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High Grade Pleomorphic Leiomyosarcoma of Ovary in Young Female: A Case Report

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Background: Primary leiomyosarcoma of ovary is a very rare tumor with around 55 cases reported so far.

Case Report: A 27 year old female presented in Gynaecology OPD with complaint of acute pain in right iliac region and abdominal distention. Per abdominal examination revealed a soft to firm mass in the right iliac region. Ultrasound imaging revealed a complex mass with solid and cystic areas in the right adnexal region measuring 9.7 x 5.2cm. Laparotomy with right ovarian cystectomy was done. Histopathology report was consistent with High Grade Pleomorphic Sarcoma. The patient was referred to Gynaecological Oncology department where debulking surgery comprising of hysterectomy, left salpingo-oophrectomy and resection of residual tumor mass on right was done with pelvic and para aortic lymph node dissection. IHC was also advised which revealed cytoplasmic positivity for desmin and smooth muscle actin in all the cells. Final diagnosis of Pleomorphic Leiomyosarcoma (High grade) was made. The patient was given chemotherapy 3 weeks after surgery in consultation with medical oncologist. Response to therapy was evaluated after 6 months by whole body CT scan and CA-125 levels both of which were within normal limits. The patient comes for regular follow-up and is doing well after 30 months of surgery.

Conclusions: We found that surgical debulking along with chemotherapy has given good response, and the patient is still surviving and is symptom free. Patient is on regular follow up, after 30 months of surgery.

Introduction

Primary leiomyosarcoma of ovary is a very rare tumor with around 55 cases reported so far and represent less than 1% of ovarian tumors [1]. Pathogenesis is uncertain with many theories including malignant degeneration of an ovarian leiomyoma or of the smooth muscle present in the wall of the blood vessels in the cortical stroma and corpus luteum, muscular attachments of the ovarian ligament, wolfian duct remnants, or totipotential ovarian mesenchyme, or arising in a teratoma. Most cases present in peri- and post-menopausal women between 45 to 60 years of age. These tumors tend to reach a very large size and wide excision is often

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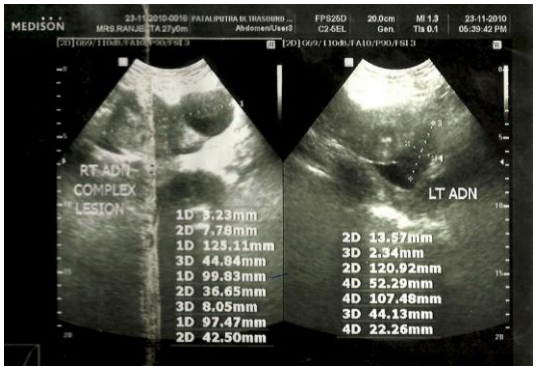

Figure-1: USG showing complex mass in right adnexa

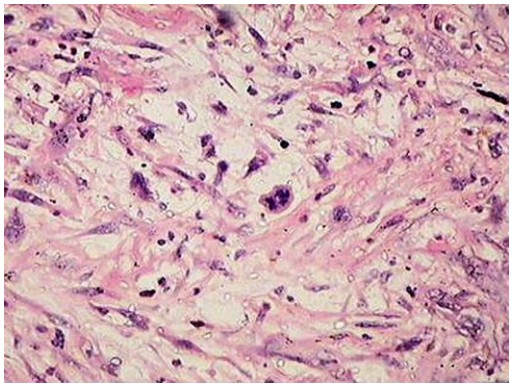

Figure 2: Photomicrograph showing fascicles of brightly eosinophilic spindle cells with ovoid to cigar shaped nuclei intersecting each other at wide angles

impossible. Majority of these tumors are well circumscribed large and softer and have a tendency for necrosis, hemorrhage and cystic degeneration. Histologic features vary with the degree of differentiation and comprise of fascicles of brightly eosinophilic spindle cells with vesicular, ovoid to cigar shaped nuclei intersecting each other at wide angles and showing uniform strong positivity for smooth muscle actin and/or desmin. Most leiomyosarcoma of ovary are highly malignant and spread by local invasion, hematogenous and by lymphatics. Metastasis is mainly to lungs and liver and overall 5 year survival is 20 to 30%. These sarcomas are characterized by marked cellular pleomorphism and brisk mitotic activity and carry a very poor prognosis [2]. Because of its extreme rarity; we present this case of ovarian neoplasm in a young woman of 27 years.

