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BILATERAL DYSGERMINOMA OF OVARY WITH DIRECT SPREAD TO UTERINE CORPUS AND CERVIX - A RARE CASE REPORT.

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Abstract : Dysgerminomas are the most common primitive germ cell tumors accounting for about 1percentage of all primary ovarian malignant tumors and constitutes 1-5percentage of all ovarian germ cell tumors. Dysgerminomas often occur bilaterally (approximately 10-15 of cases). They may present at any age from any age from infancy to old age but they are extremely rare under the age of 5 years and over the age of 50 years. Most of the cases develop in women younger than 30 years of age (mean, 21 years). We report a case of bilateral dysgerminoma ovary in a 46 years old female with direct spread to entire uterus and cervix. Patient presented with abdominal pain and bleeding pervagina for 6 months duration. USG abdomen reported as enlarged uterus with Subserosal fibroid attached to fundus of the uterus. Total abdominal hysterectomy with bilateral salphingoophoectomy was done. Histopathological examination and Immunohistochemistry confirmed the diagnosis of Dysgerminoma.

Keyword : Dysgerminoma, Germ cell tumor, Ovary.

INTRODUCTION:

Dysgerminomas are the most common malignant germ cell tumor occurring in the ovary and they are found most commonly in adolescents and young adults.1,2,3 The name "Disgerminoma," proposed by Meyer in 1931, properly defines the tumor and eliminates confusion in terminology caused by describing it as embryonal carcinoma, seminoma, sarcoma, and under many other names. Common signs and symptoms of ovarian dysgerminomas include abdominal/pelvic pain (55-85%), abdominal mass (35%), fever (10-25%), vaginal bleeding (10%), and occasionally ascites6. Unlike other germ cell tumors, Dysgerminomas often occur bilaterally in 15% of cases2, 3, and 4,6,13. Lymphatic spread of Dysgerminomas often involves the retroperitoneal and pelvic lymph nodes. Hematogenous spread to distant organs such as lungs, liver and bone occur later4, 6, 12. Direct extension occurs through the capsule of ovary with exfoliation and dissemination of cells through the peritoneal cavity3 .Recurrent disease can occur in 75% of cases within one year of initial treatment. Dysgerminomas are highly susceptible to radiotherapy4, 5, and 6.

CASE HISTORY:

We report a case of bilateral dysgerminoma ovary in a 46 years old P3L3 post menopausal women with direct spread to uterus and cervix, diagnosed by histopathological examination and immunohistochemical analysis. She presented with abdominal pain and bleeding pervagina for 6 months duration. Total abdominal hysterectomy with bilateral salphingoophoectomy was done and the specimen was sent for histopathological examination.

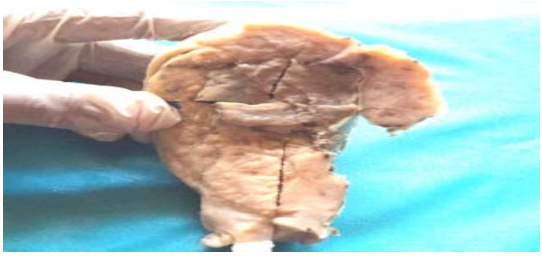

Figure -1 Showing necrotic endometrium with ovary attached to fundus of uterus.

Figure -2 Cut surface of ovary showing granular, creamy white colour with vague lobulation, foci of haemorrahage and necrosis.

GROSS EXAMINATION:

Received Total abdominal hysterectomy and bilateral salphingoophoectomy specimen with the uterus measuring 10 x 9x 4 cm with attached ovary measuring 4 x3 x3 cm(figure-1). Separately received other side ovary measuring 5x 4x 3 cm (figure-3). Cut surface of the endometrium reveals distorted endometrial cavity filled with yellowish necrotic material (figure-1). Cut surface of both ovaries revealed creamy white areas with vague lobulation and areas of hemorrhage and necrosis. (figure2, 3) Cut surface of cervix revealed similar creamy white areas (figure-4).

Figure -3 Cut surface of other side ovary showing solid tumor with lobulation , homogenous creamy white to light tan colour.

Figure-4 Cut surface of cervix shows creamy white areas.

