MALIGNANT MIXED GERM CELL TUMOUR OF THE OVARY IN A 10-YEAR-OLD GIRL

Prabhakar S Gawandi[1], Chandracant Jadhave [2], Magala .A .Shinde [3], Vidya. R. Tirankar[4].

1. Assistant professor, Department of OBGY, DR V.M.M.C Solapur Maharashtra
2. Assistant professor, Department of OBGY, DR V.M.M.C Solapur Maharashtra
3. Professor and Head, Department of OBGY, DR V.M.M.C Solapur Maharashtra
4. Associate Professor, Department of OBGY, DR V.M.M.C Solapur Maharashtra

CORRESPONDING AUTHOR:
Dr Prabhakar S. Gawandi,
Apartment Flat No 4, Near Remand Home,
Karbse Bungalow North Sader Bazaar,
Maharashtra Solapur 413003.
E-mail: dr.prabhakar.gawandi@gmail.com

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CASE REPORT: 10 year prepubertal girl came with complain of lump in abdomen, pain in abdomen, intermittent retention of urine and constipation since 1-2 months
Menstrual history – not attained menarche
Past history –not significant
Family history –not significant

ON EXAMINATION: General condition fair, thin built, no e/o any lymphadenopathy
Per abdomen examination
INSPECTION --uniform lower abdominal distension mainly up to xiphisternum.
No other obvious finding
PALPATION – a single smooth surface cystic mass arising from pelvis extending up to lower border of xiphisternum, with irregular surface, 34=36weeks size pregnant uterus ,lower border not reached .
No any Organomegaly no evidence of any free fluid
PER SPECULUM AND PER VAGINAL EXAMINATION –Not done considering age of patient
Provisional diagnosis of germ cell tumour was kept.
INVESTIGATIONS-
Hb 7.7gm%, CBC- TLC 8500/cumm, platelet count -4.63lakh BSL 90gm%
BUL 70mg%, Serum creatine- 0.8 mg%, Na^+ -138, K^+ -3.7, SGPT- 21, SGOT -29, Bilirubin- 0.8
TUMOUR MARKERS
CA 125 -18.40U/ml
B-HCG-21600.00 mIU/ml
Alpha fetoprotein -1420 ng/ml

CHEST X-RAY: within normal limit

USG ABDOMEN AND PELVIS: E/o large, well defined heterogeneous solid mass lesion of size approximately 16.8cm ×15cm ×8.5 cm with area of cystic degeneration within it seen mass almost occupying the whole abdomen and pelvis, displacing the bowel coils. Both kidneys show mild hydronephrosis. However exact origin of mass could not be commented.
Patient receives one pint whole blood preoperatively. Patient was posted for exploratory laparotomy with provisional diagnosis of germ cell tumour.

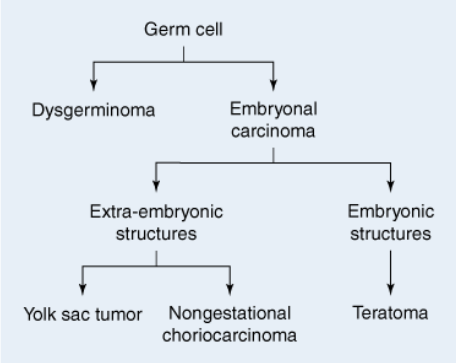
INTRAOPREATIVE FINDING: E/o ascitis, peritoneal fluid taken for cytological examination
Mass of approximately 20cm× 15cm× 10cm with irregular surface, capsule was breached in some areas. Mass was seen to be arising from the right ovary. Left sided ovary normal. E/o bilateral par aortic lymphadenopathy was present. Infracolic omentectomy was done. Mass was removed and send for frozen section. Report came as germ cell tumour. Post operative period uneventful.

HISTOPATHOLOGICAL REPORT: Gross examination Specimen consist of ovarian mass measuring 22 cm× 16cm × 12cm. External surface is bosselated. Cut surface shows a tumour with variegated appearances and bulging from parenchyma, tumour shows solid and soft areas with gelatinous material, area of haemorrhage and necrosis are also noted On microscopic examination this patient had mixed germ cell tumours with components embryonal carcinoma dysgerminoma and of yolk sac tumour. Omental biopsy shows reactive mesothelial hyperplasia and infiltrations by tumour cell with similar morphology Peritoneal seedling shows nodules composed of tumour cell with morphology similar to embryonal carcinoma.

Postoperatively patient received chemotherapy cycles of vincristine, dactinomycin, and cyclophosphamide (VAC).No complication during chemotherapy.

