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Bleeding bilateral ovarian tumor: A rare case report

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Abstract
Ovarian angiosarcomas are extremely rare and probably either occur as a component of carcinosarcomas or arise from teratoma. Ovarian angiosarcomas are generally unilateral. They can occur at all ages ranging from 7 to 81 years. The tumor favours premenopausal women and usually reported in women of reproductive age group. Very occasional cases are reported in postmenopausal women. We present a case of bilateral ovarian angiosarcoma in a 53-year-old female presented as postmenopausal bleeding. Our patient presented with severe abdominal pain for 3 days with a history of postmenopausal bleeding for the past 18 months. Imaging studies of the abdomen showed degenerative fibroids with complex solid and cystic pelvic mass involving both ovaries. Ascites was also noticed. Patient's CA 15.3 level was elevated. Microscopic examination of the resected specimen revealed a high grade malignant spindle cell lesion seen to be involving both the ovaries, uterus, fallopian tubes and omentum. Owning to vimentin and CD immuno histo chemical positivity, a diagnosis of epithelioid angiosarcoma involving both ovaries, fallopian tubes and uterus with omental deposits. Involvement of the lungs was also identified in the form of pleural effusion.

Keywords: angiosarcoma, ovary, bilateral, post menopausal

Introduction
Soft tissue sarcomas amount to <1% of all cancers. Sarcomas of the reproductive organs are very occasional and have bad patient outcomes. Among them, most are sarcomas of the uterus. Ovarian sarcomas represent only <1% of them with most of them seen as a component of carcinosarcomas [1]. Two percent of all soft tissue sarcomas are angiosarcomas [2]. They are usually situated in skin, deep soft tissue, viscera and breast. Cutaneous angiosarcoma occuring in the head and neck region in elderly people is most common. Angiosarcomas of the viscera occur in liver, spleen, heart, adrenal gland and thyroid gland. Other rare locations include vagina, vulva, cervix and very occasionally ovary.

Ovarian angiosarcomas are extermely rare. They probably either occur as a component of carcinosarcomas or arise from teratoma. It is a well-known fact that sarcomas can arise from germ cell tumors although the mechanism underlying is not clear. Such sarcomas can be a primary tumor or appear in recurrent lesions or even occur in metastatic lesions. Proposed theories include dedifferentiation, malignant change of mesenchymal component of teratoma, development from primitive germ cells or arise from blastematous stromal element of yolk sac tumor [2, 3]. Till date, only very few case series of ovarian angiosarcomas have been reported in literature. Out of which, around 25% were seen to be associated with lesions like teratoma and other tumors including epithelial ovarian neoplasms and ovarian fibromas [4-12]. Ovarian angiosarcomas are generally unilateral. They can occur at all ages ranging from 7 to 81 years. The tumor favours premenopausal women and usually reported in women of reproductive age group. Very occasional cases are reported in postmenopausal women [12].

Abdominal pain is the most common presenting complaint although some patients may present with symptoms attributed to distant metastasis more likely in the lungs. Disseminated intravascular coagulation (DIC) and pleural effusion along with ascites are some of the other presenting clinical features. Although ovarian angiosarcomas are commonly unilateral, bilateral cases are also occasionally reported. Advanced stage with disseminated disease at presentation is very common in ovarian angiosarcomas. The disease is usually aggressive with poor prognosis despite surgery and chemotherapy. Very few cases identified at an early stage also progressed to disseminated disease in a span of less than one year [13]. We present a case of bilateral ovarian angiosarcoma in a 53-year-old female presented as postmenopausal bleeding.

Case Report
A 53-year-old female presented with postmenopausal bleeding for the past 18 months. She has 2 kids, last child birth being 15 years ago. She is currently not on hormonal replacement therapy. Last Pap smear done 5 years ago was normal. She is a hypertensive on amlodipine 5mg daily. She is a non-diabetic. Patient's mother had died of lung malignancy.

Our patient recently presented with severe abdominal pain for 3 days and had to be admitted as inpatient for the purpose of pain management. There were no signs of vomiting, fever, loss of weight or loss of appetite. There was no history of any previous menstrual abnormalities. Patient had no significant past medical history. Abdominal examination revealed distended uterus and ascites. Imaging studies of the abdomen showed degenerative fibroids with complex solid and cystic pelvic mass involving both

ovaries. Ascites was also noticed. Patient's CA 15.3 level was elevated to 95.6 mmol/L and CEA level was normal. Other system examinations and investigations were unremarkable.

Surgical resection was done. Intraoperatively, bilateral ruptured, fragile and easily bleeding ovarian tumour was seen. Both tumours were seen adherent at pouch of Douglas, ovarian fossa and bowel. Multiple tumour nodules at bowel mesentery, thickened omentum with no tumour nodules, Normal sized uterus with no fibroids, bilateral enlarged pelvic lymph nodes and hemorrhagic ascites were other notable intraoperative findings. No tumor nodules were seen over the bowel, peritoneum, subdiaphragm and liver surface.