Case Report

A 27 year young female reported in Gynae OPD with complaints of pain in right lower abdomen. Her past medical and obstetric history was uneventful with two full term normal delivery. Abdomen was distended and a bulge was seen in the right iliac region. On palpation, a soft to firm lump was palpated in the right iliac region. On per vaginal examination, a mass was felt in the right fornix with restricted mobility and tenderness. Ultrasound examination revealed a complex solid cystic mass in the right adnexal region measuring 9.7 x 5.2 cm. The left ovary measured 4.4 x 2.2 cm, uterus 10 x 3.7 x 3.7 cm whereas, cervix was homogenous. Mild free fluid was seen in pouch of Duglass (Figure 1). Preoperative routine tests were normal except for mild anemia of with Hb - 8.4gm%. CA125 value was 5.2 IU/mL. Ab-initio, the patient underwent simple partial right cystectomy as malignancy was not suspected. Histopathology report was consistent with High Grade Pleomorphic Sarcoma. Hence, the patient was then referred to Gynaecological Oncology department. Patient was evaluated, Chest X-ray and a CT scan of whole abdomen was done to ensure that there was no primary tumor elsewhere. Serum Beta HCG level was5 mIU/mL On laparotomy the residual mass was found to be originating from the right ovary and was adherent to the bladder. The uterus and left ovary were not involved, however the omentum was adhered with uterus, ovaries and the abdominal wall. The mass was separated from the bladder with some difficulty. Hysterectomy with bilateral salpingo-oophorectomy, total omentectomy

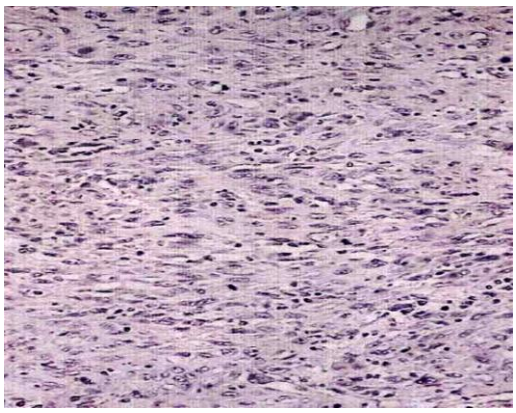

Figure-3: Immunohistochemistry revealed intense cytoplasmic positivity for Desmin in all the cells

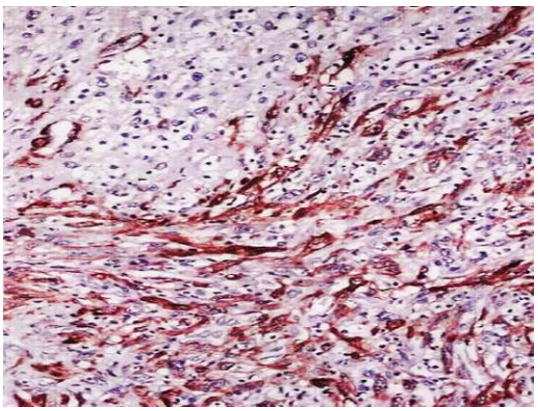

Figure-4: Immunohistochemistry revealed intense cytoplasmic positivity for smooth muscle actin in all the cells

and pelvic and para aortic lymph node dissection was done and sent for Histopathological examination .

