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Case Report

Clinicopathological characteristics and oncological outcomes in a case of primary ovarian adenosarcoma with sarcomatous differentiation: a case report and literature review

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ABSTRACT

Mullerian adenosarcoma is a rare malignant tumor generally involving the uterine corpus but can uncommonly involve extrauterine organs. Ovarian adenosarcoma is extremely rare and often diagnosed in young women. Majority of them are low grade with a good prognosis except for adenosarcoma with sarcomatous overgrowth (SO). We report a case of a large sized ovarian adenosarcoma with SO with elevated levels Ca125 and Ca19.9 and having disease free interval of approximately 1.5 years.

Keywords: Adenosarcoma ovary, Mixed mullerian tumors, Ovarian tumors

INTRODUCTION

Adenosarcoma is a rare malignancy from the mixed Mullerian tumor group (MMT), which typically originates from the uterine corpus but can occur in uterine cervix or ovary as well.[1] Ovarian adenosarcoma is very rare and represents <1 % of ovarian tumors.[2] Presence of SO, imparts poor prognosis with a high risk of recurrence.[3] We report a case of a 45-year-old female with ovarian adenosarcoma in early stage, treated with a multidisciplinary approach.

CASE REPORT

A 45-year-old lady reported to us in April, 2023 with complaints of abdominal pain and distension. All routine investigations were done which were within normal limits. On physical examination, a pelvic mass was palpable. The tumor markers were also sent and the results are as follows-CA125: 1273, CA19.9: 2605 and CEA: 1.13.

On further investigation, CT scan of abdomen and pelvis with contrast showed, bilateral ovarian lesion, mild ascites and some omental fat stranding, raising suspicion of an ovarian malignancy. Upper and lower gastrointestinal endoscopy showed no evidence of malignancy. CT scan chest did not show any spread of disease. She underwent exploratory laparotomy with total abdominal hysterectomy, bilateral saplingo-oopherectomy, posterior peritonectomy, omentectomy, bilateral pelvic and para-aortic lymph nodes dissection. Intraoperatively, right ovary showed chocolate cyst of 3×3 cm and moderate ascites.

The histopathology report showed a biphasic tumor measuring 31 cm, with minor benign epithelial component and predominantly spindle sarcoma features involving the capsular surface (Figure 1). The sarcomatous component consist of high grade endometrial stromal sarcoma showing periglandular cellular areas and hypocellular low-grade areas with the Ki-67 for sarcomatous components of 30-35% (Figure 2). Large areas of necrosis were seen with

absence of any heterologous sarcoma components. The mitotic figures were 7-8/ 10 high power field. On immunohistochemistry (IHC), the glandular component expressed positivity for CK7, EMA, estrogen receptor (ER), progesterone receptor (PR) and PAX-8 whereas p53 was wild type. The sarcomatous component expressed ER (Figure 3 A), PR, CD10 (Figure 3 B), Cyclin D1, SMA (Figure 3 C), desmin positivity and high proliferation index (Figure 3 D) and was negative for inhibin, calretinin, MyoD1 and -100p. Thus, a final diagnosis of Adenosarcoma of the ovary was made. The peritoneal lavage was negative for malignant cells and no lymph nodes showed disease spread. The pathological stage was stage IC2. She was referred for adjuvant chemotherapy based on the pathological staging.

She received 6 cycles of adjuvant chemotherapy with injection doxorubicin and ifosfamide with mesna rescue for 3 days once in every 21 days. The patient tolerated the therapy well with limited side effects. The follow up scan post completion of chemotherapy showed no evidence of residual or recurrent disease and patient has been disease free for approximately 1.5 years now.

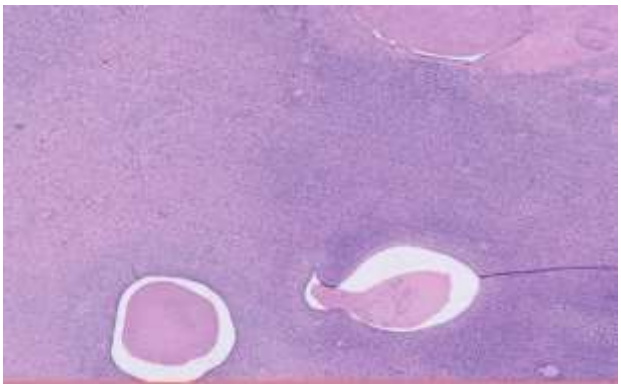

Figure 1: Biphasic tumor showing a minor benign glandular component and a predominant mesenchymal spindle cell component.
An area of necrosis is seen in the top right corner in a haematoxylin and eosin-stained slide at 20×.

