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Exophytic focal nodular hyperplasia torsion: A rare cause of sudden-onset epigastric pediatric abdominal pain☆

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ABSTRACT

Torsion of exophytic liver focal nodular hyperplasia (FNH) is a previously undescribed cause of acute abdominal pain in children. We report two cases of torsion of exophytic focal nodular hyperplasia causing acute abdominal pain successfully treated with surgical resection.

Focal nodular hyperplasia (FNH) is the second most common benign tumor in the adult population. It is found most commonly in women of 30–50 years of age [1,2]. In children, it is less common and usually diagnosed between 2 and 5 years of age [3]. Approximately 80% of all FNH cases in children are asymptomatic and require no treatment regardless of the size when the diagnosis is firmly established [4]. We describe two cases of severe acute epigastric abdominal pain in children caused by torsion of exophytic FNH that resolved with resection.

1. Case report

1.1. Case 1

A 7 year-old boy presented with a 5-day history of severe epigastric abdominal pain, fever, and tachycardia. His pain began abruptly and was constant. His fever and tachycardia developed after the first day of pain. His mother was able to describe the exact minute his pain began and he had never had a similar pain before. He was initially seen at an outside hospital where a computed tomography (CT) scan of the abdomen identified a mass (4.8 × 3.0 × 3.6 cm) near the caudate lobe of the liver with mass effect on the lesser curvature of the stomach (Fig. 1).

Given the symptoms and radiographic evidence of mass effect, the patient underwent an exploratory laparotomy. After dividing the gastrocolic ligament and entering the lesser sac, we found a solid mass in the lesser sac pushing anteriorly into the pars flaccida of the lesser curve of the stomach. The mass was attached to the caudate lobe via a pedunculated stalk that had twisted at least 360°. The mass was ischemic and there was surrounding inflammation. We mobilized the mass by dividing the torsed stalk near the caudate lobe.

His postoperative course was uncomplicated. He tolerated a regular diet and his pain resolved. He was discharged to home on postoperative day 2. Pathology confirmed the mass to be infarcted focal nodular hyperplasia (Figs. 2 and 3).

1.2. Case 2

A 13 year-old girl presented with a 3-day history of severe epigastric abdominal pain. Her pain began abruptly and was

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Fig. 1. Coronal CT image through mid-abdomen shows an oval mass between the contrast-filled stomach and left lobe of liver.

constant following onset. She was able to describe the exact minute the pain began and she had never had this pain before. She was initially evaluated at an outside hospital where a CT of the abdomen identified a mass (3.1 × 4.1 × 5.5 cm) near the caudate lobe with mass effect on the lesser curvature of the stomach (Fig. 4). Ultrasound was obtained due to possible cystic component of the mass, and it confirmed a well-circumscribed solid mass adjacent to the left lobe of the liver (Fig. 5). She was referred for evaluation and underwent an exploratory laparotomy.

After dividing the gastrocolic ligament and entering the lesser sac, we found a solid mass in the lesser sac pushing anteriorly into the pars flaccida of the lesser curve of the stomach. The mass was attached to the caudate lobe via a pedunculated stalk that had

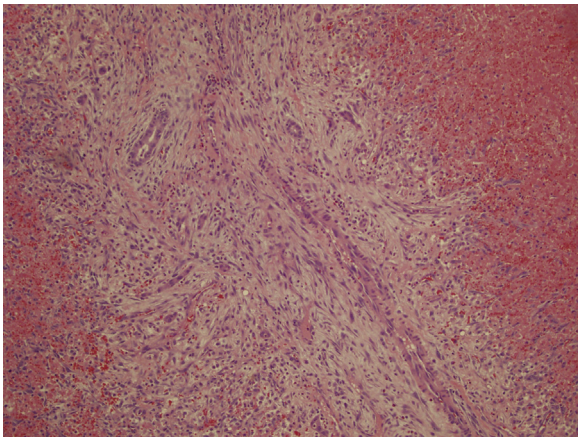

Fig. 2. H&E stain showing fibrotic portal region with abnormally thick artery, occasional bile ducts, and mixed inflammatory infiltrate.

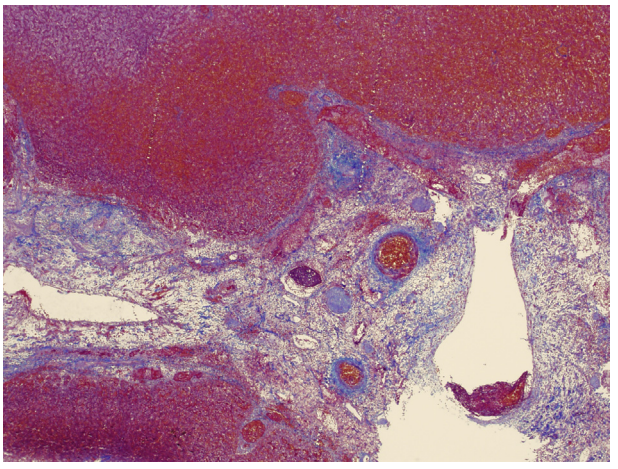

Fig. 3. Trichrome stain showing expended and fibrotic portal region with multiple blood vessels.

twisted at least 360°. The mass was ischemic and there was surrounding inflammation. We mobilized the mass by dividing the torsed stalk near the caudate lobe.

