Original Research Paper

Volume - 13 | Issue - 04 | April - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Pathology

# CLINICOPATHOLOGICAL SPECTRUM OF INTRA-ABDOMINAL CYSTIC MASSES IN PEDIATRIC PATIENTS IN A TERTIARY HEALTHCARE CENTRE

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**ABSTRACT** **Introduction:** Cystic lesions in any organ system, particularly the abdomen, are extremely common in childhood, regardless of whether the underlying aetiology is developmental or congenital. The aim of the present study is to study the clinical and histopathological features of all intra-abdominal cystic masses in the pediatric patients. **Materials and Methods:** This is an observational study which was conducted in the Department of pathology at Rangaraya Medical College, Kakinada, Andhra Pradesh from November 2019 to October 2022. A total of 25 subjects (both male and females) were included in this study. **Results:** Choledochal cysts were the most common cause of intra-abdominal cystic masses in the pediatric population in this study. The spectrum of lesions are 16 choledochal cysts, 5 intestinal cysts and diverticula which included 1 gastric antral duplication cyst, 2 cases of Meckel's diverticula and 2 omphalomesenteric duct cysts. Rare cysts, namely, 1 case of splenic true epithelial cyst, 1 case of mesenchymal hamartoma of the liver were observed. 1 case each of hemorrhagic follicular cyst of the ovary and pancreatic solid pseudopapillary neoplasm were encountered. **Conclusion:** Treatment and prognosis of these cystic masses in children is dependent not only on the radiologic features, but also on the histopathological features.

**KEYWORDS :** Cysts, Histopathology, Pediatric

## INTRODUCTION

Intrabdominal cystic lesions, whether congenital or acquired are quite common in childhood. Majority of these cystic masses are incidental findings on prenatal ultrasonography, while the rest are found in symptomatic children, where imaging plays a major role in diagnosis. However histopathology of these intra-abdominal cystic masses is the gold standard for accurate diagnosis and it determines the management. The prognosis of these cystic masses in children is highly reliant not just on the radiologic features, but also on the histopathological features owing to the fact that the presence of metaplasia, dysplasia, divergent differentiation or an accompanying focus of malignant transformation completely alters the prognosis.

## MATERIAL AND METHODS

This observational study was conducted in the department of pathology at Government general hospital, Rangaraya Medical College, Kakinada after obtaining ethical committee approval. The study material included all the pediatric patients with cystic masses of abdomen received at the department of pathology from November 2019 to October 2022 (3 years). A total of 25 cases were enlisted for this study.

**Study Design:** Prospective observational study

**Sample Size:** 25

**Inclusion criteria:** Radiologically detected intra-abdominal masses in children ≤ 15 years.

**Exclusion criteria:** specimens which were found to be solid masses after grossing.

**Method of data collection:**

- Specimens received with biopsy requisition form (filled with patient particulars) are fixed in 10% formalin.
- Fixed specimens are then subjected to grossing (strictly according to Royal College of Pathology protocols), tissue processing and section cutting.
- Microscopic features of Hematoxylin and Eosin stained sections of paraffin blocks are studied.

## RESULTS

Out of 25 patients, 13 (52%) were lesser than or equal to 4 years of age, the youngest being 2 months old male child and the oldest patient was a 15 year old male child. The mean age of presentation was 5.3 years.

Out of 25 cases, 11 were males accounting to 44% and 14 were females accounting to the rest 56% cases. There was a male to female ratio of 1:1.2.

**Table 1 : Overall Age Distribution Of Cystic Masses In Pediatric Patients**

| Age Group | Number of cases ( n=25 ) |
|---|---|
| ≤ 4 years | 13 (52%) |
| 5-10 years | 9 (36%) |
| 10 – 15 years | 3 (12%) |

**Table 2 : Sex Distribution Of Cystic Masses In Pediatric Patients**

| Sex | Number of cases (n=25) |
|---|---|
| Male | 11 (44%) |
| Female | 14 (56%) |

The study sample included sixteen (16) choledochal cysts, out of which 12 were females and 4 were males. There were five (5) cases of intestinal cysts and diverticula which included single (1) gastric antral duplication cyst, two (2) cases of Meckel's diverticula and two (2) omphalomesenteric duct cysts. This study encountered rare cysts in abdominal organs, namely, one case of splenic true epithelial cyst and one case of mesenchymal hamartoma of the liver. One case of hemorrhagic follicular cyst of the ovary and one case of pancreatic solid pseudopapillary neoplasm were encountered.

The most common location of intra-abdominal cysts in the present study, was extrahepatic bile ducts with 11 out of 25 lesions were choledochal cysts, subclassified into types I, II, III, IVb accounting for 44%. The next most common site was liver and intrahepatic bile ducts with 6 out of 25 cases presenting at this location which included 5 choledochal cysts (Type IVa and V) and a single case of mesenchymal hamartoma. This was followed by intestines (16%), where 2 cases each of Meckel's diverticulum and omphalomesenteric duct cyst were identified. The stomach, spleen, pancreas and ovary were other locations of cystic masses accounting to 1 case per site and 4% of the patient population respectively.