MICROSCOPIC FEATURES:

Histopathological examination of uterus shows proliferative endometrium (figure-7). Section from ovaries shows aggregates of polygonal cells arranged in island and cords resembling primordial germ cells. The cells are uniform, rounded tumor cells have clear to eosinophilic cytoplasm (which is almost always glycogen-rich), discrete cell membranes, and a central, large, rounded or flattened nucleus that contains one or a few prominent nucleoli (figure 5,6). Mitotic figures are also seen. The stroma consists of thin septa to broad fibrous bands infiltrated by mature lymphocytes (figure -5). Non caseating granulomas (figure-7, 8), Langhans- like giant cells are seen in endometrium and in cervix (figure-9). Additionally foci of hemorrhage, necrosis are also identified.

LABORATORY MARKERS:

Elevated serum levels of lactate dehydrogenase (LDH) of 300 U/L(reference range 120-246)

IMMUNOHISTOCHEMISTRY:

Immunohistochemistry: CD117(c-kit) shows strong membrane positivity (figure-11); cytokeratins-weak cytoplasmic positivity (figure-10). With these features, histological diagnosis of bilateral dysgerminoma with direct spread to endometrium and cervix was arrived.

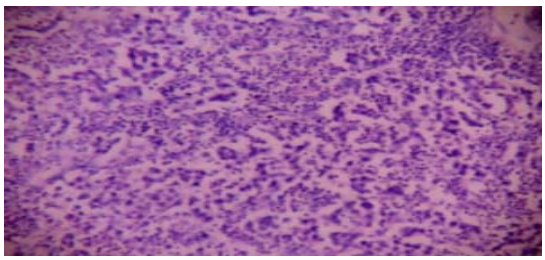

Figure -5 Ovary showing Cords of uniform tumor cells are separated by fine connective tissue septa containing lymphocytes.(10X)

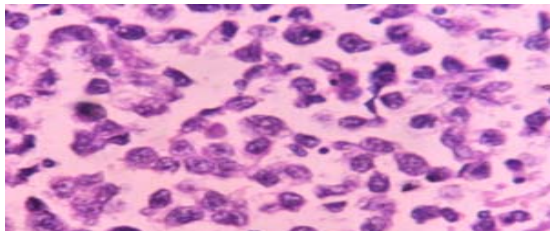

Figure-6. High power view of dysgerminoma cells in ovary showing uniform polyhedral cells with pale eosinophilic cytoplasm and prominent nucleoli.(40X).

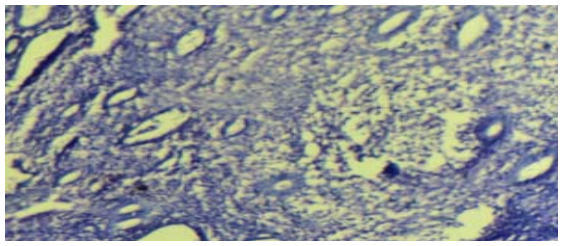

Figure-7. Microscopy of endometrium shows proliferative endometrium and granulomas admixed with lymphocytes (10x).

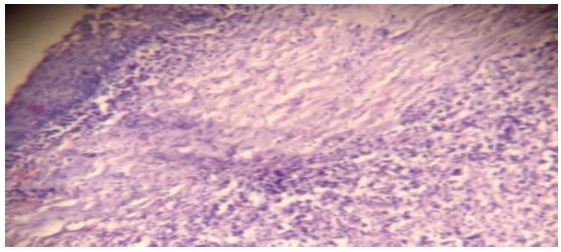

Figure-8. Micoroscopy of cervix shows aggregates of tumor cells admixed with lymphocytes(40X).

DISCUSSION: Germ cell tumors (GCTs) are rare, comprising approximately 30% of all ovarian tumors, both benign and malignant7, 9. Dysgerminomas are important beyond mere incidence because they affect women of reproductive age (i.e., <30 y) In fact, dysgerminomas make up two thirds of all malignant ovarian neoplasm's in women younger than 20 years6,7,8,10.In this study, the patient presented at the age of 46 years. Once diagnosed, dysgerminomas respond highly to the prescribed treatments, rescuing them from mortality4. Most patients have signs and symptoms related to the abdominal mass. In patients with associated gonadoblastoma, an underlying abnormality in gonadal development may dominate the clinical picture and associated with calcification2, 6, 13, Up to 95% of patients with dysgerminoma have elevated serum levels of lactic dehydrogenase (usually isoenzymes 1 and 2) at presentation, with the level varying with the size and stage of the tumor 4, 13. Rarely patients may have paraneoplastic hypercalcemia.occasionally serum inhibin levels may be elevated. Approximately 65% of dysgerminomas are stage Ia, at higher stages, the contralateral ovary, pelvic, and para-aortic lymph nodes and/or the peritoneum are typically involved4,6,8. The 5-year survival rate approaches 100% for stage I tumors12. Because of the sensitivity of dysgerminomas to chemotherapy and radiation therapy, the 5-year survival rate

for patients with higher stage disease or recurrent tumors is currently more than 80%. Most tumors recur within the first 2 years, but occasionally recurrence is late, even beyond 10 years.

