Pediatric peritoneal lesions, Radiological -Pathological correlation.

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Learning objectives

Peritoneal tumors are rare in children, including group of benign and malignant tumors. Our objectives are to:

_ Describe the clinical and pathologic features of peritoneal tumors in pediatric age group.
_ Identify the cross-sectional imaging patterns of pediatric peritoneal tumors.
_ Discuss the differential diagnosis of peritoneal tumors in children.

Background

About 30% of pediatric soft tissue malignancies occur in the abdominopelvic cavity. The rate of recurrence is higher for the peritoneal tumors compared to other anatomic sites. For abdominopelvic tumors, extension of tumor to involve the peritoneum is one form of disease progression. Pediatric patients with peritoneal metastasis have a poor long-term outcome.

Peritoneal tumors in children have a similar clinical presentation. Patient's usually present with abdominal discomfort, abdominal distention, gastrointestinal complaints, and, less commonly, pelvic pain or a palpable mass. The peritoneal masses in children include benign and malignant conditions, the metastatic malignant lesions are more common than the primary ones. Peritoneal lymphoma either primary or secondary. The primary malignant peritoneal lesions include desmoplastic small round cell tumor, germ cell tumors and less common mesothelioma. Benign conditions include mature teratoma, inflammatory myofibrobalstic tumor, lymphangioma and tuberculous peritonitis.

The knowledge about the diagnostic clues of the peritoneal lesions is crucial so that they can be appropriately included in the differential diagnosis in patients presenting with diffuse or focal peritoneal disease processes for their early diagnosis and proper management.

Normal Peritoneal Anatomy

The peritoneum is formed of thin, translucent serosal membrane covering the peritoneal cavity and its mesenteries. The visceral organs are also partially or completely cover by peritoneum.
The peritoneum provides a frictionless surface over which the viscera can move and serves as a site of fluid transport. Normally, the peritoneal cavity contains a small amount of sterile fluid acting as a lubricant and as a local bacterial defense.

The peritoneum is formed of visceral and parietal layers. The visceral layer covers the intraperitoneal organs (stomach, jejunum, ileum, transverse colon, sigmoid colon, liver, and spleen), omentum, and mesentery. The parietal peritoneum lines the abdominal walls, undersurface of the diaphragm, anterior surface of the retroperitoneal viscera (duodenum, ascending and descending colon, pancreas, and portions of the adrenal glands and kidneys), and the pelvis. The intraperitoneal organs are suspended and supported within the peritoneal cavity by peritoneal ligaments and mesenteries. The peritoneal ligaments and mesenteries subdivide the peritoneum into compartments connected together that determine the method of spread of malignant tumors.

Findings and procedure details

I. Metastatic peritoneal tumors

Metastatic peritoneal tumors are more common than primary peritoneal tumors. Understanding the mechanism of tumor spread through the abdomen and common pathways for metastases is essential for complete detection. It usually occurs due to disruption of the organ capsule by the tumor or due to peritoneal spill at the time of surgery. It may occur by hematogenous spread, or by lymphatic spread of disease and occasionally in patients with ventriculoperitoneal shunts.

The imaging findings include: ascites, mesenteric or peritoneal nodules (<3cm) or masses (>3cm), calcification may be present, bowel wall thickening and omental caking can also occur. The appearance of pseudomyxoma-like which is characterized by scalloping of the solid organs has been described in some cases such as rhabdomyosarcoma, Wilm's and Burkitt lymphoma.

Tumor dissemination can occur at any site in the peritoneum. The spread of intraperitoneal malignancy follows the peritoneal fluid circulation, and so the most common location of tumor seeding is: Douglas pouch, small bowel mesentery, the sigmoid mesocolon, the right paracolic gutter and the right sub hepatic and subphrenic spaces. The omentum is another common site of peritoneal invasion.
CT and MRI are the widely used imaging modalities for the evaluation of peritoneal tumors. The use of oral and intravenous contrast is mandatory in differentiating tumor from abdominal organs, bowel, and vessels. The most common primaries associated with peritoneal deposits in children are:

A Germ Cell Tumor

Germ cell tumours are found widely throughout the body, and encompass a wide range of individual tumours. Germ cell tumours arise from ectopic pluripotent stem cells that failed to migrate from yolk endoderm to the gonad. Because they arise from primitive cells, they have variable neoplastic potential and variable degrees of differentiation into a variety of tissues.

