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Miscellaneous conditions of the peritoneal cavity—Peritoneal tumors, pseudomyxoma, mesothelioma, fibroblastic reaction, cocoon, cystic lymphatic malformations, blue-bleb, and chylous ascites

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ABSTRACT

The peritoneum is subject to both primary neoplasia and secondary malignancy from direct, trans-coelomic, or hematogenous spread from any cancer. The knowledge base in the pediatric age group is very limited due to the rarity of peritoneal conditions in children, and much of the information is extrapolated from adult literature. There have been few reports in the pediatric population on the diagnosis and management of peritoneal conditions including peritoneal malignancy. In this article, we aim to highlight some of these conditions and the treatments available with a special emphasis on the evolving role of cytoreduction surgery and hyperthermic intraperitoneal chemotherapy in the treatment of certain peritoneal malignancies in children.

Tumors

Introduction

The peritoneum is a serous lining of mesothelial cells with a rich vascular and lymphatic supply. Thus, the peritoneum is subject to primary peritoneal malignancy or secondary neoplasia (due to direct or trans-coelomic spread from an intra-abdominal organ malignancy) or a hematogenous metastasis from any cancer. Though peritoneal malignancy is rare in children, peritoneal abnormalities can occasionally be seen at cross-sectional imaging or at laparotomy, or laparoscopy.

The knowledge base in children is very limited due to the rarity of peritoneal malignancy, and much of the information is extrapolated from the adult literature. Thus, for example, one of the classical primary peritoneal tumors is multicystic peritoneal mesothelioma (Figure 1), which traditionally was categorized as "benign," but its tendency to recur if not completely removed implies that it is, at best, a "borderline malignancy," and optimal treatment is now considered tumor removal combined with hyperthermic intraperitoneal chemotherapy.[1] Other primary peritoneal tumors are frankly malignant such as papillary or diffuse malignant mesothelioma, peritoneal serous carcinoma or sarcoma, and malignant tumors of Mullerian origin (Table 1).

The common secondary peritoneal tumors in adults arise from ovarian, colorectal, appendiceal, gastric, or pancreatic carcinoma and various sarcomas including gastrointestinal stromal tumors (GISTs) and uterine leiomyosarcomas.[2] The situation in children is different, with secondary deposits most commonly associated with neuroblastoma, rhabdomyosarcoma, and germ cell tumors.

There have been few reports in the pediatric population on diagnosis and management of peritoneal malignancy, though recently there have been some encouraging results especially in the field of desmoplastic small round cell tumors and rhabdomyosarcomas.[3,4] It is relevant to note that Hayes-Jordan et al.[3] report on treatment in individuals up to 21 years of age, which would generally be considered well outside the pediatric age group, though many of their patients are much younger than 21 years and similar treatment principles apply.

The commonest treatable secondary peritoneal malignancies in adults are of appendiceal, colorectal, and gynecological origin.[5] Again, there are few reports on such tumors in children. Late diagnosis is common in the children unfortunate enough to develop these primary cancers.

In adults, there has been an increasing interest in attempting curative treatment of peritoneal malignancy by a combination of complete macroscopic tumor removal [cytoreductive surgery (CRS)] and hyperthermic intraperitoneal chemotherapy (HIPEC)[5] particularly for appendiceal tumors.[6] One particular area of increasing global interest has been in what is best described as resectable "colorectal peritoneal metastasis" from primary colorectal cancer.[7]

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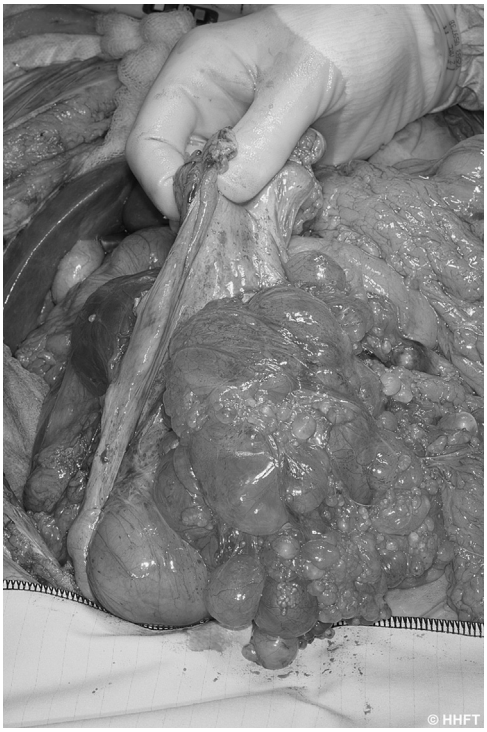

Fig. 1. Multicystic intraperitoneal mesothelioma.

