



Case Report

Huge ovarian embryonal cell carcinoma in an adolescent girl: A case report

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ABSTRACT

Pure ovarian embryonal carcinoma is a rare type of germ cell tumor, usually presenting in adolescent age group with only 30 odd cases reported in literature. They are mostly reported as one of the component of mixed malignant ovarian germ cell tumor. Tumor markers such as Human Chorionic Gonadotropin (HCG), and alpha-fetoprotein (AFP) and Lactate Dehydrogenase (LDH) also contribute to the diagnosis, prognosis and follow - up of germ cell tumors. Here, we report a rare case of a pure ovarian embryonal cell carcinoma in an adolescent girl with atypically raised LDH levels whereas the other markers such as AFP, HCG, CA-125 were normal.

A 13 year old adolescent girl presented with pain abdomen and abdominal mass. Ultrasound and CT scanning showed a huge multi-cystic septate abdomino -pelvic mass involving the right ovary. We performed fertility sparing staging laparotomy with right salpingo-oophorectomy, saving the uterus and the left ovary. Presently, patient has completed 3 cycles of adjuvant chemotherapy (BEP regimen).

Conclusion: Pure embryonal carcinoma, being an extremely rare tumor, the treatment outcomes and long-term disease-free survival is unknown, often data is extrapolated from its testicular counterpart, however treatment is standardized with fertility sparing USO and combination chemotherapy as the patients are very young. Recurrences occur mostly in the first 2 years following primary treatment. Also, secondary malignancies due to chemo-toxicity is a concern, as patients are very young, warranting a long and intensive surveillance.

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1. Introduction

Pure embryonal carcinoma of the ovary is an extremely rare tumor, with scarce reporting in the literature.¹ Commonly they are encountered as a component in mixed malignant germ cell tumor. Germ cell tumors account for 20% to 25% of all benign and malignant ovarian neoplasms across all age group, but only 3% of these are malignant.¹ Interestingly, almost 70% of childhood ovarian tumors (i.e., in the first two decades of life), are germ cell tumors, and a staggering one-third of them are malignant ovarian germ cell tumors (MOGCT), thereby, warranting a thorough and rigorous investigation to detect malignancy at the earliest.^{1,2} Embryonal carcinoma present in adolescent

age group.³ Tumor markers such as Human Chorionic Gonadotropin (HCG), and alpha-fetoprotein (AFP) and Lactate Dehydrogenase (LDH) also contribute to the diagnosis, prognosis and follow up of germ cell tumors.² We report a case of a pure ovarian embryonal cell carcinoma in an adolescent girl with atypically raised LDH levels.

2. Case Report

A 13 year old girl presented with complaints of right sided lower abdomen pain, mild to moderate intensity, on and off since 6 months, non-radiating, aggravating with physical exertion, not completely relieved with pain killers. She also had complaints of abdominal distension since last 5 months. No h/o dyspepsia, bloating sensation. No h/o fever, burning micturition, altered bladder/bowel symptoms. No

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h/o weight loss, evening rise of temperature, chronic cough.

Patient attained menarche at 11 years of age and had normal regular cycles. Her last menstrual period being 10/02/2019. On examination patient had moderate degree of pallor, no icterus, pedal edema or lymphadenopathy. Her vitals were normal. Systemic examination pertaining to central nervous system, cardiovascular and respiratory systems were unremarkable. Abdomen examination revealed an abdomino-pelvic mass of size 15*10 cm arising from the pelvis and reaching about 3 cm above umbilicus, firm in consistency, mobility present side to side, non tender, margins not well defined and lower pole could not be reached.

Her Hemoglobin was 8.3g%, total leukocyte counts were 78,00 /ml, platelets were 652T/ml, Packed cell volume was 27.9%. Tumor markers were done which showed increased LDH- 4435 U/L. Serum bet a HCG, AFP and CA-125 were normal. Her Liver function test, Renal function test, Thyroid profile and serology were unremarkable. Ultrasound and CT scan showed multicystic septate abdomino-pelvic mass of size 8.3* 16* 16 cms with maintained fat planes. The right ovary was not seen separately from the lesion. Uterus and left ovary were normal. No evidence of abdominal lymphadenopathy seen. No loculated/free intraperitoneal or pelvic fluid seen. Patient underwent exploratory laparotomy with systematic exploration of abdomen. Peritoneal washings were taken from paracolic gutters, and pouch of Douglas for cytology. Per-operatively an 18*15*6 cm mass of mixed consistency arising from the right ovary with intact capsule was present. The pedicle was enlarged to 10cm length and had torsion of 1 and a half turns (Figure 1 ab). Right salpingo-oophorectomy was done without surgical spill. The left tube and uterus were normal. The left ovary was mildly enlarged and a small nodule of 0.5cm present over the left ovary, which was excised and sent for histopathology examination along with the right tube and right ovarian tumor. Frozen section revealed malignant germ cell tumor. Omental biopsy along with appendectomy was done. No enlarged pelvic or para-aortic lymph node were encountered.

