystic stromal tumor is another benign sex cord-stromal tumor comprising small-to-large coalescent microcystic spaces forming large channels mimicking vascular spaces of hemangioma. The cellular areas are intersected by paucicellular hyaline and edematous stroma. Our case also had these paucicellular hyaline and edematous areas with large anastomotic blood vessels mimicking large cystic spaces of microcystic stromal tumor. On immunohistochemistry, microcystic stromal tumors are CD10 and vimentin positive, and weak or focally positive for sex cord-stromal marker (calretinin or inhibin).[8] The diagnosis of lymphangioma was refuted by the absence of pale homogenous material within vascular channels.

Hemangiomatous component of teratoma also enters the differential diagnosis, and careful inspection of teratomatous component is imperative to rule this differential.[10] In addition, various vascular endothelial markers such as CD31 and CD34 can also be used to differentiate and highlight the anastomosing vascular channels.

Among ovarian tumors, sex cord-stromal tumors are the most common tumors associated with functional stroma. In our case, ovarian hemangioma is associated with stromal luteinization, possibly due to excessive estrogen production which possibly explains its misdiagnosis as sex cord-stromal tumor on intraoperative consultation. Rezk *et al*. also reported a similar case of ovarian AH in a 60-year-old female with raised CA-125 and hilus cell hyperplasia. Due to its coexistent secondary changes, it was diagnosed with a stromal tumor on intraoperative consultation.[6]

Frozen sectioning/intraoperative consultation also has its own pitfalls. Limited tissue sampling which may underestimate the tumor element and frozen artifacts that distort the architecture may lead to erroneous diagnosis. Careful gross examination before taking sections and early and rapid freezing may prevent the misdiagnosis. The frozen sections must be followed up by permanent sections as a part of quality control. In our case, on the frozen section, ovarian mass was signed out as benign sex cord-stromal tumor which came out to be ovarian hemangioma on permanent sections. Since both are benign entities, the discrepancy did not have any adverse effect on patient outcomes.

Conclusion

This case highlighted the rare presentation, potential mimics, and difficulty in intraoperative diagnosis of hemangioma at this site. Careful gross examination and meticulous microscopic examination are advised for the correct diagnosis to prevent overtreatment and devising adequate follow-up strategy.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Mitra B, Sengupta S, Rai A, Mehta J, Quader AR, Roy S, *et al.* Ovarian haemangioma: A rare case report. Int J Surg Case Rep 2013;4:981-4.
2. Uppal S, Heller DS, Majmudar B. Ovarian hemangioma – Report of three cases and review of the literature. Arch Gynecol Obstet 2004;270:1-5.
3. Metodiev D, Ivanova V, Omainikova B, Boshnakova T. Ovarian anastomosing hemangioma with stromal luteinization: A case report. Akush Ginekol (Sofiia) 2015;54:58-61.
4. Bayramoğlu H, Güngör T, Öz M, Doğan NU, Reyhan H, Beşli M. Primary ovarian hemangioma: Case report and review of literature. Med J Islam World Acad Sci 2012;20:106-10.
5. Subbarayan D, Devaraji A, Senthilnayagam B, Ramanujam S, Nandagopalradha R. Anastomosing hemangioma of the ovary clinically masquerading as epithelial malignancy: A rare case report. J Midlife Health 2019;10:48-50.
6. Rezk A, Richards S, Patricia Castillo R, Schlumbrecht M. Anastomosing hemangioma of the ovary mimics metastatic ovarian cancer. Gynecol Oncol Rep 2020;34:100647.
7. Huang RS, Covinsky M, Zhang S. Bilateral ovarian capillary hemangioma with stromal luteinization and hyperandrogenism. Ann Clin Lab Sci 2013;43:457-9.
8. Irving JA, Young RH. Microcystic stromal tumor of the ovary: Report of 16 cases of a hitherto uncharacterized distinctive ovarian neoplasm. Am J Surg Pathol 2009;33:367-75.
9. Bairwa S, Satarkar RN, Kalhan S, Garg S, Sangwaiya A, Singh P. Sclerosing stromal tumor: A rare ovarian neoplasm. Iran J Pathol 2017;12:402-5.
10. Itoh H, Wada T, Michikata K, Sato Y, Seguchi T, Akiyama Y, *et al.* Ovarian teratoma showing a predominant hemangiomatous element with stromal luteinization: Report of a case and review of the literature. Pathol Int 2004;54:279-83.