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Kaposiform hemangioma of jejunum in a newborn: A case report and review of literature

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ABSTRACT

INTRODUCTION: Kaposiform hemangioendothelioma is a rare tumor, of vascular origin, which predominates in childhood. Although it is frequently a vascular cutaneous anomaly, it is known that some cases can involve bones, mediastinum and retroperitoneum, being even rarer when it occurs in the small intestine, representing only 0.05% of all intestinal neoplasms.
PRESENTATION OF CASE: Female patient, 21 days old, born at 38 weeks, is admitted to the hospital due to bilious vomiting, dehydration and major abdominal distension. An X-ray of the acute abdomen suggests type I jejunal atresia. An exploratory laparotomy was performed, with the presence of a stenotic area in the proximal jejunum, which was chosen for resection of the stenotic area and end-to-end terminal anastomosis, with post-pyloric jejunostomy. The presence of kaposiform hemangioma was confirmed by immunohistochemistry, and the patient had no further complications.
DISCUSSION: The presence of hemangiomas in childhood is frequent, is but it very rare when it appears in the gastrointestinal location, as reported in the present case. A common presentation is intestinal bleeding, being manifested as anemia or, sometimes, acute and potentially fatal anemia. Other forms of rare presentation include intussusception, perforation and obstruction, the latter being out patient's last manifestation. The diagnosis of this alteration can be difficult, and often performed only in the intraoperative period of an exploratory laparotomy.
CONCLUSION: The presence of bilateral vomiting and intestinal obstruction in newborns opens the possibility of a wide range of possible diagnoses, such as the presence of intestinal atresia. Kaposiform intestinal hemangioma is an uncommon cause in these clinical manifestations, but it should be included in the differential diagnosis.

1. Introduction

The presence of bilious vomiting in the newborn is associated with surgical pathologies, most commonly with intestinal atresia [1]. However, some rare differential diagnoses must be remembered [2]. Intestinal kaposiform hemangioma – so far, not described in the literature in children under one month – is a possibility. This article aims to describe the diagnosis, approach and follow-up of neonates with bilious vomiting and suspicion of intestinal atresia, diagnosed and being followed up at the Pediatric Surgery Department of a tertiary academic hospital in Brazil, with histopathological diagnosis confirmed by immunohistochemistry of kaposiform hemangioma in jejunum. It also aims to discuss the scarce existing literature. The work was reported according to the SCARE criteria [10].

2. Presentation of case

Newborn female patient, 38 weeks pregnant, normal labor, APGAR score of 9/10, had prenatal care without complications. There wasn't use of drugs during pregnancy neither family history of genetic diseases. Ultrasonography of nuclear translation and normal morphology shows amniotic fluid index in the last ultrasound within normal limits. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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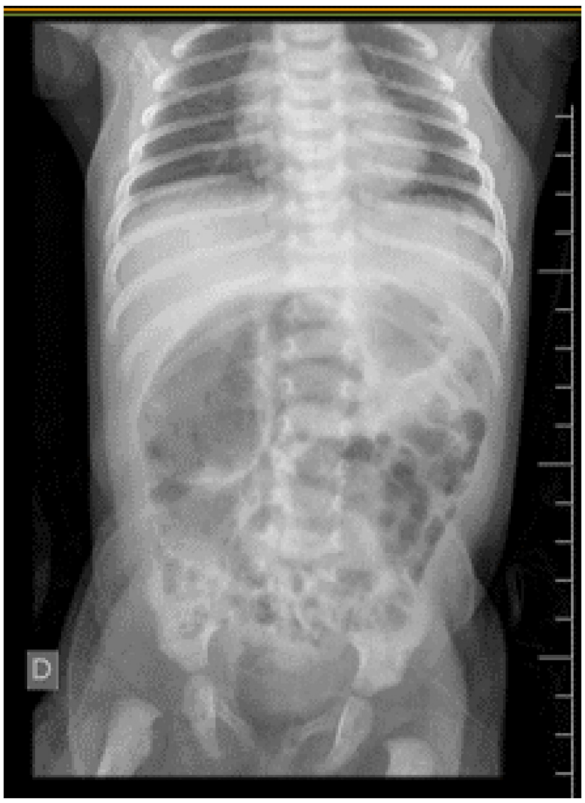

Fig. 1. Simple X-ray of the abdomen, showing a small intestine loop distension.

