Original Article

# Histopathological spectrum of ovarian neoplasms in children

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## Abstract

**Objective:** To study the various ovarian tumours in the paediatric age group in a tertiary care hospital. **Methods:** The cases of paediatric ovarian tumours were retrieved from the archives of the Department of Pathology. All the clinical details and laboratory investigations were recorded. A detailed histopathological study was done and the ovarian tumours were classified according to the current WHO classification. **Results:** There were 7 cases of paediatric ovarian tumours in this study. Age of the patients ranged between 7 months and 18 years. Pain was the most common symptom, presence of an abdominal mass was also frequent. Out of the 7 cases, 2 were that of mature cystic teratoma, 3 were that of immature teratoma, 2 were that of mixed germ cell tumour (immature teratoma, dysgerminoma, embryonal carcinoma, polyembryoma and yolk sac tumour) **Conclusion:** All the ovarian tumours in our study were found to have originated from germ cells. Majority (71.4%) of these ovarian tumours were found to be malignant. Mixed germ cell tumours are not uncommon and thorough sampling is required to identify them.

**Keywords:** Ovarian, paediatric, germ cell, teratoma.

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## INTRODUCTION

Childhood ovarian masses are rare. They include neoplastic and non-neoplastic processes. Non-neoplastic conditions include follicular cysts, corpus luteal cysts, and endometriomas. Neoplastic processes include both benign tumours such as mature cystic teratomas as well as highly malignant tumours.[1] Up to 64% of the ovarian masses have been reported to be neoplastic, majority of which arise from germ cells. The importance of considering pat ient's age when evaluating ovarian pathology lies in the fact that, in adults the majority of ovarian tumours are epithelial and in children they form less than 20%.[2]

## MATERIAL AND METHODS

The cases of paediatric ovarian tumours were retrieved from the archives of the Department of Pathology. All the clinical details were recorded. A detailed histopathological study was done and the ovarian tumours were classified according to the current WHO classification.

## RESULTS

Out of all the ovarian tumours received in the Department of Pathology, 7 cases were seen in children, all of which were germ cell tumours. Age of the patients ranged between 7 months and 18 years. Pain was the most common symptom, presence of an abdominal mass was also frequent. Out of the 7 cases, 2 were that of mature cystic teratoma [Figure 1-4], 3 were that of immature teratoma [Figure 5], 2 were that of mixed germ cell tumour [Figure 6] (immature teratoma, dysgerminoma, embryonal carcinoma [Figure 7], polyembryoma and yolk sac tumour [Figure 8]).

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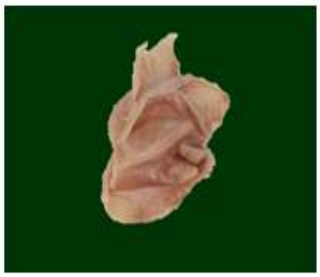
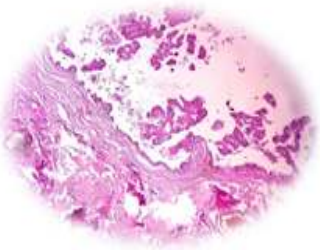


Figure 1

Figure 2



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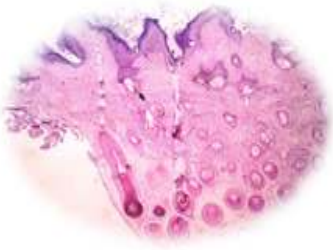