Macroscopic examination of the resected specimen showed bilateral ruptured and hemorrhagic friable ovarian tumours. Uterus showed endometrial polyp with features of adenomyosis. Omentum thickening was noticed. Microscopic examination revealed a high grade malignant spindle cell lesion seen to be involving both the ovaries, uterus, fallopian tubes and omentum (Figure 1 and Figure 2). Pelvic lymph nodes showed no metastatic deposits. A panel of immunohistochemistry markers including vimentin, actin, desmin, s100, CD3, Inhibin, CD99, CD10, and Ki67 was done. Vimetin (Figure 3) and CD31 (Figure 4) were strongly positive in the tumor with 35% proliferative index. A diagnosis of epithelioid angiosarcoma involving both ovaries, fallopian tubes and uterus with omental deposits was made. Involvement of the lungs was also identified in the form of pleural effusion. Patient was administered with adjuvant chemotherapy.

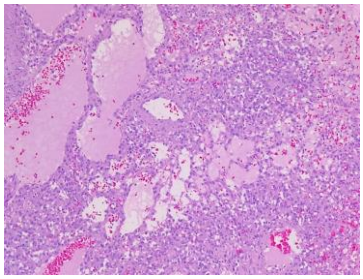

Fig 1

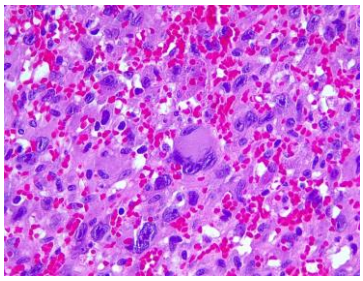

Fig 2

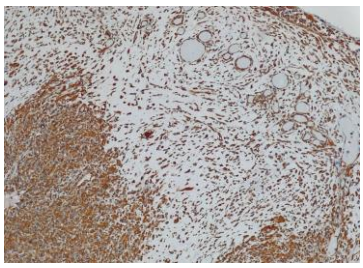

Fig 3

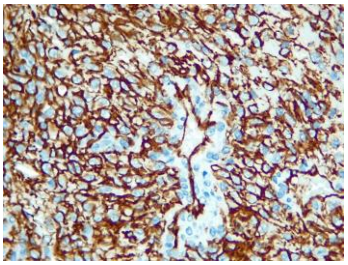

Fig 4

Discussion

Angiosarcomas are uncommon malignant tumors with characteristic features of rapid proliferation, extensive infiltration containing anaplastic endothelial cells. A wide variety of vascular malignant lesions that occur in various organs are encompassed in this category. Over 50% of angiosarcomas are seen to occur in the head and neck region [14]. Gynaecological sarcomas are uncommon amounting to around 4% of uterine and 1% of ovarian tumors [15]. Primary ovarian angiosarcomas are even rarer with fewer cases reported in the literature. Most of the cases reported are in women of child bearing age. Our case is one among the rare reported in postmenopausal age. Like most of the previously reported cases, our patient also presented with abdominal pain [16, 17, and 18]. But unlike other cases, our patient presented with postmenopausal bleeding probably attributed to tumor spread into the uterus. Bleeding PV is common in uterine angiosarcoma but uncommon in ovarian angiosarcoma. Our patient also showed other features of hyperestrogenism such as endometrial polyp and adenomyosis. While the role of estrogen in uterine sarcomas is well established, its association with ovarian angiosarcoma is not known yet [19, 20].

The main challenge in the diagnosis of angiosarcoma is histopathologic identification. The differential diagnosis of angiosarcoma based on histopathologic features should include sex-cord stromal tumors, germ cell tumors, malignant melanoma, undifferentiated carcinoma and metastatic angiosarcoma. Histologically, angiosarcoma varies from low grade tumor cells lining vascular spaces to high grade solid areas containing highly pleomorphic cells [21]. Epithelial markers and HMB 45 can help rule out undifferentiated or poorly differentiated carcinoma and melanoma. Inhibin and calretinin can be done to rule out sex cord stromal tumor. WT1 can be used to rule out ovarian tumor overall including germ cell tumor and sex cord stromal tumor [22, 23, 24].

We did a wide panel of markers (Vimentin, Actin, Desmin, S100, CD31, Pan CK, Inhibin, CD99, CD10 and Ki67). Vimetin and CD31 were strongly positive indicating vascular origin was confirmed. However, angiosarcoma is frequently confused with other vascular tumors of intermediate grade including hemangioendotheliomas and malignant endovascular papillary angioendotheliomas. A diagnosis of angiosarcoma is made in our patient, since CD 31 positivity is mostly attributed to poorly Differentiated vascular tumors [16]. Also, our patient had advanced disease involving the lungs.

Conclusion

We present a rare case of bilateral angiosarcoma occuring in postmenopausal women. Palliative surgical treatment and adjuvant chemotherapy might increase survival in patients

with late stage angiosarcomas. From this case report, we would like to emphasize on the extreme rarity of the bilateral ovarian angiosarcoma occuring in postmenopausal women. The differential diagnosis along with diagnostic markers of ovarian angiosarcoma were also described.

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