Similarly, the techniques of CRS and HIPEC have applications in children,[3] and all who are involved in imaging or operating in the abdominal cavity may occasionally encounter a suitable case of primary or secondary peritoneal malignancy.

There are a number of other intra-abdominal entities that may simulate peritoneal malignancy in children such as blue-bleb, fibroblastic reaction and cocoon, and chylous ascites, which will be discussed later.

Background

The pathophysiology of intraperitoneal tumor cell distribution has been classified as the "redistribution phenomenon" whereby free-floating intra-abdominal cells concentrate in the pelvis and the paracolic gutters by the effects of gravity and at sites of normal peritoneal fluid absorption, particularly the greater and lesser omentum and the under-surface of the diaphragm, especially on the right side.[8] The motile organs, particularly the small bowel, are generally spared, allowing tumor removal with preservation of the small bowel.

Peritoneal malignancy, as in adults, may be suspected at cross-sectional imaging (US, CT, or MRI) or may be detected at laparoscopy or laparotomy. Unfortunately, in children, the low index of suspicion for abdominal malignancy often results in presentation at an advanced stage.[9] Thus, CRS and HIPEC may not be of benefit in most such cases due to extent of disease.

Table 1
Classification of peritoneal tumors (College of American Pathologists—www.cap.org).

Benign
Adenomatoid tumor
Benign multicystic mesothelioma[a] (multilocular peritoneal inclusion cyst)
Mesothelial cyst(s) (unilocular) (free or attached)
Well-differentiated papillary mesothelioma
Solitary fibrous tumor (fibrous mesothelioma) (usually benign)
Malignant
Diffuse malignant mesothelioma
Epithelioid type
Sarcomatoid type
Biphasic type
Rare types[b]
Serous tumor of borderline malignancy (of low malignant potential)[c]
Serous carcinoma
Malignant tumors of other Mullerian types
Sarcomas

[a] Multicystic mesothelioma tends to recur or persist despite surgery; this condition is at best a "borderline malignancy" compared to "benign."

[b] Rare types include desmoplastic, small cell, lymphohistiocytoid, deciduoid, and undifferentiated types.

[c] When this tumor involves the extraovarian peritoneum significantly and the ovarian surface minimally or not at all, it is generally considered to be of peritoneal origin.

Tumors

Desmoplastic small round cell tumors (DSRCT)

There have been recent reports of CRS and HIPEC as part of the treatment of children with DSRCT.[10] DSRCT was first described in 1989, with approximately 200 cases subsequently reported in the literature.[4] The overall survival with DSRCT is about 30–55% after a combination of chemotherapy, radiotherapy, and surgery.[11] Traditional treatment of DSRCT has been with the "P6 regime" (cyclophosphamide, doxorubicin, and vincristine alternating with ifosfamide and etoposide) followed by debulking surgery.[11] As DSRCT presents with multiple tumor implants, aggressive debulking may result in significant residual tumor, and thus, CRS and HIPEC has recently been advocated as a "novel treatment" strategy.[4]

Mesothelioma

Mesothelioma is more common in adults and rarely seen in children. It derives from mesothelial cells and can arise from the serosal lining of the pleura, peritoneum, pericardium, or tunica vaginalis testis in the male.[12] It is estimated that one-fifth to one-third of all mesotheliomas are primary peritoneal.[13] The more frequent pleural mesothelioma has been linked with asbestos exposure, often several decades previously, though the exact pathogenesis of pleural or peritoneal mesothelioma in many cases is largely unknown.[14] Over 50% of patients with mesothelioma are between the ages of 45 and 65 years, though mesothelioma can present at any age, including in children.[15] The main variants of mesothelioma are cystic, epithelioid, and sarcomatous, with cystic having the best outcome and sarcomatous the least favorable. Epithelioid variants have intermediate prognosis. The combination of CRS and HIPEC has been shown to be beneficial in treating adults with abdominal mesothelioma, with a median survival up to 60 months and 5-year survival of 50% in selected patients.[12]

Rhabdomyosarcoma

Rhabdomyosarcoma is a soft tissue malignant tumor that is thought to arise from primitive mesenchymal cells. Overall, 70% of children with rhabdomyosarcoma are under 10 years of age.[16] Although head and neck (40%) and genitourinary (20%) are the most common primary sites, rhabdomyosarcomas have been known to arise from, and metastasize to, nearly all body organs. Overall, 10% of children with metastases have intraperitoneal involvement in the form of ascites, omental caking, peritoneal nodules, or masses either at the time of diagnosis or subsequently thereafter.[16] Recent reports suggest a role for CRS and HIPEC in selected cases.[3]