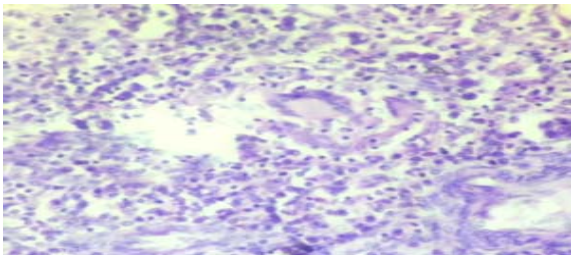

Figure -9. Low power view of cervix showing giant cells resembling the langhans cells admixed with dysgerminoma cells.(10X)

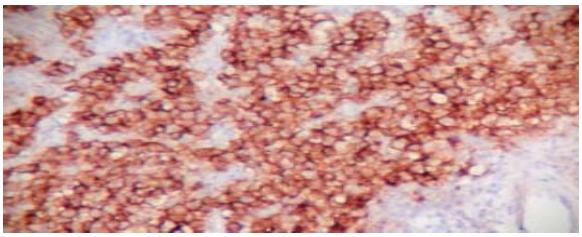

Figure -10. Immunohistochemistry with Cyto keratin showing focal cytoplasmic positivity in ovary(40x).

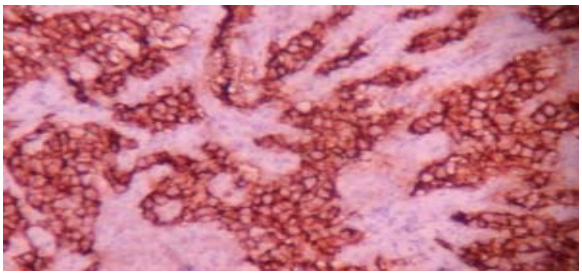

Figure-11. Immunohistochemistry with CD-117 in ovary showing diffuse strong membrane staining(40X).

GROSS FEATURES: (2,3,6,7,13) Dysgerminomas are generally solid tumors with a median diameter of 15 cm. In this study the average diameter is 5 cm. The serosal surface is smooth or bosselated. The sectioned surfaces are typically soft, fleshy, lobulated, and cream-colored, gray, pink, or tan . Areas of cystic degeneration, necrosis, and hemorrhage are occasionally present and should be sampled to exclude other types of malignant germ cell tumor, which may require different therapy. Calcification suggests the possibility of an underlying gonadoblastoma. The tumor is grossly bilateral in approximately 10% of the cases which is seen in our case too.

MICROSCOPICALLY(4,5,6,7,8) the cells resembles primordial germ cells with predominantly diffuse or insular arrangements, but other patterns like cords, solid tubules, trabeculae, small clusters, or single cells are also seen. The uniform, rounded tumor cells have clear to eosinophilic cytoplasm (which is almost always glycogen-rich), discrete cell membranes, and a central, large, rounded or flattened nucleus that contains one or a few prominent nucleoli. Mitotic figures are usually numerous. The characteristic stroma consists of thin septa to broad fibrous bands infiltrated by mature lymphocytes, sometimes with lymphoid follicle formation. Sarcoid-like granulomas are present in 20% of cases and occasionally are confluent, sometimes partly obscuring the underlying tumor **Immunohistochemistry:(5,7,8,11 13)** shows positive for placental alkaline phosphatase (PLAP), CD- 117, OCT3/4,SALL4 (nuclear transcription factor) and variably for cytokeratin. CD-30 negative Useful tumor markers for dysgerminomas are

- bHCG
- AFP
- Lactate dehydrogenase (LDH)
- Cancer antigen 125 (CA125)

These markers also can be used for postoperative follow-up care or for tracking the success of adjuvant therapy.

MOLECULAR AND GENETICS: Isochromosome 12 (i(12p)) is seen in dysgerminomas, as it is seen in seminomas of the testis8.

DIFFERENTIAL DIAGNOSIS2,5,6,8:

The chief disorders in the differential diagnosis of dysgerminoma are

- Yolk sac tumor
- Embryonal carcinoma
- Sertoli cell tumor
- Lymphoma/leukemia
- Large-cell lymphoma typically occurs bilaterally;

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