She presented a physical evaluation in the delivery room without changes, being discharged from the maternity hospital after 48 h, with guidance for usual childcare. However, the mother mentioned that the child had frequent vomiting after being breast-fed, initially milky and then bilious. She performed anti-reflux measures with no success. She received medication for the symptoms, but they continued to worsen. We started investigating the patient when she was 21 days of age and 3 kg. She was dehydrated, icteric, irritated and presented significant abdominal distension.

A nasogastric tube was performed with immediate output of voluminous bilious content. A simple abdominal X-ray (Fig. 1) and contrasted abdominal X-ray (Fig. 2) were performed, suggesting a hypothesis of type I jejunal atrophy (in membrane). After correcting the hydroelectrolytic disorders, the patient underwent a supraumbilical transverse exploratory laparotomy, with an intraoperative procedure of a stenotic area in the proximal jejunum (Figs. 3 and 4), measuring the entire length of the small intestine and colon without other areas of stenosis. It was ressected 7 cm of the proximal jejunum, fifteen centimeters away from the Treitz angle. We also performed a liver inspection without evidence of injuries, a resection of the stenotic area and an end-to-end anastomosis, and a post-pyloric jejunostomy, with a probe allocated after the anastomosis and externalized by an opening in the gastric antrum. The surgery was performed by the pediatric surgery team.

The patient showed good evolution in the postoperative phase with discharge after the complete transition from parenteral to enteral nutrition. In the first outpatient follow-up, 45 days after surgery, we removed the jejunostomy tube due to a complete acceptance of an oral diet and a good gain of weight.

When evaluating the anatomopathological examination, the presence of kaposiform hemangioma was evidenced, confirmed by immunohistochemistry. Slides revealed a lobular proliferation of small vessels, infiltrating the small bowel wall, from submucosa through muscularis propria and subserosa (Fig. 5). The neoplastic vessels were of various sizes and shapes, some slit-like, some glomeruloid, with protruson to some of the larger ones (promontory sign). Endothelium had slight atypia with elongated and slightly enlarged nuclei. Immunostains showed positivity of the neoplastic endothelium to CD31 and CD34 (Fig. 6) and low ki-67 index (Fig. 7). HHV-8 was negative (Fig. 8).

After reviewing the literature, we opted to perform a serial abdominal ultrasound every six months as a follow-up treatment. The patient presented a development and a growth within the expected ones, without occurrence of new injuries per hour. The patient is receiving follow-up every six months with abdominal ultrasound in the last eight months.

3. Discussion

Classically, newborns with bilious vomiting are considered to be potentially surgical and require higher attention. Also, they cannot be discharged until thorough evaluation [1]. Intestinal atresias are mostly suspected in the prenatal period through ultrasound with polyhydramnios due to the non-swallowing of the swallowed amniotic fluid. In some cases and with experienced ultrasound technicians, it is possible to observe fetal gastric dilation. In cases of type I stenosis or atresia, the amniotic fluid may be within normal limits and the symptoms may start later, as in the case reported [2] herein.

The presumed diagnosis is made with a contrast exam showing a failure to fill or a thinning of the intestinal loop, associated with dilation of proximal loops. There are theories to elucidate the etiopathogenesis of intestinal atresia and the most accepted one today is based on the intrauterine ischemic insult, which causes oxygenation failure, ischemia and reabsorption of the affected segment [3].

Differential diagnoses include the duplication cyst, the mesentery cyst and the extrinsic tumors, which can cause compression.

