Retroperitoneal lipoblastoma causing chronic constipation in a young boy

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Constipation is a frequent occurrence in infants and children and a common indication for referral to pediatric gastroenterologists. Although most children will be diagnosed with functional constipation and respond well to laxatives, dietary changes, and behavioral modifications, a smaller subset of patients will not improve on first-line therapies. In these children, it is necessary to consider organic etiologies like an anatomic malformation, metabolic derangement, or neuropathy. Occasionally, abdominal or pelvic tumors will present with constipation. We report an unusual case of a retroperitoneal lipoblastoma causing constipation in a young boy. ***Ann Pediatr Surg*** **13:59–61 © 2017 Annals of Pediatric Surgery.**

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Introduction

Constipation occurs frequently in infants and children and is a common indication for referral to pediatric gastroenterologists. The majority of these children will be diagnosed with functional constipation and respond well to a combination of laxatives, dietary changes, and behavioral modifications. In a smaller subset of pediatric patients experiencing constipation, an organic etiology like anatomic malformation, metabolic derangement, or neuropathy is the cause. Occasionally, abdominal or pelvic tumors will present with constipation [1]. In this paper, we report an unusual case of a lipoblastoma causing constipation.

Case report

A 3-year-old boy was referred to our institution for further evaluation of chronic constipation. Although he passed meconium in the newborn period, his problems with constipation began around 2 weeks of age. Multiple days would pass between bowel movements and he would be fussy and irritable. At first, rectal stimulation with a lubricated applicator was used until polyethylene glycol was started at several months of age, which improved his symptoms until ~18 months of age. At that point, both osmotic and stimulant laxatives did not help. He had a typical pattern of not passing stool for several days, accompanied by abdominal pain and distention, after which he would receive an enema, pass a large hard stool, followed by explosive diarrhea, and then improve for a couple of days until the cycle restarted.

Initial evaluation by an outside specialist included normal abdominal plain films and a normal unprepped barium enema with no transition zone. Laboratory studies including complete blood count, comprehensive metabolic panel, celiac serology, and thyroid function studies were normal. Upper endoscopy was endoscopically and microscopically normal with normal disaccharidases. Flexible sigmoidoscopy was endoscopically normal with rectal mucosal biopsies showing only mild melanosis coli and no other abnormalities. Rectal suction biopsies were obtained but were insufficient to evaluate for ganglion cells. Because of the equivocal rectal biopsy results, the patient was referred to us for anorectal manometry to rule out Hirschsprung's disease.

Because of his young age, anorectal manometry was performed under general anesthesia. While under anesthesia but before starting the procedure, an abdominal exam was performed, which revealed a soft, compressible and poorly demarcated mass palpable in the right lower abdomen. The anorectal manometry was subsequently performed and was normal as a rectoanal inhibitory reflex was demonstrated with insufflation of 50 ml of air in the balloon. A rectoanal inhibitory reflex was also demonstrated in a dose-dependent manner with subsequently smaller volumes of air down to 20 ml.

After the procedure, imaging was obtained to further evaluate the newly discovered mass. An abdominal ultrasound revealed a very large solid-appearing mass in the right hemiabdomen with fatty features. Subsequent computed tomography scan the same day demonstrated a retroperitoneal mass measuring $10.8 \times 14.7 \times 10$ cm and extending from the infrahepatic space to the dome of the bladder (Fig. 1). The mass displaced the large and small bowel to the left and was extremely low density, consistent with a fatty mass. One week later, pediatric general surgery took him to the operating room for an exploratory laparotomy and he underwent an en bloc resection of a very soft, well-encapsulated mass consistent with lipoma (Figs 2 and 3). Pathologic examination revealed mature fat cells separated by thin fibrous septa and a small myxoid focus consistent with retroperitoneal mature lipoblastoma (Fig. 4). Following the surgery, the patient experienced significant improvement in his constipation and is now passing stool regularly without medication.