There was no residual tumor left after surgery. Histopathology showed fascicles of brightly eosinophilic spindle cells with ovoid to cigar shaped nuclei intersecting each other at wide angles. These cells showed marked atypia and contain pleomorphic nucleus with prominent nucleoli. More than ten mitotic figures per ten high power fields were seen (figure 2). Immunohistochemistry revealed intense cytoplasmic positivity for desmin (figure-3) and smooth muscle actin in all the cells (figure-4.). It was negative for CD 10 and CD 34 and diagnosis of Pleomorphic Leiomyosarcoma (High grade) was conclusively made. The patient was put on chemotherapy after consultation with medical oncologist after 3 weeks of surgery. She was given 6 cycles of docetaxel (80mg/m^2) and gemcitabine (1000mg/ m^2) after evaluating Body

Surface area (BSA), complete blood count (CBC) and serum chemistry (LFT, KFT, CA 125). Chemotherapy was uneventful throughout all the six cycles. Response to therapy was evaluated by CT scan according to WHO response criteria. Serum CA 125 was within normal limits. Patient is on regular follow up and is doing well after 30 months of surgery.

Discussion

Pure primary sarcomas originating in the ovary are rare (< 1%), and only a few cases of fibrosarcoma, leiomyosarcoma, angiosarcoma and other histologic types of sarcoma have been reported. The histology is similar to that of the sarcoma originating elsewhere in the body and the prognosis is usually poor. On gross examination these tumors are indistinguishable from other sarcomas. Usually these tumors are solid but cystic degeneration is often seen in large tumors [3]. Leiomyosarcomas are immunoreactive for smooth muscle actin (SMA), desmin, and caldesmon. They are negative for S-100 protein. About one third of cases exhibit positivity for cytokeratins and epithelial membrane antigen (EMA) and the nuclei of a leiomyosarcoma have blunted or truncated (rather than rounded) ends and cytoplasm is denser [4].

Primary ovarian leiomyosarcomas usually occur in postmenopausal women but this is rare case in which a 27 year young woman was affected. Chun-Chieh Chia *et al.*,

reported a rare type of ovarian sarcoma that occurred in a 60-year-old female. Anil K. Sood, *et al*., reported retrospective analysis of 47 women with primary ovarian sarcomas, S Dixit, S Singhal, reported leiomyosarcoma of ovary was in a 60 year old female. Dai Yi *et al*., Between 1988 and 2007, 24 patients with primary ovarian sarcoma who underwent treatment at Peking Union Medical Hospital were reviewed retrospectively.

The International Federation of Gynecology & Obstetrics (FIGO) staging and treatment of ovarian leiomyosarcoma have been the same as those for ovarian carcinoma [5]. There is no established treatment for these sarcomas other than surgery [6]. Various adjuvant therapies have been proposed , including radiotherapy and chemotherapy, with no additional benefits [7-9].

This particular case needs further labeling by molecular genetics to identify the mutant gene responsible for the tumor. Leiomyosarcoma differs from benign counterpart by hypercellularity, diffuse atypia and presence of increased mitotic rate (more than 5 per 10 high power fields) [8]. Diagnosis of leiomyosarcoma should be strongly suspected in tumors that are overly large, necrotic or hemorrhagic, even if the mitotic count is low. These tumors are most often radio-resistant. Mainstay of treatment is debulking surgery, consisting of total abdominal hysterectomy, bilateral salpingo-opherectomy, and extirpation of the tumor masses. The prognosis of ovarian leiomyosarcoma is extremely poor, and depends on the tumor stage, tumor size, and mitotic index. Taskin *et al*., reported that 63% of stage -1 patients survived with no evidence of the disease after a mean follow-up period of 41.7 months, while 81.25% of patients at a higher stage died after a mean follow-up period of only 14.7 months. The 5-year survival rate was 32% overall, 63% for Endometrial stromal sarcoma**,** 30% for mixed mesodermal sarcomas, and 18% for leiomyo-myosarcoma [9].

Conflict of Interest

The authors declare that there are no conflict of interests.

Authors' contribution

SP performed the literature search and prepared the manuscript.
VC contributed to the pathology part of manuscript
RKS contributed to Medical Oncology part of manuscript
RH prepared the draft manuscript and helped with editing of manuscript

Ethical Consideration

Written informed consent was taken from the patient for publication of this case report.

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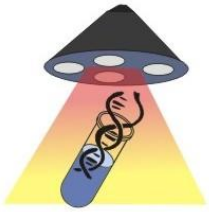

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