Figure 4

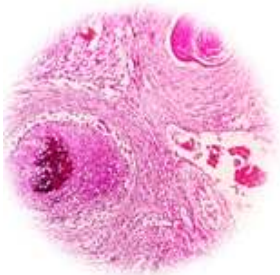

Figure 5

Figure 6

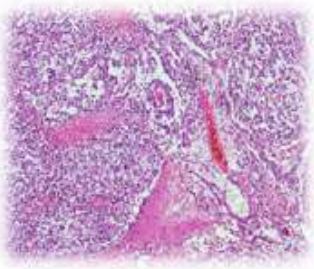

Figure 7

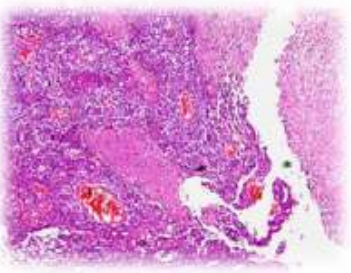

Figure 8

**Legend**

**Figure 1:** Benign Cystic Teratoma; **Figure 2:** Dermoid Cyst; **Figure 3:** Mature Cystic Teratoma Showing Choroid Plexus (H and E, x100); **Figure 4:** Mature Cystic Teratoma showing keratinized stratified squamous epithelium and skin appendageal structures (H and E, x400); **Figure 5:** Mature elements of teratoma (H and E, x400); **Figure 6:** Mixed germ cell tumour; **Figure 7:** Embryonal carcinoma component of mixed germ cell tumour (H and E, x400); **Figure 8:** Yolk sac component of mixed germ cell tumour (H and E, x400)

## DISCUSSION

Ovarian tumours are rare in the paediatric population and represent approximately 1.5% of childhood malignancies. Shih-Ming Chu *et al* showed that the symptoms are often insidious and commonly the tumour is quite large, by the time the diagnosis is finally determined and that at times, an ovarian tumour may be discovered, when a patient undergoes an operation for symptoms consistent with appendicitis. Such symptoms are more likely to be associated with a simple ovarian cyst, cystic teratoma torsion, or corpus luteal cyst rupture.[4] In our study 2 girls had this mode of presentation. Many studies have shown that in adult women, the vast majority of ovarian tumours are of epithelial origin. The germ cells are the most common cells of origin for ovarian neoplasms in the pediatric population.[2] This finding is consistent with our present study. As classified by the World Health Organization, the histologic subtypes of ovarian GCTs include teratoma (mature, immature, or monodermal), dysgerminoma, polyembryoma, embryonal carcinoma, yolk sac tumour, nongestational choriocarcinoma, and mixed GCT.[6]

TERATOMAS are GCTs composed of mature or immature tissues derived from more than one of the three primitive embryonic layers (ectoderm, mesoderm, and endoderm). They are the most common germ cell tumours observed in most published series. This subgroup of tumours may be further divided into mature teratomas, which are benign, or immature teratomas, which may be either malignant or benign. Most benign teratomas are composed of mature cells, but 20-25% also contain immature elements, most often neuroepithelium.[4]

Mature cystic teratoma (MCT), called dermoid cyst when the ectodermal elements predominate, is the most common ovarian tumour in children and adolescents, accounting for approximately 50% of all pediatric ovarian neoplasms. MCTs are bilateral in up to 10% of pediatric cases and in 10%–25% of adult cases.[6] Although MCTs typically are asymptomatic, patients may present with abdominal pain, a palpable mass, or acute onset of pain secondary to acute complications. Rarely, paraneoplastic syndromes, including autoimmune haemolytic anemia and immune-mediated limbic encephalitis, have been reported.[6] MCT is associated with various complications including torsion (3%–16%), rupture (1%–4%), infection (1%), and malignant transformation (1%–2%).[6] Grossly, an MCT is a well-circumscribed cystic mass with a smooth surface that is surrounded by a capsule of variable thickness and is filled with sebaceous material. Most MCTs are unilocular (88%), but some are multilocular. Histologically, the neoplasm often has a focal internal protuberance, known as a Rokitansky nodule, that may contain hair, bone, teeth, muscle, or cartilage, and the cyst

wall is lined by squamous, respiratory, or gastrointestinal epithelium.[6]

Immature teratoma, typically affects a younger age group; the younger the patient, the more likely that the teratoma will be the immature germ cell type. Immature teratoma frequently occurs between the ages of 10 and 20 years, with a median age of 17 years, and represents 10%–20% of all ovarian malignancies in patients younger than 20 years.[6] The neoplasm has a more aggressive behavior and a worse prognosis than does mature teratoma and thus has been treated as malignancy, although it is not truly malignant.[6] Immature teratoma is usually unilateral, large, and predominantly solid. Coexisting ipsilateral and contralateral MCTs occur in 26% and 10% of cases of immature teratoma, respectively. Histologically, immature teratoma contains a variable amount of primitive immature embryonal tissues derived from three germ cell layers admixed with mature tissues. The histologic grade is determined by the quantity of the immature neuroepithelial tissue.

