

Classic Dysgerminoma: A Case Report

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Abstract: *Ovarian dysgerminoma is rare malignant ovarian tumor seen in reproductive age group women. It accounts for 1 -2 % of all primary ovarian neoplasms. Clinically presents with abdominopelvic mass with abdominal pain with or without menstrual disturbances with elevated tumor markers of LDH & Beta HCG. Majority of cases will be diagnosed in Stage I and it has an excellent prognosis as it is highly sensitive to chemotherapy. Dysgerminomas are the only germ cell tumor with a significant rate of ovarian involvement bilaterally about 15 to 20 percent. This case report demonstrates that both the clinical and sonographic findings are unique to this rare type of ovarian malignancy.*

Keywords: Dysgerminoma, Germ cell tumor, Tumor markers

1. Introduction

Ovarian dysgerminoma is rare malignant ovarian tumor seen in reproductive age group women. It accounts for 1 -2 % of all primary ovarian neoplasms¹. The clinical presentation of abdominal mass, pelvic pain and specific tumor marker elevation clinches the differential diagnosis of Ovarian dysgerminoma. The majority of dysgerminomas are diagnosed early, Stage IA, and respond well to conservative fertility-sparing treatment of a unilateral salpingo-oophorectomy²

2. Case Report

A 35 yrs old multiparous lady presented with abdominal distension for past one month, associated with mild abdominal pain, H/O loss of weight, H/O loss of appetite, H/O nausea. Her physical examination was remarkable with abdomino pelvic mass of size 18 * 15 cm, firm to hard mass, mobile, irregular surface, not tender occupying supra pubic region, right iliac fossa, umbilicus region was felt.

Further evaluation of the mass with 18F FDG PET CECT revealed FDG avid bulky right adnexal soft tissue mass noted involving the right side of pelvis and lower abdomen,

measuring 185 * 114 * 174 mm, SUV max 10.8 (Figure 1) Mild ascites noted. No FDG avid lesions noted in the uterus and left adnexa. FDG avid following nodes noted: Mesenteric nodes largest 13 mm, SUV max 4.4, Retroperitoneal nodes, largest 9 mm, SUV 2.5. Periportal and celiac axis nodes, largest 10 mm, SUV 3.8. In addition to elevated serum Lactate dehydrogenase (LDH) 463 U/L, Beta HCG 76.89 mIU/mL, was suggestive of malignancy. Alpha Fetoprotein was normal 1.36 ng/ml, Serum CEA 1.07 ng/ml.

Based on patients age, clinical findings, biochemical analysis and radiological findings the most probable diagnosis was Ovarian dysgerminoma. Then Surgical staging laparotomy was done and proceeded with Right Ovariectomy With TAH with Left Salpingo-oophorectomy with omentectomy with pelvic lymph node dissection. Histological diagnosis Right ovarian tumor – Dysgerminoma, pT1a pn0. Immuno Histochemistry Marker CD 117 Positive, D 240 Positive. Peritoneal fluid cytology negative for malignancy. Lymph nodes shown reactive lymphadenitis features without any evidence of malignancy. As tumor size was big, surgical oncologist suggested chemotherapy in spite of Stage I A disease and our patient received 3 Cycles of bleomycin, etoposide, and cisplatin. Patient is in regular follow up.

