Case Report

Mixed Germ Cell Ovarian Tumour in an 8-year-old Child — A Case Report

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In paediatric population occurrence of ovarian cancer is rare. In this study, an 8-year-old girl was diagnosed mixed malignant ovarian Germ Cell Tumuor (GCT), with predominantly Embryonal Carcinoma component (95%) and focal dysgerminoma component (5%). The patient presented with dull aching pain abdomen and mass in abdomen. On examination mass occupied whole of abdomen, hard in consistency, mobile and lower border per vaginally occupying right fornix. CECT abdomen was done which showed a well-defined solid cystic mass measuring 24.1x 18.7x 14.9 cm mass arising from the right adnexa causing suggestive of malignancy. Uterus, right ovary and rest of the peritoneal cavity was normal. CECT chest was normal. Tumour markers S. Beta-hCG was 27,601.44 mIU/mL, S. LDH was 1735 IU/mL, S. AFP was >400 ng/ mL and S.CA-125 was 114.5IU/mL. After multidisciplinary tumuor board discussion patient was planned for staging ovarian laparotomy (fertility preserving surgery). Intraoperatively ascites was present, right ovary was enlarged measuring about 25x15 cm occupying whole of abdomen, adherent to small bowel, fundus of uterus and right fallopian tube. Left ovary, Fallopian tube and Uterus was normal. Right pelvic and paraaortic lymph nodes was enlarged. Liver, bilateral diaphragm and rest of the peritoneal cavity normal. Procedure done was excision of right ovarian mass with right salpingectomy, bilateral pelvic lymph node dissection, retroperitoneal lymph node dissection, greater omentectomy and peritoneal biopsies. On histopathology right ovary was reported as poorly differentiated neoplasm. In retroperitoneal lymph nodes 1 out 15 lymph nodes showed tumour deposits measuring 0.2cm. Right fallopian tube, bilateral pelvic lymph nodes, greater omentum, peritoneal biopsies and ascitic fluid were free of tumour. On immunohistochemistry (IHC), right ovary tissue was positive for SALL4 and PLAP (germ cell tumour marker). CD30 (Embryonal carcinoma marker) was diffusely positive and CD117 (dysgerminoma marker) was focally positive. It was negative for Beta-hCG (choriocarcinoma marker), AFP (yolk sac tumour marker), EMA (epithelial marker), Calretinin (sex cord stromal tumour marker) and Inhibin (sex cord stromal tumuor marker). From above findings, diagnosis of FIGO stage III A1 (ii) (T1cN1aM0) mixed malignant GCT was made. Patient was advised adjuvant chemotherapy with Bleomycin, Etoposide and Cisplatin (BEP).

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Key words **: Mixed Germ Cell Tumuor, Ovary, Child.**

In paediatric population ovarian neoplasms are rarely seen. Annual incidence of ovarian cancer in paediatric population is about 2.2 cases per 1 lakh girls. 27% of all ovarian tumours in females less than 16 years are malignant[1]. Ovarian cancer represents 1.1% of all childhood malignancies[2]. In contrast to adults in whom epithelial carcinoma of ovary are common, whereas in paediatric population malignant Germ Tumours (GCTs)of ovary are more common[3]. The overall prognosis for patients with malignant Germ Cell Tumour is excellent[3]. Though understanding about etiology and pathogenesis of epithelial ovarian cancer has improved recently but understanding of etiopathogenesis of malignant ovarian GCTs has not improved[4]. Reporting a case of mixed malignant GCT diagnosed in an 8-year-old girl.

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Editor's Comment :

- Ovarian cancers are rare in paediatric age groups. Multidisciplinary team is required to treat the patients.
- Staging ovarian surgery is the standard of care.
- Fertility preservation surgery should be done whenever possible.
- Neoadjuvant chemotherapy should be used to decrease the tumour burden to increase the chance of fertility preservation.

