ORIGINAL RESEARCH PAPER

Paediatric

KEY WORDS: Fetus; Neoplasms; Congenital abnormalities; Antenatal anomalies; Pediatric cancer

RARE PRESENTATIONS OF NEONATAL ABDOMINAL LUMPS

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ABSTRACT

Even though the differentials of pediatric tumors are dependent on patient's age, etiology of tumors may vary as per cause such as congenital anomalies, antenatal tumours, trauma, organ enlargement and infections. Ultrasonography evaluation should be performed early during the work up which helps in quick and accurate identification of the mass, the organ of origin or association and about nature of mass whether it is cystic or solid. Contrast enhanced tomography scan with or without intravenous contrast may be required to confirm the diagnosis. Many of the abdominal masses are found by parents incidentally while bathing a child or by a physician during routine checkup. Improvement of imaging techniques has helped significantly to increase the incidence of fetal tumors detection during prenatal evaluation. The early detection of a fetal tumor and understanding of its imaging features are very important for fetal, maternal, and neonatal care. Herewith presenting case series of various presentations of lumps in abdomen in neonatal age with varying etiologies.

INTRODUCTION

True prevalence of all congenital tumors is difficult to predict as many cases of stillborn or aborted may not be reported although literature reported as the range of 1.7-13.5 per 100,000 live birth. Incidence of congenital tumours is higher as per recent studies probably due to improvement of imaging techniques and prenatal evaluation. The variety of possible differential diagnoses and the variable course during intrauterine and neonatal period create dilemma about diagnosis and management protocol of the fetal masses.

Many of pediatric tumors are presented as an incidental finding with poor prognosis if detected in early age, although there are some exceptions. **(1)**

Herewith presenting with our such experiences encountered with neonatal lumps in abdomen.

Case Report

Case 1 :-

A newborn female child has been referred immediately after birth with very gross distended tensed abdomen having dilated superficial veins. Antenatal sonography at 36 weeks of gestation showed fetal intra-abdominal mass solid in nature extending from pelvis to upper abdomen. Previous fetal scans were normal with no any additional fetal and maternal morbidities. Rest of fetal parameters were normal, so after planned normal delivery baby was evaluated immediately. Clinically there was very gross and distended abdomen with dilated abdominal veins. Baby passed meconium immediately after birth. Baby presented with persistent tachypnea due to compression effect on the diaphragm. Baby was admitted in neonatal intensive care unit. Hematological work up was normal. For further evaluation of intra abdominal mass, computed tomography scan with intravenous contrast was done showing huge solid intra-abdominal mass extending from pelvis to mid abdomen with compression effects on ureters on either sides and urinary bladder **(Fig.4)**. An emergency exploratory laparotomy was done on day 3 of life. On exploration there was a huge solid capsulated, friable mass arising from retroperitoneum extending from deep pelvis up to mid abdomen **(Fig.1,2,3)**. Due to excessive friability mass was excised in piece meal fashion and intra operative specimen was sent for frozen section. On histopathology report it was diagnosed to be extra renal wilms' tumour. Complete tumor mass is excised along with the capsule as much as possible. Intra abdominal drain was kept in situ and abdomen was closed in layers. Postoperative histopathology report confirmed same findings in favor of extra renal wilms' tumour **(Fig.5)**. Post-operative baby was kept under observation in intensive care unit. Baby showed gradual symptomatic improvement and was planned to shift to higher center for further chemotherapy. But till postoperative day 10 baby developed Gross abdominal distension same as before the exploration. On repeat sonography study of abdomen showed recurrent of mass of similar size as before the operation. It was really confusing presentation of rapid recurrent tumor. Due to noncompliance of parents for further treatment in view of chemotherapy and radiotherapy, baby could not be succumbed.