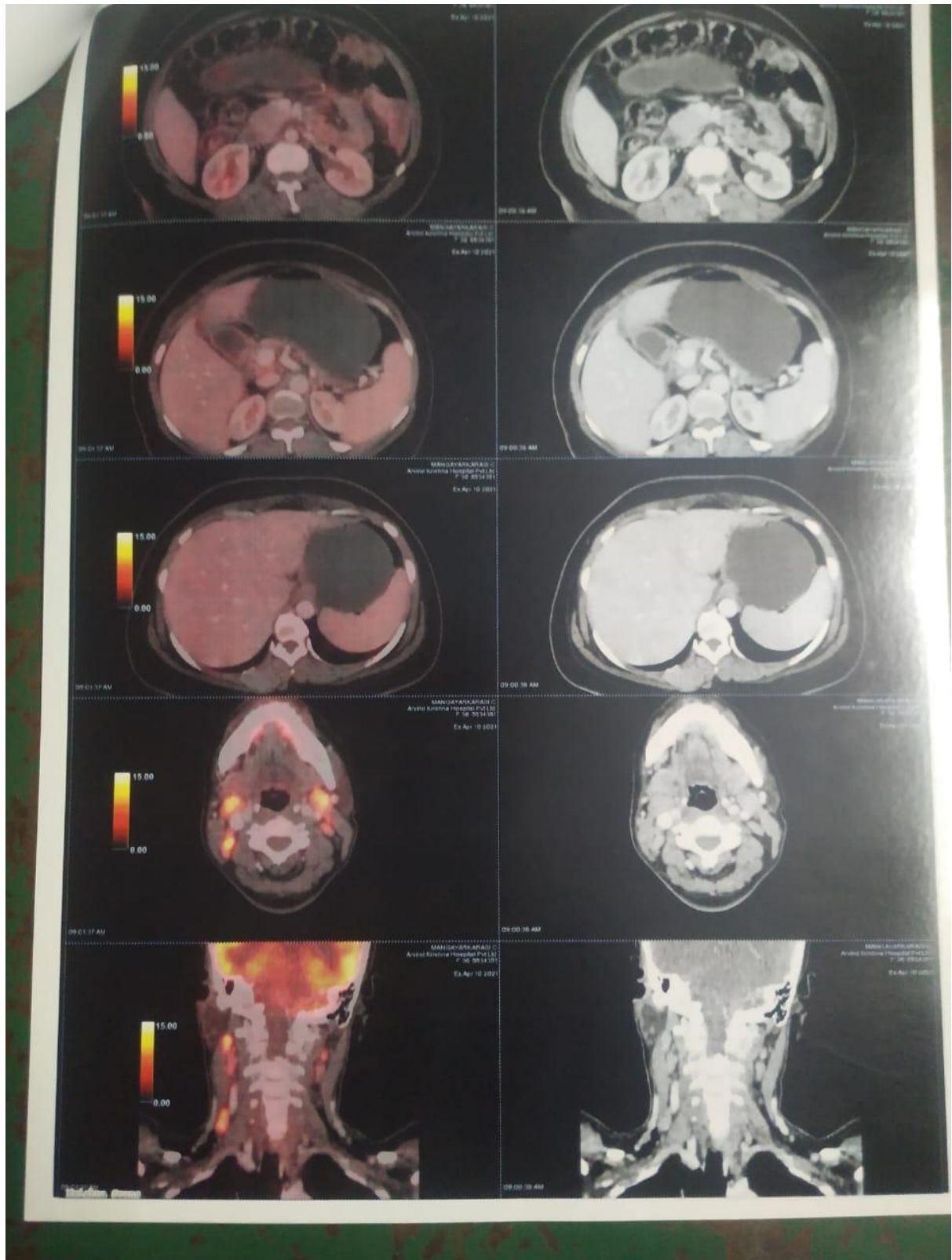


Figure 1: 18F FDG PET CECT revealed FDG avid bulky right adnexal soft tissue mass