It classified into seminatous and non seminatous types. In children yolk sac tumor is the commonest and most aggressive subtype. The peritoneal deposits associated with YST considered as one of the poor prognostic factors (Fig. 1).

The immature teratoma also can associated with peritoneal deposits, the typical appearance of teratoma as a mixed solid and cystic mass with calcification and fat components may help in diagnosis, yet frequently, the immature teratoma lack considerable amount of the calcification and fat, in these cases the pathological diagnosis is mandatory. (Fig. 2&3)

B. Rhabdomyosarcoma

Rhabdomyosarcoma is a malignant soft tissue tumor of childhood that arises from primitive mesenchymal cells. It is one of the most common malignant soft tissue tumors in children, representing about 5-15% of all solid malignant tumors. Most of cases are less than 10 years old at time of diagnosis.

It may arise from almost all body organs and metastasize to different sites. It can occur in the peritoneum and retroperitoneum. The most common sites are head and neck and genitourinary.

Peritoneal metastasis may present at time of diagnosis or over the course of the disease.

The tumor under the microscope has characteristic skeletal muscle lineage-either by its appearance or by the pattern of chemical staining (“immunostaining”). There are two main types of RMS, embryonal and alveolar. The embryonal type is more common in children.

It may present with a focal mass or ascites or signs of abdominal complications, such as bowel or urinary obstruction or generalized spread.

The imaging findings include multiple enhancing masses, omental caking or pseudomyxoma peritonei-like picture. (Fig. 4)
The rate of recurrence after surgical excision is very high, and thus it has a poor prognosis.

**C. The atypical teratoid rabdoid tumor (ATRT)** is a recently identified aggressive tumor of unknown origin. It usually occurs in infancy and young children. The primary site of ATRT is the kidney and brain yet it can occur at any age. We report a case of pelvic ATRT at age of 10 year associated with peritoneal deposits (Fig.5).

**D. Wilm's Tumor or nephroblastoma**

Wilm's tumor arises from the metanephros, the mesodermal precursor of the renal parenchyma. It may arise in the extrarenal retroperitoneum from mesonephric remnants.

Wilm's tumor represents about 87% of renal masses in children and its incidence is about 1:10,000 children. It represents about 7% of all cancers in children. The most common age is at 3-4 years with 80% occurring under five years.

The most common presentation is palpable mass. Hematuria and pain are less common. Arterial hypertension may be present in up to 25%. Tumor pathology is very important and determines prognosis. Tumors are classified as having favorable (FH) or unfavorable histology (UH). Tumors with prominent tubular differentiation, no anaplasia, no sarcomatous elements are considered favorable.

The metastases are most commonly found in the lungs (85% of cases). Metastases to the liver and lymph nodes also occur.

Peritoneal extension occurs if the tumor breaks through the renal capsule before surgery or rupture during surgery.

Peritoneal cavity invasion results in thickening, nodules, and masses in the omentum or mesentery. The pelvis is also a common site of seeding metastases. Pseudomyxoma-like appearance may also occur (Fig. 6).

**II. Primary peritoneal tumors**

**1. Desmoplastic Small Round Cell Tumor (DSRCT)**

Desmoplastic small round cell tumor is a rare malignant tumor. It occurs most commonly in the young adult. It has a poor prognosis, the 3-year survival rate not exceeding 30 %. It is a distinctive clinic pathologic entity in the family of primitive pediatric tumors that are
composed of small, round, blue cells; a group that includes Wilm's tumor, Ewing sarcoma, peripheral primitive neuroectodermal tumor, and Askin tumor.

The most common primary site is the peritoneal cavity. Other primary sites including paratesticular, pleural, and nonserosal desmoplastic small round cell tumors. It may occur in older age group and in females.

The desmoplastic small round cell tumor may be a single or multiple, gray to white, firm discrete and confluent peritoneal masses. The desmoplastic small round cell tumor is composed of cords or nests of undifferentiated malignant small round cells with surrounding dense, collagenous stroma. The malignant cells have scant cytoplasm and large, hyperchromatic nuclei with characteristic single cell necrosis and numerous mitotic figures.