DISCUSSION: Histogenesis Primitive germ cells migrate from the wall of the yolk sac to the gonadal ridge. As a result, most germ cell tumors arise in the gonad. Rarely, these tumors may develop primarily in extragonadal sites such as the central nervous system, mediastinum, or retroperitoneum (Hsu, 2002)[1]. Ovarian germ cell tumors have a variable pattern of differentiation. Dysgerminomas are primitive neoplasms that do not have the potential for further differentiation. Embryonal carcinomas are composed of multipotential cells that are capable of further differentiation. This lesion is the precursor of several other types of extraembryonic (yolk sac tumor, choriocarcinoma) or embryonic (teratoma) germ cell tumors. The process of differentiation is dynamic, and the resulting neoplasms may be composed of different elements showing various stages of development (Teilum, 1965).[2]

In 1973, the World Health Organization classified germ cell tumours as Dysgerminoma, Endodermal sinus tumour, embryonal carcinoma, Polyembryoma, Choriocarcinoma, Teratomas (Immature, Mature, and Monodermal), Mixed and Gonadoblastoma. [3] This initiative represented a major advance in terms of standardization of nomenclature and histological criteria. Malignant germ cell tumours comprise less than 5 percent of all ovarian neoplasms [4]



The average age of presentation of patients with germ cell tumours is 13.8 years (range, 4 to 27 years). The presentation includes abdominal mass, abdominal pain, and fever. Serum AFP and HCG levels may be elevated. The average tumour diameter at presentation is 15.5 cm.Germ cell tumours demonstrate variable internal consistency, ranging from completely solid to largely cystic. In children and adolescents, approximately one-third of ovarian germ cell tumours are malignant, whereas the vast majority is benign in adults.

Dysgerminoma is the most common germ cell tumour, accounting for 50% of all germ cell tumour cases. The yolk sac tumour (also known as endodermal sinus tumour) is the second most common germ cell tumour, accounting for 20% of all cases, and is common in girls and young adults with an average age of 19 years. Less common germ cell tumours are embryonal carcinoma, immature teratoma, choriocarcinoma, polyembryomas, and mixed germ cell tumours.

Embryonal carcinomas are germ cell tumors formed by primitive cells that resemble those of very early embryonic development. They are considered to be the least differentiated type of germ cell tumor. Only about 4 per cent of malignant ovarian germ cell tumours.[5]They often is combined with other forms of germ cell tumors, most commonly yolk sac tumors. Embryonal Carcinomas occur primarily in children and young adults. These tumors can produce alpha-fetoprotein or *human* chorionic gonadotropin; the latter may be associated with precocious puberty and abnormal uterine bleeding. Embryonal carcinomas are highly malignant tumors that usually have spread extensively within the abdominal cavity by the time of presentation. They metastasize early and do so primarily through the lymphatic system.

Endodermal sinus tumour 2 most common tumour form of malignant germ cell tumour these neoplasm are highly malignant they metastasis early and invade the surrounding structure α fetoprotein level are often increase in this group of patient they are characterized by extremely rapidly Growth and extensively intra abdominal spread.

The most common component of a mixed malignancy was dysgerminoma, which occurred in 80%, followed by EST in 70%, immature teratoma in 53%, choriocarcinoma in 20%, and embryonal carcinoma in 16%. The most frequent combination was a dysgerminoma and an EST. The mixed lesions may secrete AFP, HCG, or both or neither of these markers, depending on the components. Microscopically a dysgerminoma component is present in 80 percent, endodermal sinus tumour in 70 percent, immature teratoma in 53 percent, choriocarcinoma in 20 percent and embryonal carcinoma in 16 percent. A mixture of dysgerminoma and endodermal sinus tumour is the most common combination, accounting for one-third of the cases.[6]

Surgery is the initial treatment for the majority of patients with malignant germ cell tumour of the ovary. Procedures include unilateral oophorectomy, bilateral salpingo-oophorectomy, and intra-abdominal tumour debulking, with the goal of removing as much gross tumour as possible while preserving fertility. All patients except those with FIGO/AJCC stage I require neoadjuvant or postoperative platinum-based combination chemotherapy. Cisplatin, bleomycin, and etoposide (BEP) and vincristine, dactinomycin, and cyclophosphamide (VAC) are commonly used regimens

CONCLUSION: Mixed malignant germ cell tumour of the ovary is a highly aggressive neoplasm that can present as disseminated disease at initial diagnosis. A high index of suspicion is needed and early intervention for any adolescent girl. Definitive diagnosis requires histological confirmation.