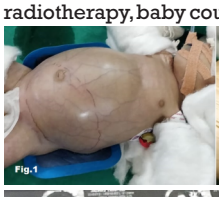
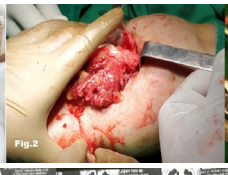


Fig.1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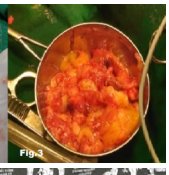
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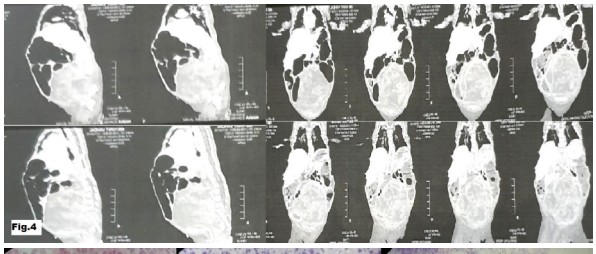
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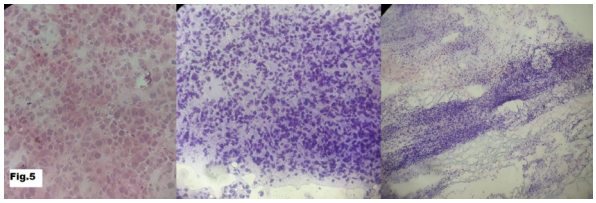
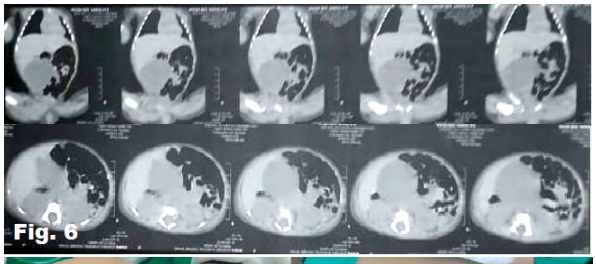


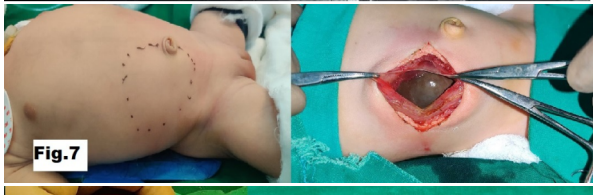
Fig.5

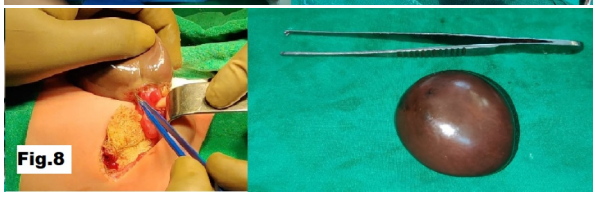
Case 2:-

A case of antenatal suspected fetal intra-abdominal cystic swelling detected at 22 weeks of gestation, got delivered at 36

weeks of gestation by caesarean section mode. On serial fetal sonographies done antenatally showed gradual increase in size of cystic swelling inside fetal abdomen with normal rest of the fetal parameters. Immediately after birth baby was shifted to intensive care unit. Along with other routine haematological evaluation, serum tumor marker values were sent CA19-9 [which were raised 43.43 (normal range 0-37 U/ml)] and For further evaluation of intra-abdominal mass, computed tomography scan was done showing huge solid intra-abdominal mass extending from pelvis to liver occupying whole of right side of abdomen with compression effects on right side ureter, inferior vena cava **(Fig.6) .** Exploratory laparotomy was performed on day 3 of life. Under anesthesia, solid mobile lump with well-defined margins was palpable in right of abdomen. On exploration there was well capsulated cystic swelling visible with dense adhesions on retroperitoneal side and with colon **(Fig.7)**. Origin of swelling could not be identified immediately after exploration. Swelling was well dissected from all sides releasing adhesions and delivered out. It was turned out to be left side ovarian cystic swelling got torted on right side occupying whole of right side of abdomen. Left side oophorectomy with fallopian tube excision was done by removing the specimen in toto **(Fig.8).** Post operatively baby was shifted to intensive care unit and was discharged on post-operative day 7 on full feeds. Histopathology study showed matured ovarian teratoma. Baby was discharged on postoperative day 14 with uneventful recovery.