Case Report

8-year-old girl with no significant family history presented with dull aching pain abdomen and mass in abdomen. On examination mass occupied whole of abdomen, hard in consistency, mobile and lower border per vaginally occupying right fornix. CECT abdomen was done which showed a well-defined solid cystic mass measuring 24.1x 18.7x 14.9 cm mass arising from the right adnexa causing right reflex hydrouteronephrosis suggestive of malignancy. Uterus, right ovary and rest of the peritoneal cavity was normal. CECT chest was normal. Tumour markers S. Beta-hCG was 27,601.44 mIU/mL, S. LDH was 1735 IU/mL, S. AFP was >400 ng/

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Discussion

In children and adolescents, GCTs are the most common variety of ovarian tumours histologically and they are mostly benign[6,7]. In India most common type of GCT is dysgerminoma followed by teratoma[8]. Incidence of mixed malignant Germ Cell Tumour is extremely rare. In one study done by Khanna, *et al* most common combination of subtypes were Yolk Sac tumour and immature teratoma. Mature and immature teratoma combination was second most common followed by combination of Yolk Sac tumuor and embryonal carcinoma being third most common[9].

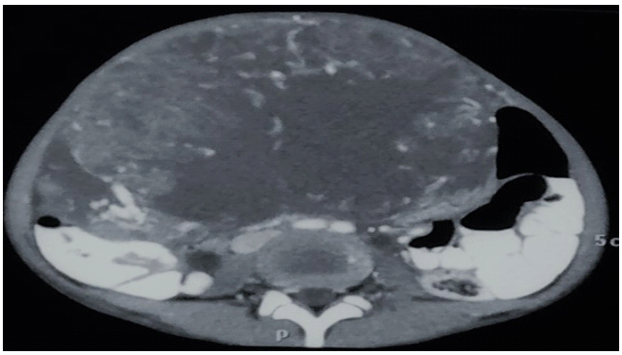

Fig 1 — CECT of abdomen and pelvis tumour arising from right ovary occupying whole of abdomen with ascites.

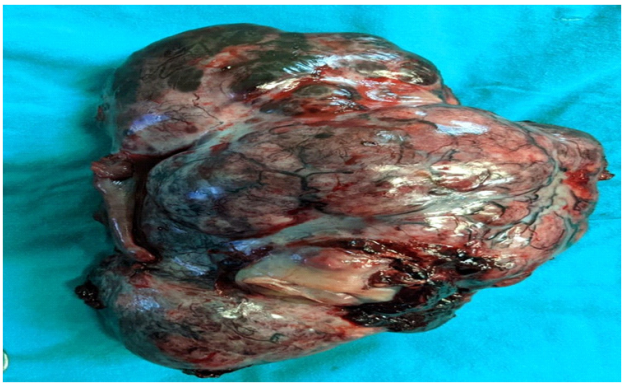

Fig 2 — Showing postoperative excised specimen of right ovary.

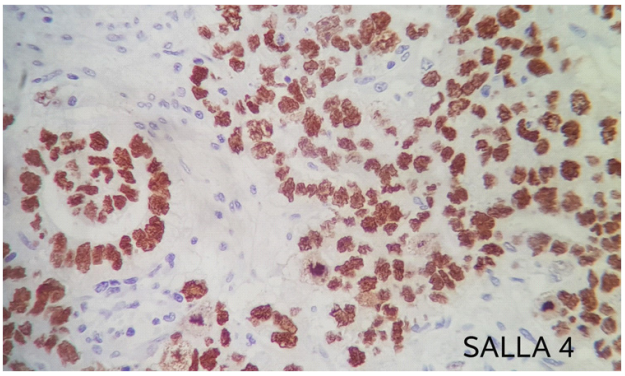

Fig 3 — Showing IHC on right ovary tumour was positive for SALL-4 (germ cell tumour marker).