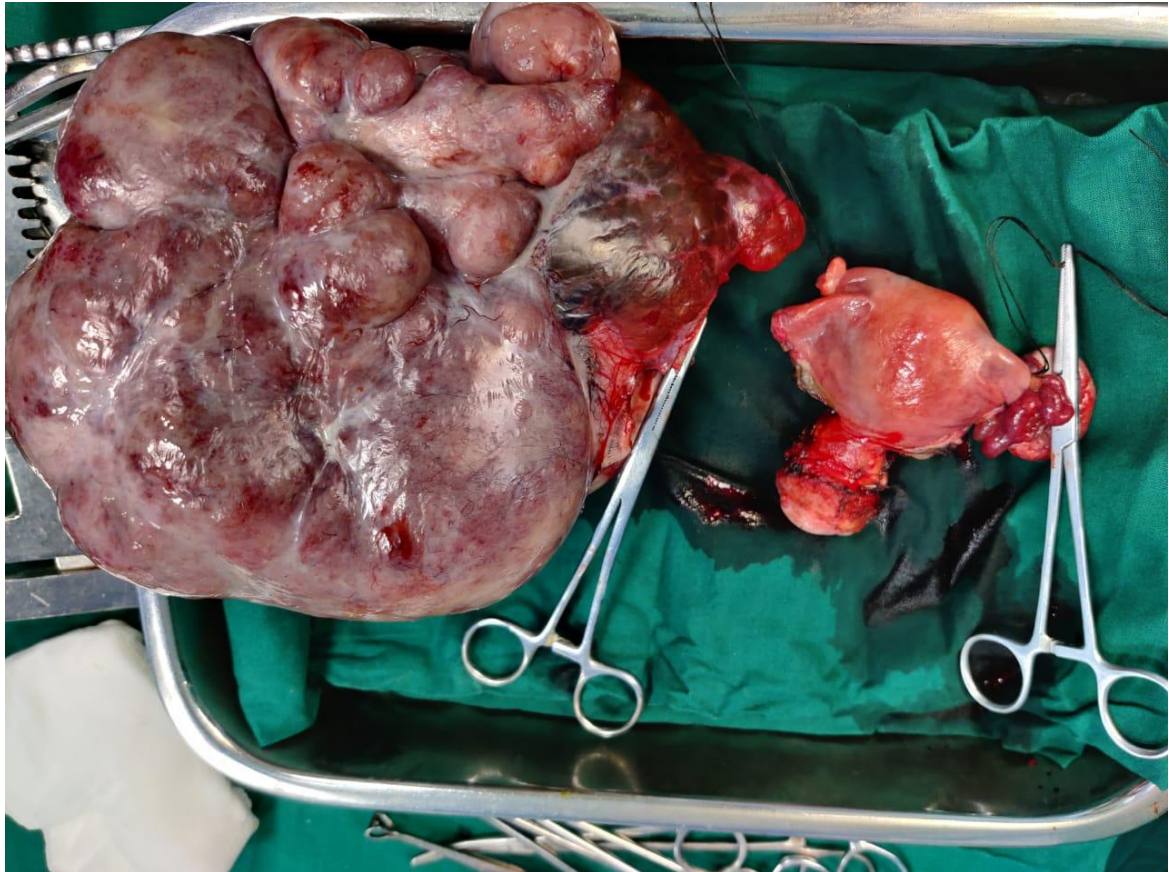


Figure 2: Specimen Right ovariectomy with TAH with left salphingo ophorectomy

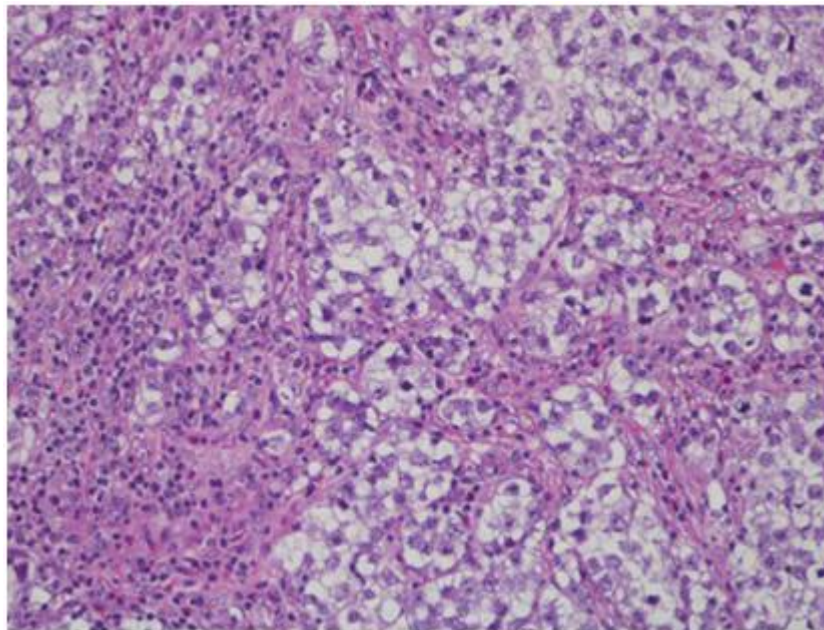


Figure 3: Histopathology Dysgerminoma, Large hyperchromatic nuclei with abundant eosinophilic cytoplasm

3. Discussion

Germ cell tumors are the tumours which arise from the ovary's germinal elements and its make upto one third of all ovarian neoplasms. Malignant germ cell tumors comprises about 2 to 3 percent of all malignant ovarian cancers³. Three-fourths of cases usually arise in the young adults and adolescents, and this same age group accounts for 33% of all ovarian malignancies⁴

Three features distinguish malignant germ cell tumors from epithelial ovarian cancers. First, individuals will typically present at a younger age, usually in teens or early 20s but our patient belongs to 3 rd decade. Second, most of them have stage I disease at diagnosis. Third, prognosis is excellent, even or those with advanced disease—due to excellent tumor chemosensitivity. Fertility sparing surgery is the primary treatment for women who seek future pregnancy, although most of them will require postoperative chemotherapy. As our patient completed her family and as

there is an risk of 15% chance of occult clinical occurrence or recurrence in other ovary, our patient opted for pan surgery and willing to have hormonal replacement therapy in future for hormonal support.