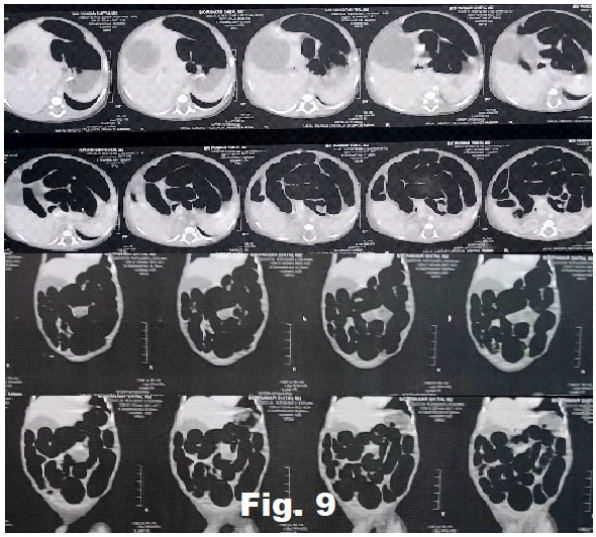


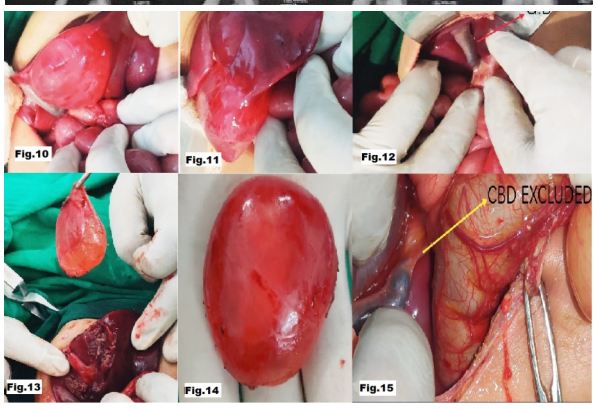




Case 3:-

Antenatal suspected case of fetal intra-abdominal cyst diagnosed at 26 weeks of gestation with normal rest of parameters got delivered on 36 weeks of gestation with planned caesarean section. Serial fetal scans were done till delivery showed gradual increase in size of intra-abdominal fetal cyst. After birth, baby was shifted to intensive care unit. In first 24 hours of life after birth baby started developing jaundice and gross distension of abdomen. Immediate ultrasound study of abdomen showed huge cyst in right hypochondriac region compressing gall bladder and swelling was involving right lobe of liver. Computed tomography study of abdomen **(Fig.9)** revealed same picture with gaseous distension of bowel loops suspecting the case of duplication cyst. Due to compression effect on diaphragm, tachypnea, jaundice (Direct bilirubin > indirect bilirubin) and increasing abdominal distension, emergency exploratory laparotomy was planned. Rest of hematological work up was normal. On exploration there was huge cyst arising from left lobe of liver compressing gall bladder and common bile duct and crossing over to right lobe of liver. Rest of the bowel was absolutely normal. Liver was delivered out and complete cyst enucleation was performed sparing gall bladder and common bile duct. Complete in toto excision of cystic swelling could be done **(Fig.10 – 15)**. Swelling was invaginating into liver parenchyma on either side. Hemostasis has been confirmed. Postoperative baby was shifted to intensive care unit and after 48 hours of surgery feed were started. Baby was discharged on post-operative day 10 without any complaints. Jaundice got resolved immediately after surgery. Histopathology study showed findings suggestive of Simple hepatic cyst with clear fluid as content. On follow up baby presented with good weight gain with no any complaints.