Elevated serum AFP levels have been reported in 33%–65% of patients with immature teratoma.[6]

DYSGERMINOMA, The histologic profile of dysgerminoma is stereotypic and identical to that of testicular seminoma and midline germinomas.[7] It originates from undifferentiated germ cells that are similar to primordial germ cells. It is the most common malignant GCT in childhood and adolescence.[6] Large, clear, neoplastic, primitive-type germ cells are consistently associated with a variable cytotoxic Tcell lymphocytic response that helps to identify the tumour, even in cases with complex histology.[7] Grossly, the tumour is commonly well-encapsulated, lobulated, and solid, with a white to light tan color. Irregular necrosis, cystic change, or calcifications are seen in the tumour, and there may be focal hemorrhagic areas.[6] Histologically, the tumour is composed of monotonous polygonal tumour cells that aggregate in cords and clumps, with abundant cytoplasm and flattened central nuclei. The tumour cells are separated by fibrous or fibrovascular septa.[6] Rarely, microcysts or pseudoglandular spaces distort the usual architecture, creating differential diagnosis problems with the highly malignant small cell carcinoma of hypercalcemic type or even struma ovarii.[7] In our study, dysgerminoma was found to occur not in its pure form but as one of the components of a mixed germ cell tumour.

YOLK SAC TUMOUR, the new term primitive endodermal tumours, is a more apt definition of their complex, multifaceted histologic features, which comprise early endodermal differentiation into secondary yolk sac and primitive gut, and their derivatives, such as intestine, liver, and lung. The other terms like endodermal sinus tumours, are no longer accepted, as the structure it purports to represent is nonexistent in human embryogenesis. It is a rare malignant GCT and mostly occurs in the 2nd and 3rd decades of life, at an average age of 19 years.[7] Yolk sac tumour is usually aggressive, characterized by rapid growth and extensive spread to the abdomino-pelvic cavity. Hematogenous and peritoneal metastases are common. The tumour produces AFP, which can be used as a tumour marker at initial diagnosis and postoperative follow-up.[6] In our study yolk sac tumour was found to occur as one of the components of a case of mixed germ cell tumour.

EMBRYONAL CARCINOMA, is a rare and highly malignant tumour that accounts for approximately 3% of malignant GCTs. The tumour occurs primarily in children and adolescents, with a median age of 14 years. Unlike in other malignant GCTs, isosexual precocity or menstrual irregularity related to b-hCG secretion occurs in up to 60% of cases.[6] Histologically, embryonal carcinoma frequently appears mixed with other malignant germ cell types. Pure embryonal carcinoma is extremely rare. Grossly, the tumour manifests as a large, predominantly solid, variegated mass. Extensive areas of haemorrhage and necrosis are commonly seen, with cystic spaces containing mucoid material.[6] In our study embryonal carcinoma was seen as one of the components of a mixed germ cell tumour.

POLYEMBRYOMA, is a very rare ovarian tumour. It consists of numerous embryoid bodies which morphologically resemble normal embryos. It is usually found in association with other malignant germ cell components such as dysgerminoma, yolk sac tumourr, embryonal carcinoma, and mainly immature teratoma.[13]

## CONCLUSION

All the ovarian tumours in our study were found to have originated from germ cells. Majority (71.4%) of these ovarian tumours were found to be malignant. Mixed germ cell tumours are not uncommon and thorough sampling is required to identify them.

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