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Infantile hemangioma as cause of neonatal gastrointestinal bleeding

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ABSTRACT

Introduction: Gastrointestinal infantile hemangiomas (IHs) are extremely rare and represent a diagnostic challenge, especially in newborns for the limited applicability of diagnostic approaches.

We report two cases of neonates with gastrointestinal bleeding from intestinal hemangioma and a literature review of gastrointestinal IHs.

Case 1: Full-term baby, asymptomatic at birth. On the seventh day of life she experienced failure to thrive, weight loss and hypotonus followed by melena and vomit. The imaging showed an intestinal vascular anomaly. We performed exploratory laparotomy and intestinal resection to remove the lesion. No complication occurred.

Case 2: A 20-day old girl had melena and severe anaemia. The scintigraphic evaluation excluded gastric heterotopia. We opt for explorative surgery and we performed a laparoscopy that showed a congested and oedematous intestinal segment that was resected. Post-operative course was uneventful.

Discussion: Gastrointestinal IHs may cause intractable bleeding, intussusception and intestinal perforations with bad prognosis. A prompt and accurate diagnosis is vital but not always easy. Colour-coded Duplex Scan are a good first-line imaging, MRI and CT give a better definition but the latter exposes to radiation. In the "era" of minimally invasive surgery, laparoscopy can have a role in the management of patients with gastrointestinal IHs.

Conclusions: Gastrointestinal IHs should be considered in the differential diagnosis of neonates with anemia and gastro-intestinal bleeding. In selected cases, the minimally invasive approach permits diagnosis and treatment.

1. Introduction

Infantile hemangiomas (IHs) are common benign vascular tumours affecting 4–10% of infants [1,2]. The cutaneous localized (focal) and the segmental forms account respectively for 67,5% and 13% of cases, whereas the remainder are indeterminate (16,5%) or multifocal (3,6%) [3]. Extracutaneous intestinal manifestations are extremely rare and represent a diagnostic challenge, especially in neonates and small infants for the limited applicability of diagnostic approaches [1,4,5]. We report two cases of neonates with intestinal bleeding that were found to have infantile intestinal hemangioma.

2. Case reports

2.1. Case 1

A Caucasian female was born at 38 weeks of gestational age. The pregnancy was complicated by intrauterine death of one twin. The girl was asymptomatic at birth and discharged home after standard observation period. At 7 days she was admitted to the hospital with failure to thrive, weight loss and hypotonus. Laboratory tests showed high inflammatory makers (reactive C protein and white blood cell count) with negative infection screening. After two days she had liquid malodorous stools, vomit and painful abdomen. Blood samples showed progressive reduction of haemoglobin: after 18 days there was abundant melena.

MRI showed enhancement of part of the small bowel with increased calibre of the superior mesenteric vein, splenic vein and portal vein at the hepatic hilum. CT scan was performed for a better definition: the proximal ileum was involved by a vascular anomaly that leaded to increased venous blood flow through the superior mesenteric and portal veins (Fig. 1).

Systemic corticosteroids were administered but the episodes of melena frequently recurred causing persistent severe anaemia. The decision was to perform explorative laparotomy through a midline incision: the middle ileum had a thickened and red wine wall that extended for about 40 cm. The lesion appeared well marked (Fig. 2a) and was resected leaving a residual normal intestine of 57 cm. A termino-

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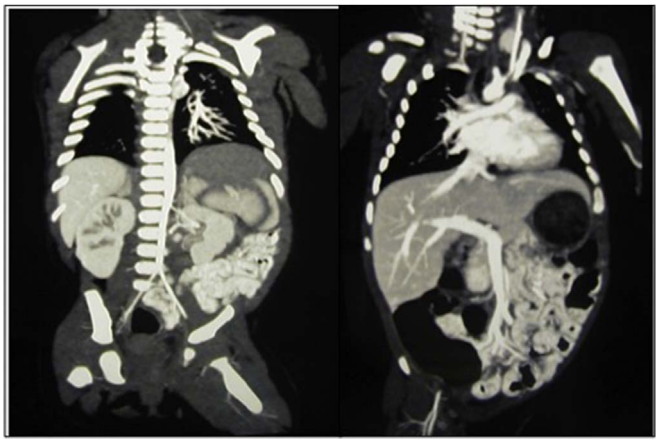

Fig. 1. CT evaluation showing increased venous blood flow through the superior mesenteric and portal veins.

terminal anastomosis was performed after resection. Post-operative course was uneventful.

Feeding stared after 5 days and the girl was discharged after 23 days. Histology permitted to diagnose a benign vascular tumour with features of infantile hemangioma (GLUT1 positive) (Fig. 2b). The girl had a regular follow-up without evidence of disease.