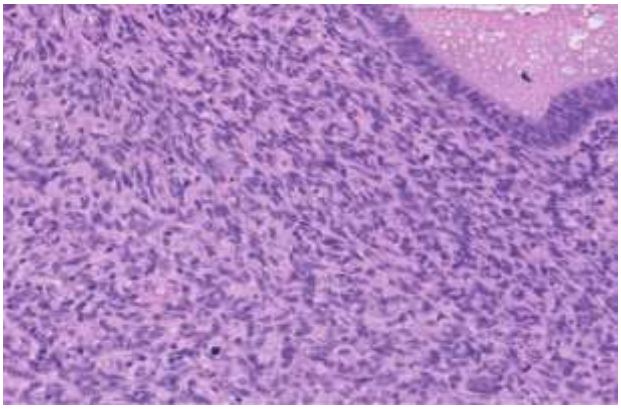

Figure 2: The spindle cell component is hypercellular and shows many mitotic figures.
The glands are benign.

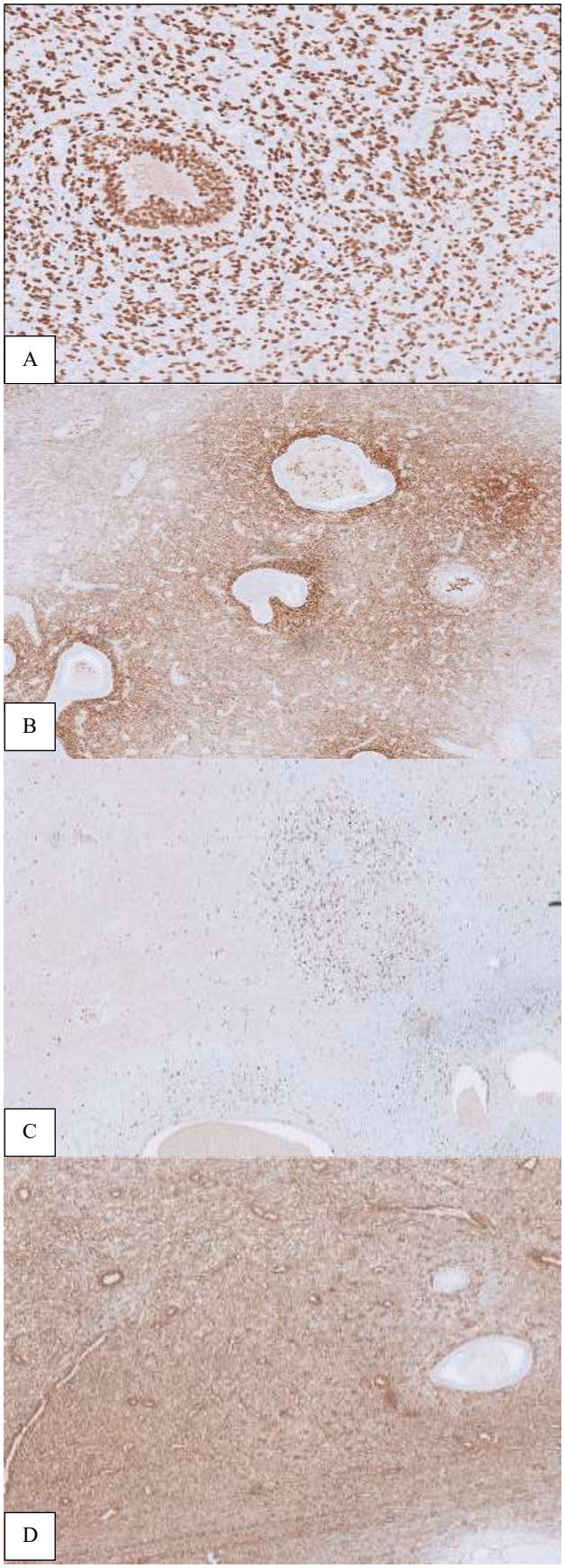

Figure 3 (A-D): The spindle cells express A) estrogen receptor, B) CD10, C) SMA and D) a high Ki-67 proliferation index.