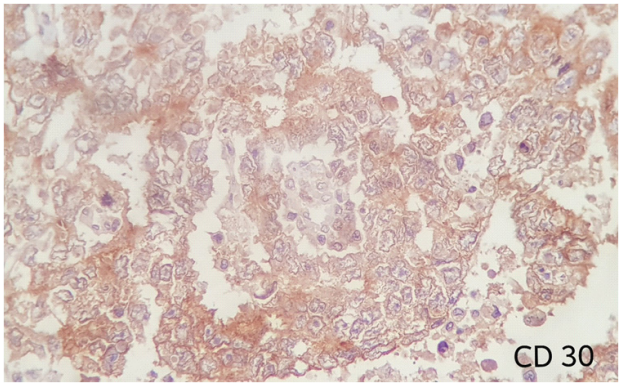

Fig 4 — Showing IHC on right ovary tumour was diffusely positive for CD 30 (Embryonal carcinoma marker).

World Health Organisation (WHO) classifies ovarian GCTs into many histological subtypes. Subtypes are : (1) Dysgerminoma, (2) Yolk Sac tumours, (3) Embryonal carcinoma, (4) Polyembryoma, (5) Choriocarcinoma, (7) Teratomas and (8) Mixed GCTs[10]. Dysgerminoma is most common followed by Yolk Sac tumuor also known endodermal sinus tumour[10]. In our patient, on histopathological examination and IHC revealed a combination two types tumours hence term as mixed GCT. 95% of tumour stained with IHC marker CD30 (Embryonal carcinoma marker) and 5% of tumour stained with CD117 (dysgerminoma marker). Most paediatric

patients with ovarian GCTs present between age 20-30 years[11]. Patients generally remain asymptomatic till ovarian mass grows to a large size causing abdomen pain adjacent organ compression. Less common presentations are vaginal bleeding, constipation and amenorrhoea. Sometimes they present as complication secondary to ovarian mass such as rupture of themass, infraction or torsion[1]. One-third of GCTs have extra-gonadal origin such as vagina, mediastinum, pineal gland, cervix, endometrium and sacrococcygeal area[12-14].

Surgery is main treatment of ovarian GCT. Fertility preservation surgery should be done whenever possible ie, preservation of uterus and opposite side ovary. When in doubt, opposite ovary biopsy should be taken before excision. Staging ovarian laparotomy is should be done to stage the disease[15]. Principles of staging surgery is defined by Children Oncology Group (1) Intact ovarian removal without rupture of the tumuor capsule. A salpingectomy must be performed if the fallopian tube is adherent. (2) Examination of the contralateral ovary with biopsy if a suspicious aspect is seen. (3) Inspection of the peritoneum, the liver and the omentum and resection of any abnormal tissue. (4) Inspection of aorto-caval and iliac lymph nodes and biopsy of suspicious ones. 5. Sampling of ascitic fluid for cytological examination. If ascites is absent, a washing is required[16,17]. Adjuvant chemotherapy should be used for tumours of FIGO any stage embryonal carcinoma and yolk sac tumour, stage II-IV dysgerminoma, Stage I with grade 2 or 3 and above immature teratoma[18]. BEP regimen is treatment of choice for treating ovarian GCTs[19]. Whenever extensive is disease is noted, neoadjuvant chemotherapy followed by surgery can be considered.In a study done by Rudaitis, *et al* showed that use neoadjuvant chemotherapy can significantly decrease the tumour size which minimizes the extent of surgery and thus helps in preserving fertility[20]. Overall prognosis of patients with ovarian GCT is excellent[3].

Conclusion

Ovarian tumours are rare in paediatric age group specially mixed ovarian Germ Cell Tumours. Multidisciplinary team is required to treat the patients. Coordination between paediatricians, surgical oncologist, medical oncologists, radiologists and pathologists is important for diagnosis and treatment. Surgery (staging ovarian laparotomy) is the standard of care. Fertility preservation surgery should be done whenever possible. Neoadjuvant chemotherapy should be used to decrease the tumour burden to enhance fertility preservation. Adjuvant chemotherapy should be used for tumours of FIGO any stage embryonal carcinoma and Yolk Sac tumour, stage II-IV dysgerminoma, Stage I with grade 2 or 3 and above immature teratoma.

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