Case 4:-

A full term normally delivered baby boy with normal antenatal scans presented with abdominal distension with history of not passed meconium for 72 hours even after glycerin rectal wash. Plain x ray abdomen on day 2 of life reveled gas shadows in small bowel with no gas shadow in colon **(Fig.16)**. All hematological work up was normal. Rectal dye study was performed after 72 hours which revealed evidence of microcolon suspecting small bowel atresia **(Fig.17)**. On emergency exploratory laparotomy performed on day 3 of life showed pus in the abdomen with ruptured cyst just beneath the inferior border of left lobe of liver extending towards stomach. There were lots of pus flakes with plastered peritonitis and lump of small bowel loops. Complete excision of ruptured cyst was done (Antenatal intrauterine rupture of duplication cyst) **(Fig.18,19,20)**. On exploring the lump of small bowel there was evidence of distal jejunal atresia **(Fig.21)**. Jejunal anastomosis was done. Abdominal cavity was washed thoroughly and closed in layers after putting intra-abdominal drain in situ. Baby was shifted to intensive care unit. On post-operative day three baby passed motions. No leakage from anastomosis site was confirmed with oral water-

soluble dye study and gradually feeds were started. Baby was discharged on post-operative day 10 with uneventful recovery. Histopathology report of sent specimen confirmed about ruptured duplication cyst showing the cystic wall with formation of the intestinal metaplasia, showing a sheet of enteric type mucosa rich in goblet cells and Paneth cells, which were infiltrated with myriads of erythrocytes.

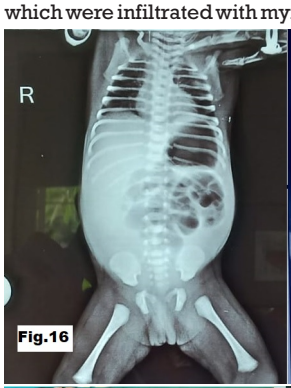
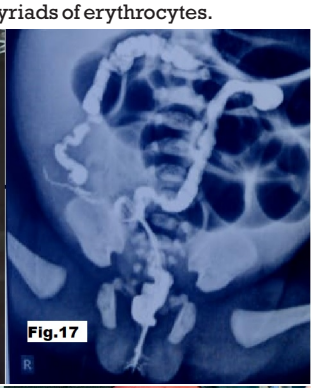


Fig.16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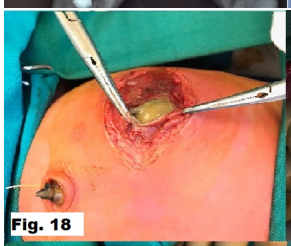
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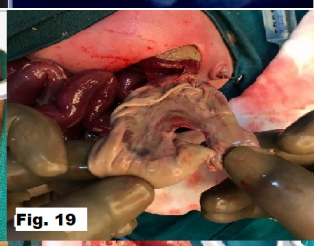
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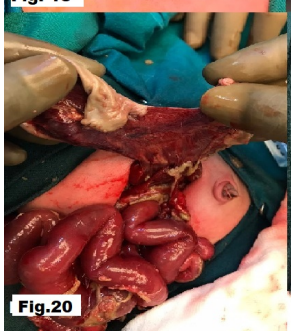
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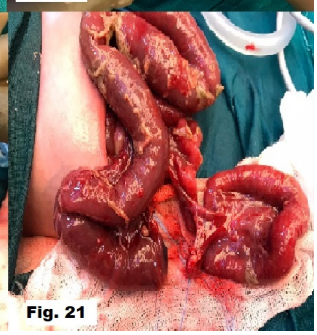
Fig. 21

All patients are kept under closed follow up.