The desmoplastic small round cell tumor spreads throughout the peritoneal surfaces presenting initially as diffuse peritoneal thickening, nodules or masses. The initial presentation may be a solitary peritoneal mass. The dominant peritoneal mass reaches large size (>10 cm) in most of cases. The peritoneal masses are heterogeneous showing necrotic areas showing heterogeneous pattern of enhancement and may show small calcifications. (Fig.7). They elicit heterogeneous low T1 and bright T2 signal at MR imaging with heterogeneous pattern of enhancement after intra venous contrast administration. The tumor may be complicated by intestinal or ureteric obstruction. The ureteric obstruction is more common in large pelvic masses. Metastases, whether lymphatic or hematogenous, are common at time of diagnosis (about 50 % of cases have metastasis at initial presentation) to the liver, lung, and bone.

Desmoplastic small round cell tumor is considered in the differential diagnosis of solitary or multiple peritoneal masses in a young male. Other possibilities which should be also considered include peritoneal carcinomatosis and lymphoma as well as benign entities such as splenosis. The presence of dominant single or multiple masses within the diffuse peritoneal process as well as presence of calcification and necrosis is suggestive of desmoplastic small round cell tumor rather than other lesions.

2. **LYMPHOMA**

Peritoneal lymphomatosis is not common. Many subtypes of lymphoma may be associated with peritoneal involvement. It is more common with aggressive pathological subtypes of high- grade lymphoma. The most common type is diffuse large B-cell lymphoma.

Peritoneal lymphomatosis may be similar to peritoneal carcinomatosis. Lymphomatous involvement of the peritoneum is usually associated with bowel and mesenteric infiltration.
The most common CT findings of peritoneal lymphomatosis are peritoneal thickening, enlarged lymph nodes, omental and mesenteric infiltrations, lymphomatous mass, bowel wall thickening, hepatosplenomegaly with focal lesions and ascites (Fig. 8).

CT findings in Burkitt lymphoma could be similar to mucinous tumor in adults or may be confused with desmoplastic small round cell tumor in children and adolescents, and may have a similar clinical presentation (Fig. 9&10).

At ultrasound, it appears as hypoechoic mesenteric masses or, rarely in Burkitt lymphoma, as omental caking.

PET/CT delineates metabolically active tumor and is helpful for detection of distant metastases as well as in staging and monitoring response to therapy.

3. **Mesothelioma**

Mesothelioma is the primary malignant tumor of the pleural, peritoneum and pericardium as well as the tunica vaginalis.

It is rare in childhood and in contrast to adults, there is no evidence of relationship between mesothelioma in children and asbestos exposure.

Peritoneal mesothelioma may occur in children at any age and is more common in girls.

The prognosis is poor, however has improved recently with aggressive multimodality therapy.

Imaging findings include thickened peritoneum, omental and mesentric nodules or masses and ascites.

C. **Benign peritoneal lesions**

1. **Plexiform neurofibroma**

The neurofibromas and plexiform neurofibromas of the mesentery and gastrointestinal tract arise from nerves of the mesenteric, subserosal, and myenteric plexuses. Mesenteric plexiform neurofibromas manifest as infiltrating lesions extending from the root of the mesentery to the wall of the intestine. By U/S, they appear as well-defined homogeneously or heterogeneously hypoechoic masses. By CT, they appear as iso- or hypoattenuating masses (Fig.11). The appearance may mimic adenoapthy in lymphoma.
By MRI, it gives characteristic ring like or septated fascicular pattern, which is not seen in adenopathy.

Mesenteric neurofibromas can encroach on the adjacent bowel and causing mass effect on the serosal surface, or may infiltrate into the adjacent bowel wall, producing submucosal or mucosal masses. Neurofibromas which infiltrate the adjacent intestinal wall appear as focal or diffuse mural thickening.

**Mesenteric inflammatory myofibroblastic tumor** IMFT

An inflammatory myofibroblastic tumor (IMFT), also called an inflammatory pseudotumor, is a rare pseudosarcomatous inflammatory lesion that consists of inflammatory cells and myofibroblastic spindle cells. An IMFT commonly involves the lung and the orbit, but the tumor has been reported to occur in a variety of tissues and organs. IMFTs that involve the mesentery or omentum present with abdominal masses that are associated with fever and abdominal pain.

Fever may be a manifestation of an inflammatory response, and abdominal pain maybe related to the compression effect of the tumor.

Mesenteric or omental IMTs appeared as well-defined solid, mixed-echogenic masses within the mesentery as seen on sonography and prominent vascularity as depicted on Doppler sonography. On CT scans, a mesenteric IMT shows typically heterogeneous attenuating enhancement. (Figs.12). these lesions were considered sarcomas, lymphomas, or IMTs.

