# A CLINICOPATHOLOGICAL STUDY OF OVARIAN TUMOURS IN CHILDREN

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## SUMMARY

**Ovarian tumours in childhood were studied with a view to analyse the problems they post in diagnosis and management. They accounted for 6% of all ovarian tumours. The tumours were malignant in 92% of the cases and as a group germ cell tumours were the most predominant (72% of cases). Laparotomy was followed by varying extent of surgery. Postoperative radiotherapy and chemotherapy were given. However, 6 patients expired within a year. When first seen disease was in an advanced stage in as many as 22 cases. This precluded us from resorting to conservative surgery followed by adequate chemotherapy/radiotherapy which is the ideal treatment of ovarian tumours in childhood.**

### *Introduction*

Ovarian neoplasms in children are rare, notoriously lethal when malignant and there is controversy regarding their management. The rarity of these ovarian tumours does not allow the practitioner to become familiar with the specific problems that are inherent in the clinical and pathologic features of these neoplasms. The management of malignant ovarian tumours in children differs from that in the adult because of the desirability of maintaining the patient's reproductive and menstral functions. We have reviewed 26 cases of ovarian tumours in children below 17 years with a view to analyse the clinical presentation, the difficulties in diagnosis and the problems encountered in their management.

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### *Material and Methods*

All cases of ovarian tumours encountered in children (17 years or less) admitted to the Jipmer Hospital from January 1981 through August 1988 were considered for the study. All the patients received referral care at our hospital and were treated in the Departments of Gynaecology, Pediatric Surgery and Radiotherapy. A systematic search of the records was carried out. The laparotomy findings were recorded and frozen section study guided the extent of surgery. The tumours were then sent to the Department of Pathology for further study. The histological classification was based on that proposed

by World Health Organisation (WHO) and a similar one used by Scully (1979).

### *Observations*

There were 513 cases of ovarian tumours during the study period of which 26 were encountered in girls below the age of 17 years. Thus ovarian tumours in children accounted for 6% of all ovarian tumours. The symptoms with which the cases presented are shown in Table I. Abdominal mass was the most frequent symptom (80%) followed by abdominal pain (72% cases).

**TABLE - I**
**SYMPTOMS WITH WHICH THE CHILDREN PRESENTED**

| Symptoms | No. of patients | % |
|---|---|---|
| Abdominal mass | 20 | 80 |
| Abdominal pain | 18 | 72 |
| Abdominal distension | 6 | 24 |
| Loss of weight | 4 | 16 |
| Fever | 3 | 12 |

Physical examination revealed Mullerian duct agenesis, vestibular anus and bilateral iguinal hernia and mastocytosis in one case each. X-ray of the abdomen revealed calcified areas in two cases of mature teratoma. In 6 cases, IVP revealed pressure on ureter due to an extrinsic mass. Ultrasonogram revealed a solid tumour in 9 cases. Multilocular cystic mass was diagnosed in 11 cases.

The histological types of tumours encountered are shown in Table II. As a group, the germ cell tumours accounted for 92% (24) of the tumours. We came across 6 cases of mature teratoma; they accounted for 25% of germ cell tumours. The details are shown in Table III.

**TABLE - II**
**HISTOPATHOLOGY OF THE TUMOURS**

| Types of tumours | No. of cases |
|---|---|
| Mature teratoma | 6 |
| Immature teratoma | 3 |
| Teratocarcinoma | 1 |
| Dysgerminoma | 6 |
| Endodermal sinus tumours | 5 |
| Embryonal carcinoma | 1 |
| MIXED TUMOURS | |
| Endodermal sinus tumour + teratoma | 1 |
| Dysgerminoma + endodermal sinus tumour | 1 |
| EPITHELIAL TUMOURS | |
| Papillary serous cystadenocarcinoma | 1 |
| Mucinous cystadenocarcinoma | 1 |

In this series, there were 3 cases of immature teratoma. The relevant information is depicted in Table III.

The only case of teratocarcinoma occurred in an 8 years old girl. At laparotomy she had extensive metastasis. Hence debulking was done and patient was started on Actinomycin-D and Cyclophosphamide.

Dysgerminoma occurred in 6 cases and accounted for 25% of all germ cell tumours. Table IV illustrates the age group, stage and management of these cases.

Endodermal sinus tumour was diagnosed in 5 cases. The youngest patients were in this group. The salient features are shown in Table IV. Laparotomy was followed by radiotherapy and chemotherapy with Cyclophosphamide, Methotrexate and 5-Fluorrouacil (CMF). Two patients expired within 6 months and other 3 were lost to follow up.