Subacute abdominal pain is the presenting symptom in 85 percent of patients and reflects rapid growth as occurred in our patient of a large, unilateral tumor undergoing capsular distention, hemorrhage, or necrosis. Individuals typically seek care within 1 month of the onset of abdominal complaints, although some may note subtle waxing and waning of symptoms or more than a year.

Diagnostic Procedures

Surgical resection is required for definitive tissue diagnosis, Staging and treatment. The surgeon should request a frozen section analysis for confirmation of diagnosis, but discrepancies between frozen section interpretations and the final paraffin histology can occur⁵. And also, specific immunostaining is often required to resolve equivocal cases⁶.

Histogenesis

Primitive germ cells will migrate from the wall of the yolk sac to the gonadal ridge. Hence most germ cell tumors arise in the gonad. Very Rarely, these tumors may develop primarily in extragonadal sites such as the central nervous system, mediastinum, or retroperitoneum⁷. Dysgerminomas are primitive neoplasms and they do not have the potential for further differentiation. Dysgerminomas are the only germ cell tumor with a significant rate of ovarian involvement bilaterally about 15 to 20 percent. And also patients with bilateral lesions will have grossly obvious disease, but cancer in the remainder will only be detected microscopically. 5% of women may have elevated serum hCG levels due to intermingled syncytiotrophoblast. Similarly, serum lactate dehydrogenase (LDH) and the isoenzymes LDH-1 and LDH-2 may also be useful in monitoring individuals or disease recurrence⁸.

Dysgerminomas will have a variable gross appearance, but in general, usually they are solid, pink to tan to cream-colored lobulated masses.

Microscopically, a monotonous proliferation of large, round, polyhedral clear cells which are rich in cytoplasmic glycogen and contains uniform central nuclei with one or a few prominent nucleoli. The tumor cells will closely resemble the primordial germ cells of the embryo and they are histologically identical to seminoma of the testis. The standard treatment of dysgerminoma involves fertility-sparing surgery with unilateral salpingo-oophorectomy (USO). In some situations, ovarian cystectomy may be considered⁹. Surgical staging is generally extrapolated from epithelial ovarian cancer but lymphadenectomy is highly important. Of the malignant germ cell tumors, dysgerminoma has the highest rate of nodal metastases, approximately 25 to 30 percent¹⁰. Preservation of the contralateral ovary leads to recurrent dysgerminoma in 5 to 10 percent of retained gonads during the next 2 years. This finding thought to reflect the high rate of clinically occult disease in other ovary rather than true recurrence. Indeed, at least 75 percent of recurrences develop within the first year

of diagnosis¹¹. Other frequent recurrence sites are within the peritoneal cavity or retroperitoneal lymph nodes. In spite of this significant incidence of recurrent disease, a conservative surgical approach do not adversely affect long-term survival because of cancer's sensitivity to chemotherapy¹². Dysgerminomas carries the best prognosis of all malignant ovarian germ cell tumor variants. 2/3rd are stage I at diagnosis, and the 5-year disease-specific survival rate approximates 99%. Even with advanced diseases have high survival rates following chemotherapy. For example, with stage II-IV disease may have a greater than 98-percent survival rate with platinum-based agents¹³. Despite the fact that all dysgerminomas are malignant, they have excellent prognosis after a simple salpingo-oophorectomy—up to 96% cure rate of a unilateral tumor without capsular invasion or spread. Because of its excellent response to chemotherapy, those that have extended beyond the ovary can often be cured, with overall survival of greater than 80%.¹⁴ In management of dysgerminoma, surgery is not only therapeutic but also required for diagnosis and staging, with scope of procedure dependent on intraoperative findings and patient's desire to retain fertility or avoid exogenous estrogen.¹⁵⁻¹⁷ Patients with completely resected stage IA disease usually receive 3 cycles of bleomycin, etoposide, and cisplatin while higher stages with stage III C disease—receive 4 cycles.

4. Conclusion

Ovarian dysgerminomas are included in the differential diagnosis for reproductive age female who presents with non-acute lower quadrant pain, palpable pelvic mass, and elevated β -HCG and LDH. Supporting radiographic findings include a solid, heterogeneous, lobulated adnexal mass with low resistance internal blood flow. The majority of tumors are Stage IA at the time of diagnosis and it can be conservatively treated with a unilateral salpingo-oophorectomy to preserve fertility. This case study demonstrates the unique characteristics of this rare type of malignant ovarian germ cell tumor, including age of presentation, symptoms, elevated lab values, and sonographic characteristics.

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