Discussion

Lipoblastoma is a rare, benign mesenchymal tumor in infants and young children. It consists of proliferating immature fat cells of embryonal origin, and is in the

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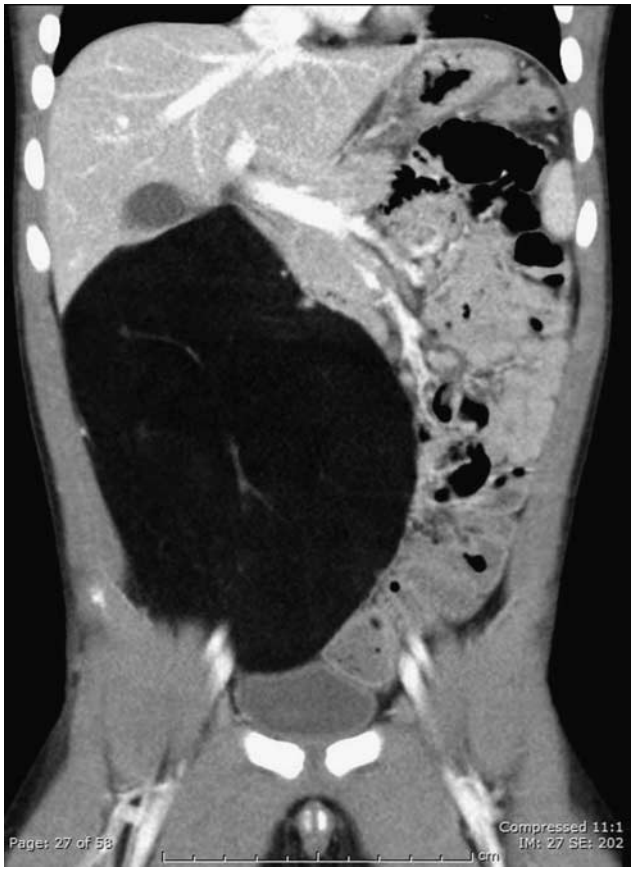
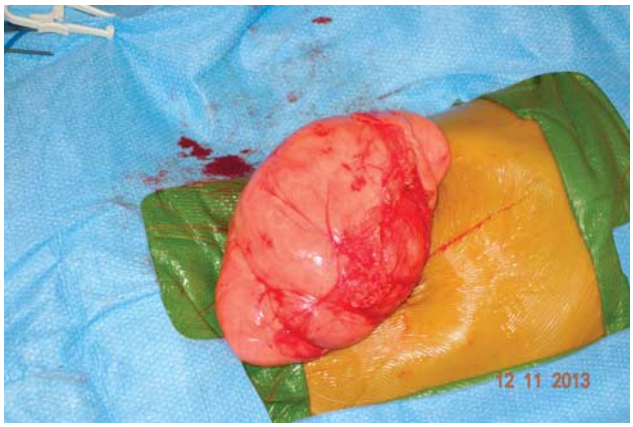
Fig. 1



Coronal image from contrast-enhanced CT scan showing a very large fat-density mass extending from the liver to the bladder and displacing the bowel leftward. CT, computed tomography.

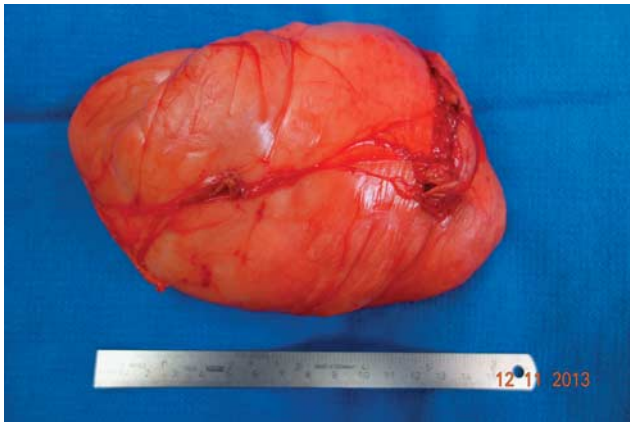
Fig. 2



Intraoperative image showing the smooth, soft, well-encapsulated mass immediately before resection.