On gross pathologic examination, sections through the ovary exuded 200 ml of clear fluid. Solid and cystic areas present with haemorrhage alongwith grey-white multiple nodules. Microscopic examination revealed round to polygonal cells with increased nucleo-cytoplasmic ratio, vesicular nucleus, with marked anisokaryosis, arranged in sheets, trabeculae & cords (Figure 2). Abnormal mitotic figures and areas of necrosis were seen, these findings consistent with embryonal cell carcinoma. Peritoneal washings, omental biopsy and contralateral ovarian nodule were free of tumor.

The patient had a usual post-operative course and received 3 cycles of chemotherapy with BEP (Bleomycin, etoposide and cisplatin) regimen every 3 weeks. Her LDH

levels have drastically reduced to 161 U/L presently.



Fig. 1: a,b: Showing intra-operative huge right ovarian tumor with pedicle elongation and torsion.

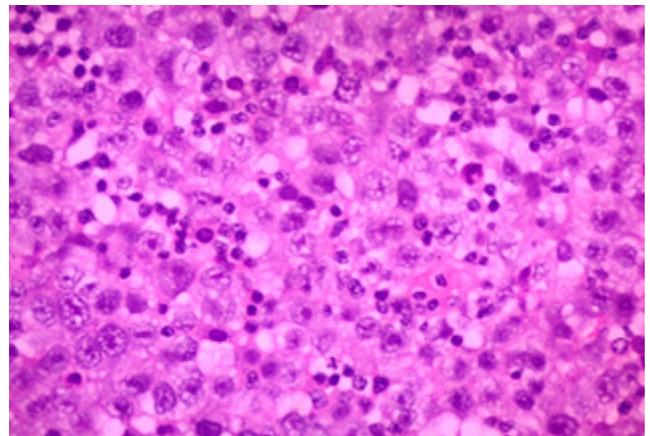


Fig. 2: Showing photomicrograph consistent with Embryonal carcinoma of ovary

3. Discussion

Germ cell tumors originate from the ovary's germinal elements.¹ They are histologically classified as dysgerminoma, embryonal carcinoma, choriocarcinoma, endodermal sinus tumor and mixed germ cell tumor and others.^{1,2} Pure ovarian embryonal carcinoma is quite rare, with about 30 odd cases reported in literature.³ The patients are usually young, the mean age being 14 years.³ They usually secrete AFP & HCG more so if they are a component of mixed germ cell tumor,⁴ nevertheless in the pure variety these markers, peculiarly, are absent, as seen in our case. Embryonal carcinomas are unilateral tumors, size averaging more than 15 cm.⁵ They are largely solid-cystic tumors containing mucoid material. Sectioning shows a variegated appearance with widespread areas of hemorrhage and necrosis. Easy identification of these rare tumors can be done by noting solid disorganised sheets of large anaplastic cells, distinct papillary structures and gland-like spaces.⁵

Clinically, majority of patients with germ cell tumors present with abdominal pain, distension or a pelvic mass.

Approximately 10 percent of them will present with an acute abdomen, secondary to rupture, intraperitoneal haemorrhage, or torsion of the ovarian mass.⁶ Both HCG and alpha-fetoprotein are secreted by some germ cell malignancies, and are useful in the diagnosis and in monitoring the response to treatment.^{5,6} Placental alkaline phosphatase (PLAP) and LDH are produced by upto 95% of dysgerminomas. But in our case, LDH level was raised to 4435 U/L, yet no component of dysgerminoma was evident. Ultrasonography and CT is helpful in delineating the size and complexity of these tumors, and also lymph node status can be known.

Treatment is standard fertility sparing unilateral salpingo-oophorectomy (USO), in young patients.^{7,8} Adjuvant chemotherapy (ChT) with combination of BEP (Bleomycin, etoposide, cisplatin) achieves excellent remission, with overall disease-free survival rates of greater than 95%.^{7,8} Lowe JH et al in 2000⁹ and Zanetta et al in 2001,¹⁰ reported normal gonadal function and fertility in patients of MOGCT treated with surgery and adjuvant combination chemotherapy. In 2013, Solheim et al. reported dose-dependent risk of gonadal dysfunction in females.¹¹ Also, patients receiving less than 4 cycles or no adjuvant treatment had a higher cumulative probability of conception.¹¹ The infertility rate in women attempting pregnancy after treatment for GCTs is 5%–10%,¹² which is similar to that seen in age-matched general population.^{13,14} Our plan of care included staging laparotomy with USO (right-sided) with appendectomy and omental biopsy with adjuvant 3 cycles of BEP chemotherapy.

3.1. Follow-up

Careful clinical, radiological, and serological surveillance is to be done every 2 months for first year, 3 monthly for second year, every 6 months during the third year and then annually upto 10 years.¹⁴ Measurement of tumor markers is done at each visit, imaging is done if indicated by history & examination.¹⁴ Regular surveillance is safe with an excellent chance of cure with platinum-based salvage chemotherapy for the 20–40% that do relapse.¹⁵

3.2. Follow-Up

Surveillance of patients following surgery and chemotherapy for germ cell tumours should be intensive in the initial follow-up period. Most recurrences of embryonal cell tumours, together with other ovarian germ cell malignancies, will occur in the first 12 months following treatment. This risk of recurrence reduces after the first two years

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None.

5. Conflict of interest

None.

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