**TABLE - III**
**CLINICAL DETAILS OF THE TERATOMAS**

| Type of tumours | No. of cases | Age range (yrs) | Clinical Stage | Surgery performed | Chemotherapy and/or Radiotherapy |
|---|---|---|---|---|---|
| Mature teratoma | 6 | 11-17 | – | Unilateral ovariectomy | – |
| Immature teratoma | 3 | 8-12 | I (2) | Unilateral ovariectomy | Vincristine Actinomycin Cyclo-phosphamide |
| | | | III (1) | Unilateral ovariectomy, hysterectomy and resection of sigmoid. | |

**TABLE - IV**
**CLINICAL DETAILS OF DYSGERMINOMAS AND ENDODERMAL SINUS TUMOURS**

| Types of tumours | No. of cases | Age range (Yrs) | Clinical stage | Surgery performed | Chemotherapy and/or Radiotherapy |
|---|---|---|---|---|---|
| Dysgerminoma | 6 | 7-14 | IIa (1) III (4) IV (1) | Abdominal hysterectomy with bilateral salpingo-oophrectomy (3); Dubulking (3) | All had Radiotherapy Vincristine Actinomycin Cyclophos-phamide |
| Endodermal sinus tumour | 5 | 1 yr (2) 11-13 (2) | II (1) | Abdominal hysterectomy with bilateral salpingo-oophrectomy (2) Debulking (3) | Cyclo-phosphamide Methotrexate 5-Fluorochain |

Mixed germ cell tumours were noted in two cases. Endodermal sinus tumour was associated with teratoma in one case and with dysgerminoma in another. In the former ipsilateral ovariotomy was performed and the patient was put on postoperative chemotherapy (CMF). She did not turn up for the second course. The second patient was a 10 year old girl and belonged to stage IIIb. Abdominal hysterectomy with bilateral salpingo-oophrectomy was performed. She received postoperative radiotherapy, but did not come for chemotherapy. She was readmitted in a moribund state a year latter and she died.

Borderline mucinous cystadenocarcinoma (Stage I) was encountered in a 15 year old girl. Right-sided salpingoovariotomy was performed. Patient did not turn up for further follow up and therapy. Papillary serous cystadeno carcinoma (Stage III) was found in a 12 year old

girl; debulking was followed by 8 courses of CMF. Patient was alive for 3 years.

***Discussion***

Ovarian tumours account for approximately 1% of all malignant neoplasms in the age group of 0 to 17 years (Young and Miller, 1975). In the present series, ovarian tumours in this age group comprised 6% of all ovarian tumours as also recorded by Norris and Jensen (1972). Sawai and Sirsat (1973) studying ovarian tumours upto the age of 20 years at the Tata Memorial Hospital, Bombay recorded an incidence of 11.2%. In this age group 76% of the tumours were malignant. A range of 13.3 to 85% was recorded on reviewing the literature from 1965-75 (Breen and Maxson, 1977).

Abdominal mass and abdominal pain were the most common symptoms as also noted by Raney et al (1987). One of our patients with torsion of teratoma presented with acute abdomen. One has to be careful in not mistaking it for acute appendicitis (Breen and Maxson, 1977). The diagnosis of ovarian neoplasms in the pediatric patient is difficult primarily because of low incidence and a very low index of suspicion. An average delay of 3-4 months between the onset of clinical symptoms and confirmation of ovarian tumours has been reported (Lindfors, 1971). Many of our patients were treated at primary level for abdominal pain and fever for 3 to 6 months in some cases. When there was no response they were referred to the pediatricians and then either to Pediatric Surgeons or the Gynaecologists. This caused an undue delay in starting therapy.

Ultrasonogram is a valuable tool and should be employed liberally in children with history of abdominal pain, abdominal mass and an altered clinical profile due to hormonal effects. It was performed in 20 of our patients.

Germ cell tumours, as a group were encountered in 24 (92% of cases). In the series of Sawai and Sirsat (1973), 77.5% of the tumours below the age of 20 years orginated from the germ cell. Breen and Maxson (1977) reviewing 1002 ovarian tumours in series from 1963 to 1975, found that tumours of germ cell origin accounted for 67.2% of tumours. Lack and Goldstein (1984) reported an incidence of 71% and Raney et al (1987) an incidence of 77.7%.

