Primary retroperitoneal mucinous cystadenoma: An unusual cause of an abdominal mass in a child

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Abstract

Primary retroperitoneal mucinous cystadenomas are uncommon tumors whose incidence in the pediatric population is very low. We present the case report of a 17-year-old female with a primary retroperitoneal mucinous cystadenoma, borderline type, and describe the successful laparoscopic resection of the mass.

Key words:
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Primary retroperitoneal mucinous cystadenoma (PRMC) is an exceedingly rare type of neoplasm found primarily in adult women, with the total number of documented cases less than 50. Its incidence in the pediatric population is described in only one other case that the authors are aware of [1]. We present the case of laparoscopic resection of a PRMC of borderline type occurring in a 17-year-old female, and briefly review the literature on the subject.

1. Case report

The patient is a 17-year-old female with no past medical history who initially presented to her pediatrician with complaints of abdominal fullness and vague abdominal pain. Physical exam revealed an obese abdomen, soft, tender to palpation in the left upper and left lower quadrants, with no palpable masses. Review of systems was remarkable for mild diarrhea which, according to the patient, was attributable to anxiety. The only medication the patient was taking was low estrogen oral contraceptives. CT scan of the abdomen and pelvis was performed with 5 mm thin sections, with administration of intravenous and oral contrast. The scan revealed a large simple cyst situated in the left pelvis and extending into the left mid-abdomen, measuring 10.3 × 7.1 × 10.9 cm (Fig. 1). The cyst was without septations or enhancement and demonstrated a mass effect of the adjacent distal descending colon and sigmoid, pushing the sigmoid anteriorly, suggesting a retroperitoneal origin. The cyst was separate from the left ovary. Preoperative laboratory workup included a complete blood count with differential (WBC = 12.3), quantitative beta-HCG (normal), and alpha fetoprotein measurements (normal).

The patient underwent a laparoscopic resection of the mass. A 12 mm trocar was inserted through the umbilicus. Two additional 5 mm trocars were inserted in the suprapubic region and right lower quadrant. The position of the cystic mass was identified on the left side within the retroperitoneum pushing the left colon out of its usual position. The retroperitoneum was opened lateral to the colon on top of the mass using the harmonic scalpel (Fig. 2).
and the colon was mobilized medially (Fig. 3). The mass was identified and appeared blue and cystic in nature (Fig. 4). Blunt dissection was used to enter the retroperitoneum and the mass was freed from the surrounding tissues. The umbilical trocar was removed and a 15 mm bag was placed into the peritoneal cavity. The mass was placed in the bag and the edges were brought through the umbilicus (Fig. 5). Fluid was aspirated and sent for cytopathology. The mass was then decompressed of approximately 650 cc of clear, slightly mucinous fluid. The bag containing the mass was then removed. The patient was discharged home on the same day.

The surgical pathology report demonstrated the mass to be a primary retroperitoneal mucinous cystadenoma, borderline type. The cyst showed a fibrous wall with scattered calcifications and focal ovarian type stroma without oocytes. It was lined by columnar mucinous epithelium with focal complex architecture and papillary formations. Scant mitotic figures were identified within the epithelium. There was no cytological atypia, and no invasion into the stroma. Immunohistochemical staining was reported as below: Cytokeratin 7 (+), Cytokeratin 20 (−), PAX-8 (+), CA-125 (+), CA 19.9 (+), CDX-2 (−), Estrogen receptor [(+)] nuclear staining in stroma, (−) epithelium, Progesterone receptor [(+] nuclear staining in stroma, (−) epithelium).

The above findings are indicative of a mucinous cystadenoma of probable Müllerian origin. The mucinous epithelial complexity is taken as evidence that this represents a borderline lesion. The lesion did not communicate with the ovaries, fallopian tubes, bowel, pancreas or biliary system, and is therefore interpreted as being primarily retroperitoneal in origin.

2. Discussion

Primary retroperitoneal mucinous cystadenomas are extremely rare. They have been reported mainly in middle aged women, although there have been multiple reports of occurrences in men [2]. There is only one other case report of a PRMC occurring in the pediatric population [1]. There are four general theories regarding the pathogenesis of PRMC. One theory is that the tumors arise as a result of ectopic ovarian tissue, and ovarian like stroma is often
found in these tumors [2,3] including our patient. However, Falidas et al. [2] note that frank ovarian tissue is rarely found and there have been reports of these tumors in men. The second hypothesis is that these tumors are a result of a teratoma in which the mucinous epithelium has overgrown all other types of tissue [4,5]. The third hypothesis is that they arise from remnants of the urogenital apparatus in which, as with the teratoma hypothesis, the mucinous epithelium has overgrown all other tissue types [2,6]. The fourth, and most widely accepted, hypothesis is that of mucinous metaplasia of the peritoneal mesothelial layer. This theory supposes that during embryological development, multipotent mesothelial cells invaginate and become trapped in the retroperitoneum [7,8]. These cells then undergo metaplasia to mucinous epithelium, thus resulting in cyst formation [7,9–11]. These cells have been known to conform to a phenotype of extragenital Müllerian system [9], as was evidenced in our case.

PRMC’s are classified into three different types: mucinous cystadenoma, mucinous cystadenoma of borderline type, and mucinous cystadenocarcinomas, with the latter two types being more common than the first. In a recent clinicopathological study of 18 cases of PRMC’s, the examined cases diffusely stained positive for cytokeratin 7, as did our case [8]. Estrogen and progesterone receptor positivity has been noted variably in the literature [8,9]. Our case demonstrated positive staining for these receptors.

Pre-operative history, physical examination, laboratory and imaging studies almost always fail to confirm the diagnosis of PRMC. Symptoms are most often vague abdominal/flank pain and/or an abdominal mass [12]. Typical tumor markers such as AFP, CA 125, and CA 19.9 are inconsistent at best. On CT scan, PRMC’s usually manifest as a homogenous, unilocular cystic mass with displacement of nearby structures [6,12]. As discussed previously, our case was consistent with this typical radiological picture.

3. Conclusion

In conclusion, we describe a very rare incident of a primary retroperitoneal mucinous cystadenoma in the pediatric population. The possible diagnosis of retroperitoneal masses in children is a long list, including those of renal and adrenal origin, neuroblastomas and other nerve cell derived masses, lymphomas, sarcomas, germ cell tumors, and benign masses such as lipomas, fibromas, GI stromal tumors, and lymphatic malformations. We now know that PRMC must be included in this differential. Importantly, we demonstrate that PRMC’s can be safely removed laparoscopically in children. By adopting this approach, our patient avoided the morbidity associated with a laparotomy, was discharged and returned home the same day, and was adequately treated with complete resection without tumor spillage.

Consent

Not required.

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Conflicts of interest

None.

References