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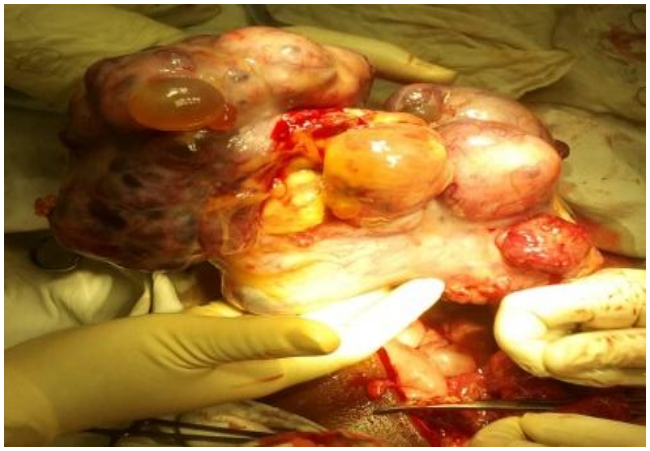

Fig 1: Intropertative finding.

Fig 2: Cut Section of the Tumour

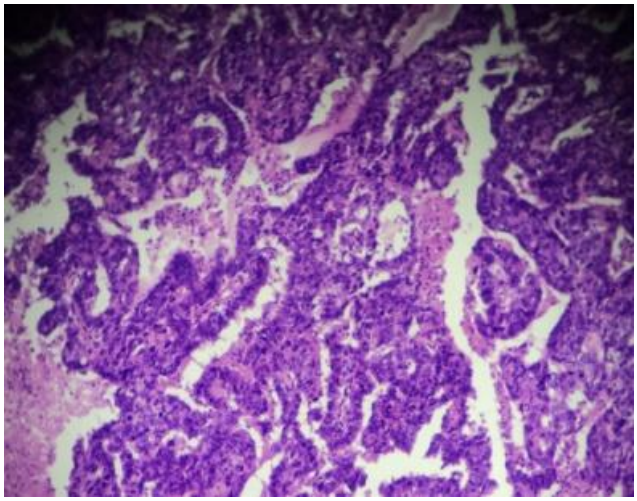

Fig 3: Microscopic picture of embryonal carcinoma

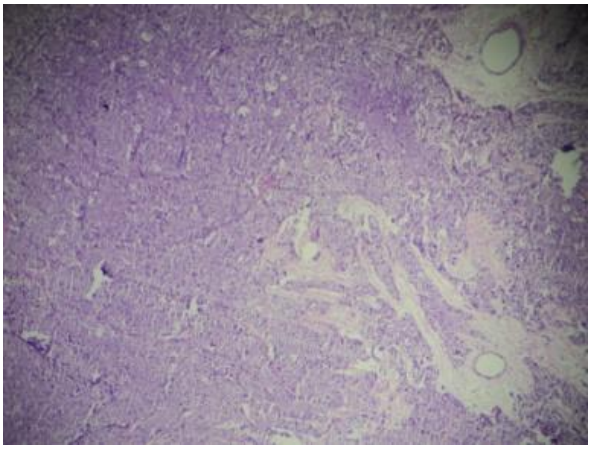

Fig 4: Microscopic picture of dysgerminoma

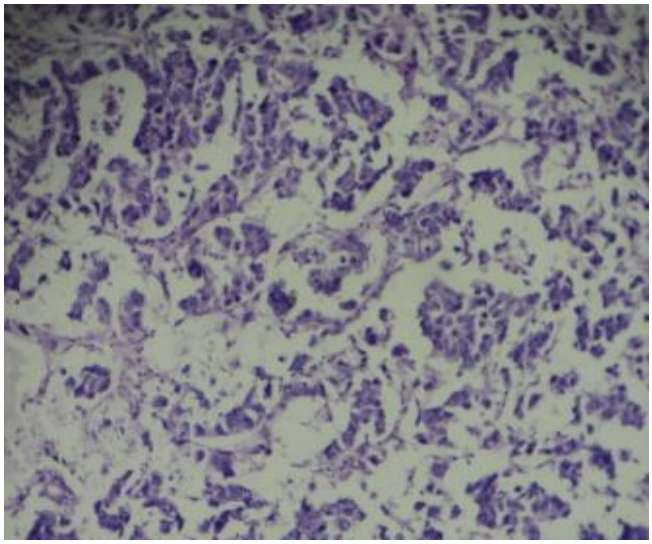

Fig 5: Microscopic picture of yolk sac tumour