Mature teratoma of ovary accounted for 25% of tumours in our study as compared to 38.6% (Breen and Maxson, 1977). In children, the recorded age span is 3 months to 19 years with an average age of 15 years. The age range in our study was 11 to 17 years. Malignant degeneration within a mature teratoma has not been recorded in children (Breen and Maxson, 1977). Therefore, ovarian cystectomy and ovariectomy is an adequate surgical procedure; the latter particularly in a situation of torsion or rupture.

Immature teratoma accounted for 12% of the tumours in our study. Breen and Maxson (1977) reported 17 cases in the age group of 4 to 20 years. In one case of Stage I, ipsilateral ovariectomy was followed by Vincristine, Actinomycin-D and Cyclophosphamide (VAC). Patient is alive for 1 year. The other two were lost to follow up. With conservative surgery and adjuvant VAC regimen 10 of 12 patients survived with a median of 43 months (Breen and Maxson, 1977). Gershenson et al, (1986) and Pipitt et al, (1988) made similar observations.

In this series, dysgerminoma accounted for 25% of all germ cell tumours. All our patients had postoperative radio-

therapy. Only one case with stage IIIa survived for 2 years. The prognosis is also influenced by the presence of other malignant germ cell component. Asadourain and Taylor (1969) reported admixture of germ cell tumours in 12 of 117 patients with dysgerminoma. Krepart et al (1978) followed up 5 cases with stage Ia disease. All were alive 3 to 10 years after treatment and 3 of these have had children. Newlands and Bagshawe (1987) and Gallion et al (1988) reviewing 7 cases of dysgerminoma have concluded that unilateral ovariectomy should be resorted to if the tumour is confined to one ovary.

Endodermal sinus tumour (EST) occurred in 7 cases. There was only one case of embryonal carcinoma (EC). No patient had any endocrine manifestation. In two patients EST occurred alongwith dysgerminoma and teratoma. Both EST and EC are biologically the most aggressive of all the germ cell tumours. Two of our patients died within 6 months. In the series of Breen and Maxson (1977), 11 of 14 cases proved to be fatal. However, the introduction of very effective cytotoxic drugs and the use of tumour markers has had a major impact in converting this often fatal disease into a curable one (Slayton, 1978; Sessa et al, 1987).

Of the malignant tumours, only 4 cases reported in the early stage of the disease and only 3 cases came for follow up as instructed, probably due to ignorance and poverty. Many succumbed to the advanced stage of malignancy. Thus in our society, mortality in children suffering from malignant ovarian tumours will continue to remain high unless the physician at first contact has a high index of suspicion. Also there should be centres for specialised care with availability to cytotoxic drugs and facilities for follow up.

### *References*


1. *Asadourain, Z.A. and Taylor, H.B.: Obstet. Gynec. 33:370, 1969.*
2. *Breen, L.J. and Maxson, W.S.: Clin. Obstet. Gynec. 20:607, 1977.*
3. *Gallion, H., Van Negell, H.R., Powell, D.R.: Am. J. Obstet. Gynec. 158:598, 1988.*
4. *Gershenson, D.M., Del Juneo, E.S., Sliva, E.G.: Obstet. Gynec. 67:624, 1986.*
5. *Gershenson, D.M., Copeland, L.J., Del Juneo, G.: Obstet. Gynec. 68:789, 1986.*

5A. *Krepart G., Smith F.P., Rutledge F. and Delclos L., Cancer 41, 908, 1978.*

6. *Lack, E.E. and Goldstein, D.P.: Current Problems in Obstetrics and Gynaecology, 1984. Year Book Medical Publishers.*
7. *Lindfors, C.: Ann. Chir. Gynec. Fenn, 177:1, 1971.*
8. *Norris, H. and Jensen, r.: Cancer 30:713, 1972.*

8A. *New Lands, E.S. and Bagshawe, K.D.: Recent Advances in Obstetrics & Gynecology 1987 Churchill Livingstone Medical Division, London.*

9. *Pippitt, C.H., Chair, J.M., Hames, T.B.: Gynec. Oncol. 31:268, 1988.*
10. *Raney, B.R., Sinclair, L., Uri, A.: Cancer, March 15:1214, 1987.*
11. *Sawai, M.M. and Sirsat, M.V.: Indian J. Cancer 10:302, 1973.*
12. *Sessa, C., Bonassi, C., Landoni, F.: Obstet. Gynec. 70:220, 1987.*
13. *Slayton, R.E., Hreshchyshyn, M.M., Silverberg, S.G. et al.: Cancer 42:390, 1978.*
14. *Young, J.L. and Miller, R.W.: J. Pediatr. 86:254, 1975.*