DISCUSSION:-

Tumours in the neonatal period are extremely rare. Many tumours of infancy are unique to that age group but virtually all childhood neoplasms have been reported in the perinatal period also. Although there is no clear difference between the type of tumours that present at birth and those in early infancy, there appears to be some distinction due to the in utero influence of the congenital tumours by the maternal hormones and immunoglobulins. The pattern of metastasis is also different due to fetal circulation **(1)**

Extrarenal Wilms' tumour is a rare but challenging entity, considering its diagnosis, histopathology, staging, treatment, and prognosis. Diagnosis of extrarenal Wilms' tumor is always postsurgical, which may jeopardize treatment planning and consulting with parents in the first step. The histopathology of Wilms' tumor is very confusing. While most authors believe that it arises from primitive ectopic nephrogenic rests, teratoid Wilms' tumor leads to the debate whether this tumor is neoplastic or embryonic. Staging of extrarenal Wilms' tumor is also a challenge when we consider the National Wilms' Tumor Study (NWTS) recommendations; all these tumors should be considered as stage II or higher as they are beyond the renal capsule. This will mandate chemotherapy for all patients while most of the reported cases have a favorable histology, and long-term tumor-free survival has been reported even with exclusive surgery in some case reports. Although treatment strategies for extrarenal Wilms' tumor are the same as those for renal Wilms' tumor, different locations and neighboring organs may invoke special considerations and scenarios while planning for surgery and adjuvant therapies. Consulting with the parents is also a problem, considering the rarity of the disease and limited publications. Many literatures have mentioned in detail after a systematic review of extrarenal Wilms' tumor cases to date in order to provide a clear perspective for confronting this rare disease **(2)**. Surgical excision remains the key step in the treatment of ERWT, especially when performed radically **(3)**. Adjuvant chemotherapy is recommended for all ERWT cases postoperatively in spite of favorable histopathology in most of them. However, there are a few cases of successful treatment of stage I ERWTs with pure surgery. Chemotherapy regimen is determined by histology and stage of the tumor, considering the NWTS protocols that consist of the administration of vincristine, actinomycin D, and doxorubicin. Regarding the current guidelines of NWTS, completely resected ERWTs with no evidence of tumor at or beyond the margins are considered as stage II and treated with vincristine and actinomycin D, while addition of doxorubicin will have benefits in stage III ERWTs. Radiotherapy is reserved for unresectable tumors or for those with gross residue, recurrence, or metastasis **(4).**

Neonatal ovarian cysts are nearly always benign and self-limiting, and many of them go unreported **(5,6,7).** In girls, the frequency of such cysts was reported as approximately 5% of all abdominal masses in the first month of life **(8).** The first prenatal detection of a fetal ovarian cyst was in 1975 by Valenti **(9,10).** Patients who were antenatally diagnosed were followed by ultrasonography twice a month until birth. Ultrasonographic evaluation was repeated within 48 hours of birth. Patients who were postnatally diagnosed were followed twice a month during the first three months, then once a month until the cysts shrank and disappeared. The cysts were classified according to their size as "small" for cysts smaller than 40 mm in diameter and "large" for those larger than 40 mm. Besides, according to their ultrasonographic features they were classified as "simple" and "complex". The Nussbaum criteria were used for discrimination between complex and simple cysts. According to these criteria, "simple cysts" are completely anechoic, homogeneous, thin-walled, and are frequently unilocular and located unilaterally. Thick-walled cysts having a solid structure and septa, and which contain blood clots and debris are "complex cysts" **(11,12).** Neonatal ovarian cysts may cause pain, irritability, vomiting, fever and abdominal distension. Peritonitis, anemia due to intracystic hemorrhage, fetal tachycardia due to peritoneal irritation or anemia, and sudden infant death syndrome may also occur **(13,14,15).** The large cysts may cause intestinal and urinary obstruction due to their size, dystocia, and therefore abdominal and thoracic mass effect gives rise to pulmonary hypoplasia and polyhyramniosis. When a cyst is torsioned, its size rapidly increases and its ultrasonographic features change to complex. The ovarian torsion frequently leads to further complications such as rupture resulting in hemoperitoneum, ascites stemming from transudation, adhesion with adjacent organs resulting in urinary and intestinal obstruction, calcification of cyst walls, and auto-amputation of the ovary. On abdominal examination, a mobile mass can be palpable if auto-amputation of a torsioned ovary occurs. Complex cysts (mostly torsioned simple cysts) cannot be distinguished from other intra-abdominal pathology and require surgical exploration. In fetal life, when intracystic haemorrhage and ovarian torsion are detected, preterm birth is necessary after lung maturity has been attained **(16).**