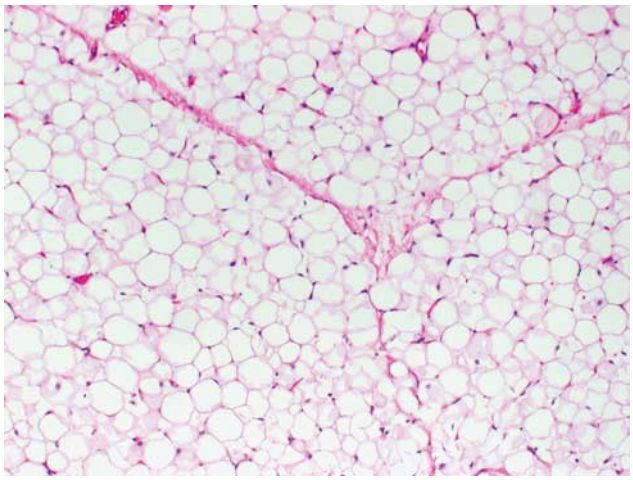
differential diagnosis for other adipose-based masses like lipoma and liposarcoma. It can be either focal or diffuse, in which case it is referred to as lipoblastomatosis, and has a higher chance of local recurrence [2]. Most are diagnosed before 3 years of age [3], with an average age at resection of

Fig. 3



The smooth, soft, well-encapsulated mass immediately following surgical resection.

Fig. 4



Microscopic image demonstrating mature adipocytes separated by thin fibrous septa and a small myxoid focus.

2.8 years [4]. A male : female predominance of $\sim 2:1$ has been reported [5]. In one of the largest case series, 64% were found on the trunk, followed by 27% on the extremities, and 8% in the head and neck [6]. Abdominal lipoblastomas are quite uncommon, generally constituting less than 10% of all reported cases [3]. When present in the abdomen or pelvis, it can present as an incidentally discovered mass during physical exam or with abdominal distention. It presents exceedingly rarely with constipation. Because of their large size, it is not uncommon for the tumor to cause problems from local mass effect. However, they are usually not invasive, although in rare instances they can involve local structures and even the great arteries [7]. Diagnosis is initially suggested by imaging, with computed tomography or MRI providing the best anatomic detail, and then confirmed by pathologic examination of the specimen. Cytogenetic testing is occasionally employed in unclear cases, with abnormalities involving chromosome eight being the most commonly

detected aberrations [6]. Treatment involves complete surgical resection, which can usually be accomplished with minimal complications [8]. Although not malignant or metastatic, lipoblastoma can recur locally, and hence the patient should be closely followed up with repeat imaging.

Conclusion

Our patient is noteworthy in many ways. His lipoblastoma is a rare tumor by itself; its retroperitoneal location is even more uncommon, and its presenting symptom of constipation may be one of the only case of its kind reported. This case also reinforces important principles in the successful management of pediatric constipation. Despite the probability that his constipation was likely functional in etiology, his failure to improve with standard therapies should and did result in further evaluation. In his case, a large mass was detected and subsequently removed, producing improvement in the patient's long-standing symptoms. This case also illustrates the importance of a thorough physical examination in patients presenting with constipation. If a procedure is planned, a meticulous abdominal exam should be performed while under anesthesia immediately before beginning the procedure, as detection of subtle findings can be very challenging in a lively young child. Finally, it reinforces the knowledge that plain radiographs have a limited role in the workup of constipation in children, as radiographs of this patient's abdomen failed to detect the large mass.

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Conflicts of interest

There are no conflicts of interest.

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