Her postoperative course was uncomplicated. She tolerated a regular diet and her pain resolved. She was discharged to home on postoperative day 2. Pathology confirmed the mass to be infarcted focal nodular hyperplasia (Figs. 6 and 7).

2. Discussion

The management of asymptomatic FNH is usually non-operative with radiologic and clinical monitoring. Symptomatic lesions can be

Fig. 4. Coronal CT image through mid-abdomen shows an oval mass projecting to the left of the left hepatic lobe.

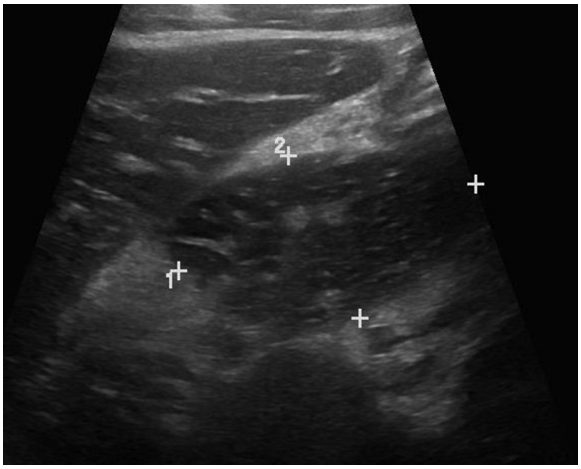

Fig. 5. Sonographic image shows the mass (outlined by cursors). Its appearance by ultrasound is similar to normal liver.

Fig. 6. Gross bisected specimen showing diffuse hemorrhagic necrosis with a central stellate-shaped scar.

either excised or treated with embolization of the feeding vessel. Atypical presentation of FNH may include with exophytic extension causing gastric outlet obstruction [5] or compression of the surrounding vasculature [6]. FNH can also originate from an accessory lobe presenting as colicky abdominal pain and vomiting [7]. In one study, exophytic growth or distortion of the hepatic contour was seen in 32% of the cases [8].

We have identified two cases where torsion of exophytic FNH caused severe acute epigastric pain in pediatric patients. Surgical exploration proved to be diagnostic and therapeutic in both cases. Complete surgical resection appears to be safe and effective. Potentially malignant tumor deterred us from initial diagnostic laparoscopy, but the suitability of the laparoscopic approach has been well described in the literature. Both cases described were followed up with uneventful one-month postoperative visits. Given

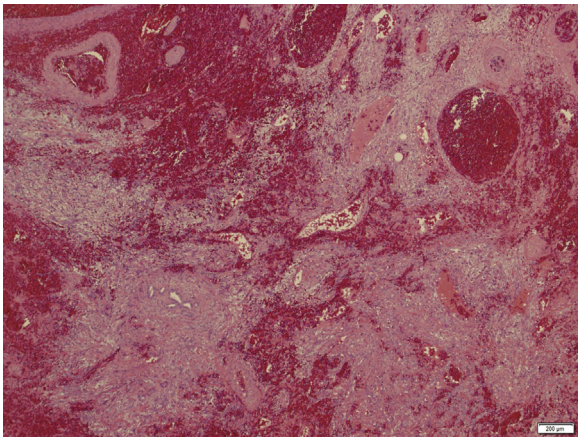

Fig. 7. H&E stain showing irregular fibrous thickening with abnormal blood vessels in a background of hemorrhagic necrosis.

that recurrence after surgical resection has been estimated to be 13% at 29–48 months, we recommend long-term follow up [2].

It is important to pathologically distinguish between FNH and liver cell adenoma since both may have similar clinical presentation. FNH usually has stellate-central scar with bile ducts and central vein, while liver cell adenoma lacks portal triad and central vein.

Our findings highlight potential for exophytic FNH to cause severe acute epigastric abdominal pain when torsion occurs.

Support

None.

Disclosure

The authors have no conflicts of interest to disclose.

References

[1] Wanless IR. Terminology of nodular hepatocellular lesions. Hepatology 1995;22:983–93.
[2] Bonney GK, Gomez D, Al-Mukhtar A, Toogood GJ, Lodge JP, Prasad R. Indication for treatment and long-term outcome of focal nodular hyperplasia. HPB 2007;9:368–72.
[3] Meyers RL. Tumors of the liver in children. Surg Oncol 2007;16:195–203.
[4] Reymond D, Plaschkes J, Luthy AR, Leibundgut K, Hirt A, Wagner HP. Focal nodular hyperplasia of the liver in children: review of follow-up and outcome. J Pediatr Surg 1995;30:1590–3.
[5] Eris C, Yildiz MK, Odabasi M, Akbulut S, Abuoglu H, Ozkan E. Gastric outlet obstruction caused by focal nodular hyperplasia of the liver: a case report and literature review. Int J Surg Case Rep 2013;4:681–3.
[6] Khan MR, Saleem T, Haq TU, Aftab K. Atypical focal nodular hyperplasia of the liver. Hepatobiliary Pancreat Dis Int 2011;10:104–6.
[7] Ruiz Hierro C, Vazquez Rueda F, Vargas Cruz V, Lasso Betancor CE, Ayala Montoro J. Focal nodular hyperplasia on accessory lobe of the liver: preoperative diagnosis and management. J Pediatr Surg 2013;48:251–4.
[8] Brancatelli G, Federle MP, Grazioli L, Blachar A, Peterson MS, Thaete L. Focal nodular hyperplasia: CT findings with emphasis on multiphasic helical CT in 78 patients. Radiology 2001;219:61–8.