Congenital hepatic cyst is a rare and asymptomatic condition in infants and children. Its incidence is 2.5% in the postnatal life with a much lower incidence in the prenatal period. Incidental finding on antenatal imaging is the most common presentation. **(17,18).** These cysts do not contain bile and arise from congenital or secondary obstruction of the biliary

glands, which normally arise from the ductal plate at the hepatic hilum around the seventh week of gestation and continue to proliferate until adolescence. They are superficially located just under the liver capsule and are coated by a single layer of cuboidal or columnar epithelium, characteristic of the bile ducts. The cystic fluid is generally clear and rarely contains bile **(19).** Cholestasis may also be observed and in some cases due to the compression of the hepatic parenchyma and the biliary system by a large cyst. Management of congenital hepatic cyst is conservative with periodic ultrasound monitoring to ensure their stability, especially for large cysts (>4cm in diameter) **(20,21).** Most simple hepatic cysts are benign in nature and have a spontaneous resolution as in our patient **(22).** Surgical intervention with aspiration, sclerotherapy, or excision is indicated only for severe cases, such as hydrops, progressive enlargement, haemorrhage, torsion, or if image characteristics prompt diagnostic doubt. **(23,24).** Compression of intrahepatic structures due to an increase in the cyst size also prompts surgery; hence, ultrasound follow-up is important **(25).** Surgical treatment with partial or total removal of the cyst is the favoured treatment in neonates with a large symptomatic lesion, with an objective of excising as much of the cyst wall as possible without causing damage to the surrounding vital organs.

As a potentially life-threatening disease, an enteric duplication (ED) or intestinal or enteric duplication cyst (EDC) has all the time been perplexing paediatric surgeons. It is really a challenging task to make a clinical diagnosis mainly due to its rarity and nonspecific presentations, unless complications ensue unexpectedly. An enteric duplication cyst is an unusual congenital deformity of the alimentary system, which is a separate entity invested with a cystic appearance, but at the same time is in intimate contact or communication with the alimentary tract **(26).** This entity consists of a wide variety of cystic lesions originating from different sites alongside the digestive tract between the mouth and the anus in conjunction with their neighbouring organs, with a predilection for the small bowel, especially the ileum **(27,28).** It takes place most commonly in infants and children, with an estimated incidence of approximately 1 in 4500 births, **(29)** but is exceedingly uncommon in adults. By definition, an ED or EDC is characterized by a well-developed muscular wall along with a lumen lined by ectopic mucosa, **(30)** which bears a striking resemblance to a portion of the normal bowel tract. It is endowed with the similar layered muscles with the normal bowel wall, and frequently even shares a common muscular wall. The ectopic mucosa is typically lined by gastrointestinal-type epithelium ranging from gastric to rectal component, which is usually compatible with the adjacent bowel, while different epithelial constituent other than gastrointestinal origin has been sometimes identified, commonly being respiratory epithelium **(31).** Normally, a single epithelial lining is present, the majority being gastric, although a combination of multiple types of epithelia coexisting in the same duplication has occasionally been disclosed **(32,33).** Intra uterine rupture of enteric duplication cyst is very rare and associated with intestinal atresia is barely reported in literature.

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