### 3. Teratoma

Teratomas are germ cell tumors composed of the embryonic germ layers and contain structures usually foreign to the anatomic site of origin. Germ cell tumors are relatively rare, except for mature cystic teratoma of the ovary. The testes and ovaries are the most common sites of germ cell tumors, but they may also occur at the anterior mediastinum, intra cranial and the sacrococcygeal region. The peritoneum is a rare site of extragondal teratomas.

Teratomas may be benign, mature, well-differentiated cystic lesions or immature, poorly differentiated with solid components and malignant transformation.

The immature teratoma contains immature neural tissue and has a great rate for recurrence.
About 60% of ovarian tumors are germ cell tumors in patients younger than 21 years, and one-third of those are malignant.

The most common presentation is abdominal distention due to the tumor and ascites. Complications can also occur as rupture or torsion. Ovarian germ cell tumors are commonly associated with ascites and intraperitoneal spread. (Fig. 13)

4. **Lymphangiomas**

It may represent either congenital malformations of the lymphatic system or benign neoplasms and typically appear as large, thin-walled, usually multiloculated cysts at CT. Mesenteric infiltration is common, and removal often necessitates bowel resection; an omental origin is less commonly seen. At CT, the cyst walls are often imperceptible, but vessels may be seen coursing between locules. Cyst contents can have an attenuation less than that of water due to their chylous nature, which can be a key distinguishing feature.

Images for this section:
12 Year female patient presented with abdominal mass

A n ill defined predominantly solid mass lesion occupying the pelvic cavity component. No definite areas of fat density or calcified foci are noted within. Associated perihepatic deposit is noted.

Pathological dx: Immature teratoma
After chemotherapy

Fig. 3

Newly developed notable calcific foci within the mass and within the prehepatic peritoneal deposit. Although the mass size increase and moderate amount of ascites appeared. Pleural deposit is also seen (arrow)
14 year old male presented with mandible swelling

Multiple nodal, peritoneal, and lung metastasis

Rhabdomyosarcoma

Fig. 4
10 year old female patient presented with abdominal swelling

A huge well-defined heterogeneous hypodense solid mass is seen occupying the lower abdomen and pelvis. It is seen extending into the right retroperitoneal region extending behind the right psoas muscle compressing it and infiltrating it. No intrinsic calcifications. No intraspinal extension. Associated moderate amount of ascites is noticed with peritoneal thickening and nodularity.

Atypical teratoid rhabdoid tumor

Fig. 5
7 year male presented with growing abdominal swelling

Large left renal mass with multiple peritoneal and mesenteric masses

Wilm’s tumor

Fig. 6
17 year old female presented with abdominal swelling

Marked ascites with multiple variable sized masses with faint marginal calcifications and central necrosis

Desmoplastic small round cell tumor

Fig. 7
4 year old male presented with abdominal swelling

Marked ascites, multiple mesenteric and retroperitoneal nodal masses and omental caking in addition to bilateral renal infiltration

Lymphoma (NHL)

Fig. 8
7 year old male presented with abdominal discomfort and swelling

Marked ascites, multiple mesenteric, peritoneal and pelvic masses in addition to large bowel mass

Burkitt lymphoma

Fig. 9
13 year old female presented with abdominal swelling

Marked ascites with bilateral ovarian masses and omental caking

Burkitt lymphoma

Fig. 10
12 year male presented with abdominal discomfort

Large mesenteric mass

Mesenteric plexiform neurofibroma

Fig. 11
1 year old boy presented with abdominal discomfort

Fig. 12
1 years old male patient presented with slowly growing abdominal mass.

An ill-defined large heterogeneous solid and cystic lesion is seen at the retroperitoneal region showing areas of fat density and calcified areas.

Pathological diagnosis: Mature teratoma

Fig. 13
Conclusion

The differential diagnosis for tumors of the peritoneum in children is wide. The secondary malignancy is the commonest with rhabdomyosarcoma, lymphoma, Wilm's tumor, germ cell tumors are the commonest primaries. The primary malignancies include DSRCT and primary lymphoma with mesothelioma rare in children. Also there are variable benign conditions can occur in peritonitis in children. The radiological awareness of different patterns of these lesions is of utmost importance helping in proper diagnosis and staging of children.

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References