**Table 3 : Location Wise Distribution Of Cystic Masses In Pediatric Patients**

| Site | Number of cases (n=25) |
|---|---|
| Liver and intrahepatic bile ducts | 6 (24%) |
| Extrahepatic bile ducts | 11 (44%) |
| Intestine (large and small) | 4 (16%) |
| Stomach | 1 (4%) |
| Spleen | 1 (4%) |
| Pancreas | 1 (4%) |
| Female genital tract | 1 (4%) |

The most common presenting symptom was pain abdomen in 8 (32%) out of 25 cases. Other presenting complaints were mass per abdomen accounting to 7 (28%) of 25 cases and jaundice accounting to 3 (12%) of 25 cases. Incidental detection of a cystic mass lesion on radiology in asymptomatic patients was seen in 7 (28%) of 25 cases.

**Table 4 : Presenting complaints of Cystic masses in pediatric patients**

| Symptoms | Number of cases (n=25) |
|---|---|
| Pain abdomen | 8 (32%) |
| Mass per abdomen | 7 (28%) |
| Jaundice | 3 (28%) |
| Radiological findings (mass lesion per abdomen) | 7(28%) |

## DISCUSSION

Choledochal cysts were the most common cause of intra-abdominal cystic masses in the pediatric population in this study.

### Choledochal Cysts:

A choledochal cyst is a congenital dilatation affecting a segment or even the entire CBD . It is more common in Asia than Western countries. The five clinical types according to Todani's classification are:

Type I - Solitary fusiform extrahepatic cyst
Type II - Extrahepatic supraduodenal diverticulum
Type III - Dilatation of intraduodenal CBD (choledochocele)
Type IVA - Multiple fusiform intrahepatic cysts
Type IVB - Multiple fusiform only extrahepatic
Type V - Multiple intrahepatic cysts only (Caroli disease)

Majority of choledochal cysts (70–90%) are of Type I. It is more prevalent in females than males. Intermittent discomfort, jaundice, and a mass per abdomen are the most common symptoms. It is the most frequent cause of obstructive jaundice in children.

Macroscopically, the fibrous cyst wall shows calcifications. The cyst, which has a maximum diameter of 15 cm, often contains 1-2L of fluid and wall thickness within 2-10mm.

Microscopically, the wall is composed of thick fibrous tissue with dispersed elastic and smooth muscle fibres. Infants have an intact columnar epithelium and inflammation. The lining is frequently discontinuos in older people, and the inflammation is more apparent. Often, scattered glands can be located. The lining is often duodenal in choledochocele (choledochal cyst involving the intraduodenal part of CBD). Choledochal cysts frequently harbour hyperplasia, metaplasia, or flat or tumoral intraepithelial neoplasia; carcinoma is thought to occur in 2–10% of cases, with a 20-fold increased chance of developing the condition compared to the general population. The wall of the cyst is most common site for carcinomas, though they can occur in gall bladder too.

### Gastric antral duplication cyst:

Foregut duplications form 35% of all gastrointestinal tract duplications, whereas stomach duplications are rare constituting only 7%. The larger curvature is more often involved in gastric duplications. Although respiratory epithelium can be seen, stomach or enteric-type epithelium typically lines gastric duplication cysts. There are frequently associated anomalies, e.g., duplications in other parts of GIT. The embryological origin of such cysts is unknown. Grossly, they are cystic or tubular masses.

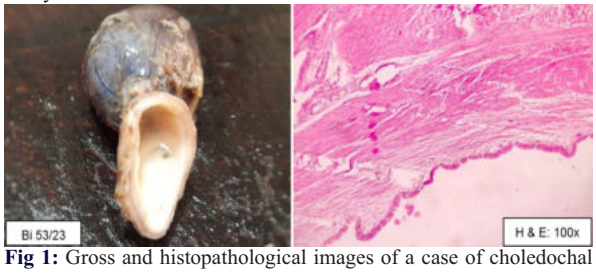


**Fig 1:** Gross and histopathological images of a case of choledochal cyst.

### Mesenchymal Hamartoma Of The Liver:

Rare benign hepatic mass lesions known as mesenchymal hamartomas consist of large, fluid-filled cysts and loose mesenchymal stroma. According to a research of more than 1,200 children hepatic neoplasms, they represent around 6% of all paediatric liver tumours, making them the second most prevalent benign paediatric tumour. Boys are more affected than girls. Children under the age of three are affected more than other age groups. Occurrence is more frequent in the first year of life.

While older patients and adults present with abdominal pain or jaundice, paediatric patients typically present with abdominal distension or an upper abdominal mass. Serum alpha-fetoprotein (AFP) levels can be moderately to significantly high, liver enzyme values are typically normal, hence a biopsy or resection helps to rule out an erroneous diagnosis of hepatoblastoma.