2.2. Case 2

A Caucasian female was born full term after regular pregnancy. Twenty days after birth she presented with melena and severe anemia (Hb 6.4 g/dL) requiring red blood cells transfusion. Screening for infections (CMV, rotavirus, adenovirus, enterovirus) and coproculture were negative. Inflammatory markers were normal. There was no sign of milk protein allergy. A scintigraphy with Tc99m was performed to rule out Meckel diverticulum: the exam showed impregnation of contrast means in the small intestine. Meanwhile melena and anaemia recurred and the girl became instable. We performed a laparoscopic exploration that permitted to identify an intestinal loop with red wine serosa 20 cm far from the ileo-cecal valve (Fig. 3a). The small bowel was exteriorized through the umbilical wound and inspected and palpated outside the abdomen. The affected segment was dilated and edematous and well-demarcated separation (Fig. 3b). We performed an intestinal resection completely removing the lesion and a termino-terminal anastomosis to restore canalization. Histology concluded for infantile hemangioma with GLUT1 positivity. Imaging evaluation performed soon after the operation included abdominal and cerebral ultrasounds: both evaluations were negative.

Two months after surgery a right cervical mass appeared without associated symptoms. Colour-Coded Duplex scan showed a large solid vascular mass with regular margins and deep extension covered by normal skin. The intralesional flow pattern was characterized by high vascularization density and presence of multiple arterio-venous shunts, consistent with the diagnosis of deep infantile hemangioma in proliferative phase.

Total body MRI (Fig. 4) was performed for a better definition of the clinical picture: the cervical mass measured 7 × 5.5 × 7 cm and extended towards the deep cervical sheath and in the opposite side of the body. The abdominal scans showed a hepatic mass (4 × 3,5 cm) in the right lobe, multiple alterations in the mesentery surrounding the vascular branches (9 × 6x5 cm) and a focal diaphragmatic lesion (1,6 × 0,7 × 1 cm). These lesions had rapid and diffuse signal enhancement during the arterial phase, persisting also in late acquisitions.

The anomalies had the characteristics of multifocal infantile hemangioma. Propranolol therapy (2 mg/kg/die) was administered after cardiological evaluation. The response occurred in a few days. After one month there was a huge clinical and US improvement.

3. Discussion

Vascular tumours include IHs, congenital hemangiomas, tufted angioma, pyogenic granuloma, and hemangioendotheliomas (Table 1) [6]. These entities were grouped together by Mulliken and Glowacki because they share the endothelial cell hyperplasia with increased proliferation activity and aberrant blood vessel architecture [7]. In contrast, vascular malformations are not true neoplasms but localized defects of vascular morphogenesis caused by embryogenetic and vasculogenetic dysphunctions.

IHs are the most common tumours of infancy and have a female predominance [1,8]. Their natural history includes a developmental phase between 2 and 8 weeks of life with rapid growth for approximately 6–12 months. Afterwards they undergo slow involution in the first years of life. During the proliferative phase, IHs variably deepen into soft tissues producing superficial, deep and combined forms. IHs may also be localized/focal (67,5% of total IHs, arising from a single focal point), segmental (13%, covering a cutaneous territory derived from a specific neuroectodermal placode), indeterminate (16,5%) and multifocal (3,6%).

The spectrum of manifestations includes the most common cutaneous forms and extracutaneous localizations (e.g. hepatic, gastrointestinal, mediastinum, brain …) [9].

The liver is the second most common localization. Hepatic involvement should be suspected when there are multifocal (> 5 lesions), large (> 5 cm) and segmental cutaneous IHs or in case of hepatomegaly and congestive heart failure [1,10,11].

Gastrointestinal IHs are extremely rare, accounting for only 0,05% of all intestinal neoplasms [8]. They are usually located in the intestinal

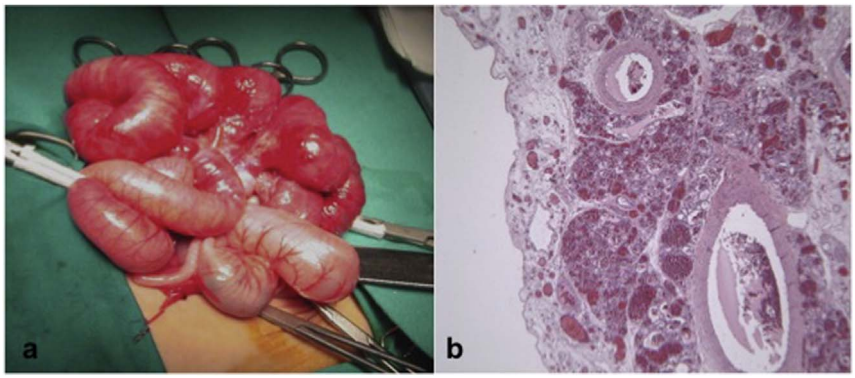

Fig. 2. Intraoperative identification of the affected bowel segment (a) and histological evaluation (b).