DISCUSSION

Mullerian adenosarcomas are mostly seen in uterus and the extrauterine sites include, cervix, ovary, fallopian tube, pelvic sidewall, peritoneum, round ligament, pouch of Douglas, vagina and colon. Clement and Scully described the first case of uterine adenosarcoma in 1974.[4] Extra-Mullerian adenosarcomas (EMA) are more commonly reported in younger patients as compared to uterine mullerian adenosarcomas.[3]

Ovarian mullerian adenosarcoma (OMA) are very rare and have poorer prognosis than uterine, mainly due to capsular rupture and adhesions. These tumors are characterized by a mixture of benign glandular epithelium and a low-grade malignant sarcomatous stroma resembling that of endometrial stromal sarcoma.[5] The average size of the adenoscaroma of uterus and ovary is reported to be 5 cm and 14 cm respectively.[3] In our case, the mass was 31 cm in diameter. To best of our knowledge, 31 cm is maximum tumor diameter reported in literature. Heterologous elements may be seen which include rhabdomyosarcomatous, chondrosarcomatous, osteosarcomatous/liposarcomatous components. The most common element seen in OMA is rhabdomyosarcomas.[3,6] Unlike other reported cases in literature, our case lacked the heterologous sarcoma components.

SO is defined as partial overgrowth comprising of at least 25% tumor mass in an otherwise typical adenosarcoma. In OMAs, SO rates are comparatively higher than uterine adenosarcoma. The mitotic counts and atypia are evaluated on the area with the highest mitotic activity. Ten or less mitoses at 10 high power field (HPF) as well as mild atypia is defined as low grade whereas more than 10 mitoses as well as severe atypia is included in high grade sarcomas. SO areas are graded on the basis of same criteria.[3] A literature review has shown that the rate of SO in Mullerian adenosarcoma ranges from 8% to 54%.[3] Our case had high grade ovarian adenosarcoma with SO with a Ki67 of 30-35% and 7-8 mitotic index/10 HPF.

Molecular genetic profiling of adenosarcomas revealed genetic heterogenicity of adenosarcoma with recurrent pathogenic driver alterations, including rare cases with ESR1-NCOA2/3 fusions. DICER1 is among the most common mutations. In addition, a subset of high-grade adenosarcomas is known to harbor TP53 mutations with associated aberrant p53 immunohistochemical expression. Mutations of genes within the PI3K/AKT/PTEN pathway, ATRX, FGFR2, and KMT2C along with amplifications of the MDM2/CDK4 locus and BAP1 deletions have been reported in literature. Our patient had no expression of p53 on IHC.[7]

They are often associated with endometriosis. The main clinical symptoms are non-specific and present with vaginal bleeding, abdominal pain, abdominal distension, ascites, and intestinal disorders and hence, are often missed at early stages. A literature review by Mandato et al reported abdominal or pelvic pain as the primary presentation in 14 out of 34 (41 %) symptomatic patients.[8] Eichhorn et al reported an abdominal mass in half of the patients during physical examination.[3] Compatible with these studies, our patient's initial presentations were abdominal pain and distention. Thus, adenosarcoma is difficult to diagnose both clinically and pathologically.

Shakuntala et al mentioned the rarity of elevated CA125 in adenosarcoma.[9] Inoue et al suspected the elevated CA125 to be a probable indicator of SO.[10] Our patient had increased level of both Ca125 and Ca19.9, simultaneously.

The recommended treatment is hysterectomy and bilateral salpingo-oophorectomy i.e. surgical removal in localised cases.[8] Due to rarity of the disease, the role of adjuvant chemotherapy/radiotherapy in avoiding disease recurrence is not defined in literature. However, certain prognostic factors guide in the management of adjuvant therapy. Poor prognostic factors include: i) High grade, SO, ii) <53 years of age, iii) Tumor rupture, iv) ovarian location and iv) Heterologous elements. Our patient was 45-year-old with high grade SO ovarian adenosarcoma with involvement of the capsule, hence, making her a risk candidate.[3] Thus, she was planned for 6 cycles of adjuvant chemotherapy.

The systemic treatment to prevent recurrence or in case of metastatic disease is commonly made with chemotherapy regimens aiming at uterine and soft tissue sarcomas or carcinosarcomas, such as ifosfamide, cisplatin, doxorubicin, and dacarbazine.[8] In our case, we used the most commonly used regimen doxorubicin and ifosfamide for 6 cycles and the patient tolerated the therapy well with minimum side effects. Unlike the cases reported by Azam et al where disease became metastatic just after 3 weeks of surgery and recurred after 4 months of surgery, respectively, our patient had been disease free of approximately 1.5 years despite the presence of poor prognostic factors.[10]

CONCLUSION

Our case reiterates the fact that rare pathological varieties of ovarian neoplasms like adenosarcoma, require an experienced oncopathologist and multidisciplinary tumor board approach for successful management. An adenosarcoma ovary should be considered as a differential diagnosis in patients presenting with a solid or cystic pelvic mass particularly in those who have a history of endometriosis.

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