They are true neoplasms and not hamartomas, despite their name. A balanced translocation involving the same breakpoint on chromosome 19q13.4 has been described in multiple studies. Nevertheless, they have the potential to turn malignant and form embryonal sarcomas.

Macroscopically, they are solitary, unencapsulated, and well circumscribed. Around 75% of the tumours are located in the right lobe, range in size from 5 to 30 cm. More than 90% of the time, sliced sections show numerous cysts, ranging in size from a few millimetres to 14 cm. The tumours in younger newborns may be solid, but in older individuals, the lesion becomes progressively cystic and often fibrotic. The cysts contain clear/mucoid fluid but are not true cysts; instead, they are a consequence of cystic degeneration.

Microscopy reveals both mesenchymal and epithelial components, although the mesenchymal component predominates. Sections show sparse nests of bland hepatocytes and small collections of bile duct-like structures which are elongated and branched. The stroma is made up of primitive mesenchymal tissue which is composed of bland stellate and spindle cells with myofibroblastic features, embedded in an edematous or myxoid matrix with varying quantities of collagen.

Extramedullary hematopoiesis is frequent and found in the stroma next to small bile ducts or capillaries near the lesion's edge.

### Splenic epithelial cyst:

Splenic cysts are uncommon and occur in only 0.07% population. Either squamous epithelium or mesothelial type cells line them. True cysts are those that have an epithelial lining, whereas false cysts lack a cellular lining. The most frequent splenic cysts are squamous-lined epithelial cysts, which are hypothesised to develop from metaplasia in mesothelial cysts.

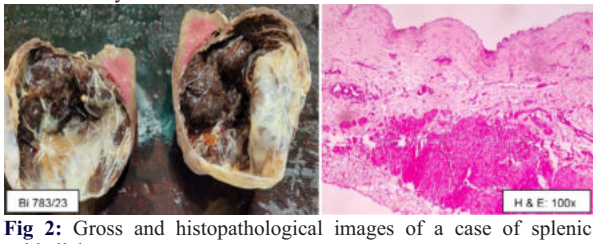


**Fig 2:** Gross and histopathological images of a case of splenic epithelial cyst.

### Hemorrhagic follicular cyst of ovary:

Now that sonography is a common practice, ovarian cysts in utero and in newborn are diagnosed more frequently. Cysts of ovary are most common reason for an abdominal mass in a fetus or newborn. Both ovaries are affected equally, however bilateral cysts are less frequent. In the first few months of life, the majority of tiny cysts involute and are of negligible clinical significance.

Large, untreated ovarian cysts complicate to hydrops, dystocia and pulmonary hypoplasia secondary to an abdominal space occupying lesion, intestinal obstruction, torsion of the pedicle resulting in infarction, hemorrhage into the cyst, or abdominal cavity and rupture of the cyst with peritonitis. Gross examination of the lesion reveals a thin-walled cyst with a smooth lining. Microscopically, the cyst wall consists of a upper layer of luteinized granulosa cells and a layer of luteinized theca cells underneath.

### Meckel's diverticulum and omphalomesenteric duct cyst:

Meckel's diverticulum occurs due to the failure of the omphalomesenteric (vitelline) duct to resorb by 8th week of life. It is twice as common in males than females. The "law of 3s" describes it as being present in up to 3% of live newborns, up to 3 inches (8 cm) in diameter, typically 3 feet (100 cm) proximal to the ileocecal valve, and

less than 3% symptomatic. Omphalomesenteric fistulas, sinuses, enteric cysts, fibrous bands connecting small bowel to the umbilicus, and intussusceptions at the umbilicus are all included under the umbrella term Meckel's diverticulum.

Vitelline cysts, also known as enterocysts, are generated when the intermediate portion of the omphalomesenteric duct undergoes incomplete stricture or apoptosis.

Mucosa of Meckel's diverticulum may contain acid-secreting gastric epithelium (30-50%) or ectopic pancreas.

**Solid pseudopapillary neoplasm of pancreas:**

These tumors of uncertain differentiation are more common in females, occurring around 30 years of age. Although they have been reported in all age groups, they tend to affect men more frequently in their youth or late adulthood. SPNs frequently develop cystic degeneration as they grow. Macroscopically, they look well-defined, hemorrhagic and yellow-brown in colour. The cysts are usually extremely asymmetrical and lined with shaggy detritus.

Microscopically, cyst walls consist of solid nests of cells surrounded by large number of small vessels. The tumour cells are not cohesive due to damaged intercellular adhesion molecules. A cuff of viable cells encircling each blood vessel gives these tumours their distinctive pseudopapillary appearance. There are no true intercellular luminal spaces.

## CONCLUSION

Because management strategies differ, careful whole histopathological examination of abdominal cystic masses in the paediatric population is critical in diagnosis. The prognosis of these cystic masses in children is dependent not only on radiologic features, but also on the histopathological features.

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