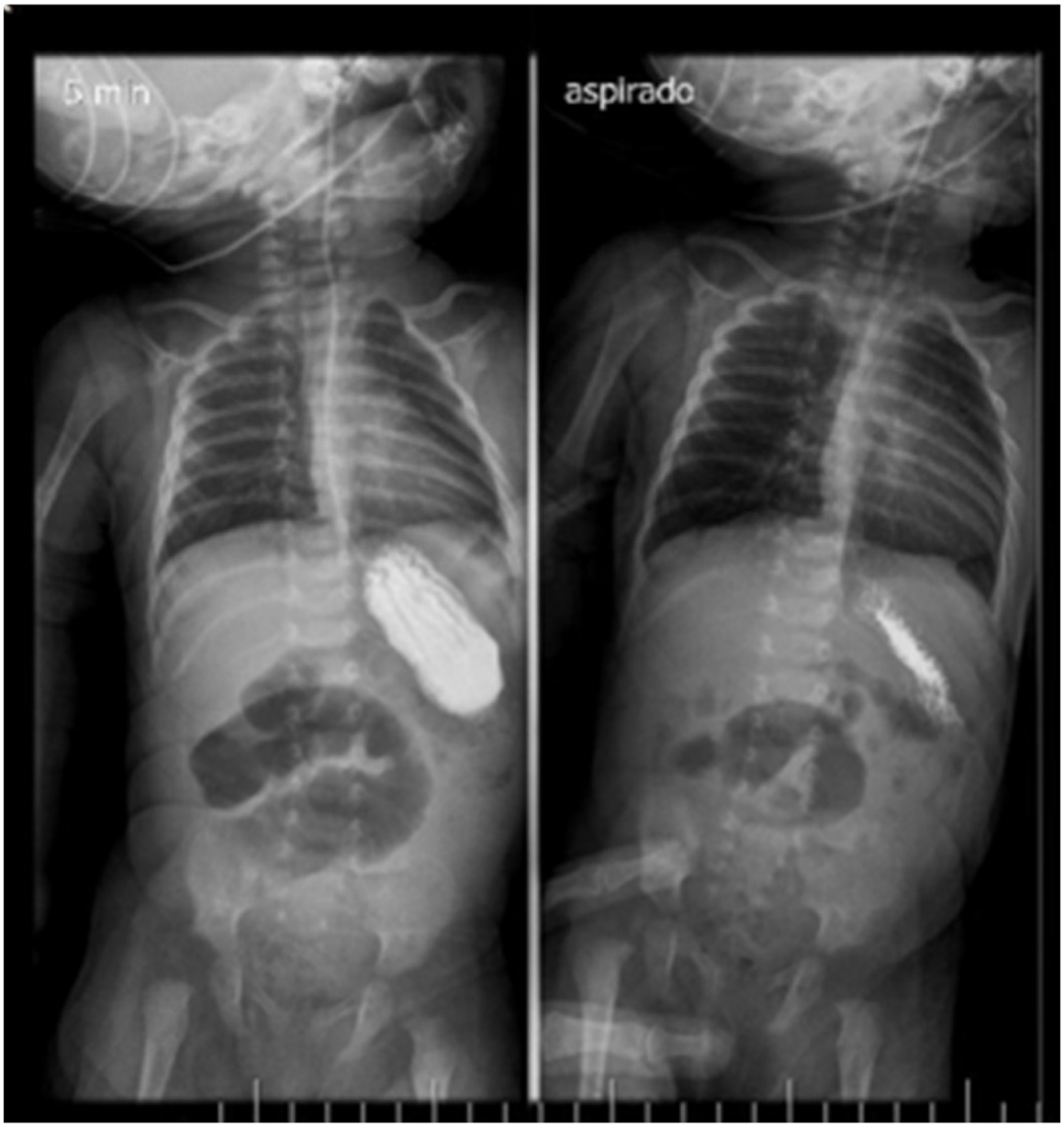

Fig. 2. Contrasted X-ray showing no contrast progress.

More rare and little described in the literature is the possibility of intestinal kaposiform hemangioma, which motivated the report of the case described [4].

Hemangiomas are congenital vascular tumors with a prevalence of 2–6% in the general population, being the most common tumors in childhood [5]. Kaposiform hemangioendothelioma is a rare tumor of vascular origin that predominates in childhood. It has benign histological characteristics, but it presents malignant behavior with local proliferation and, frequently, vascular aggression. The condition involves the skin, subcutaneous tissue and deeper tissues, reaching retroperitoneum in some cases [5].

The diagnosis made according to the described herein is even rarer. Since it is a tumor derived from vascular malformation, many patients present other lesions, mainly cutaneous, what facilitates the diagnosis. These lesions can be localized or dispersed and can also be completely asymptomatic or cause occult gastrointestinal/obscure bleeding [6]. In a recent study, there were four cases of patients with intestinal hemangioendothelioma. They all started the symptoms with hematochezia, enterorrhagia and ascites, but none of them presented a kaposiform form. In neither case was there a relationship of subocclusion or suspicion of intestinal atresia [7].

Visceral kaposiform hemangioendothelioma is an aggressive vascular tumor, and less than 100 cases were reported in the literature since its description in 1993 [6]. It has this name because it exhibits the characteristics of capillary hemangioma and Kaposi's sarcoma. It presents a better prognosis if it is limited to the intestine and not related to coagulopathies. It is characterized according to the malignancy as borderline, due to its rapid growth and invasion capacity [4].

The tumor has benign histological characteristics, but it leads to high mortality in the absence of treatment. It generally affects children under the age of two and has an increased incidence in girls. It progresses as a single lesion on the skin, and it is more common on the trunk or extremities. Tumors grow up to 18 months of life [6].

Intestinal placement is extremely rare and was first reported in 2012. An article describes a case that is similar to the one reported

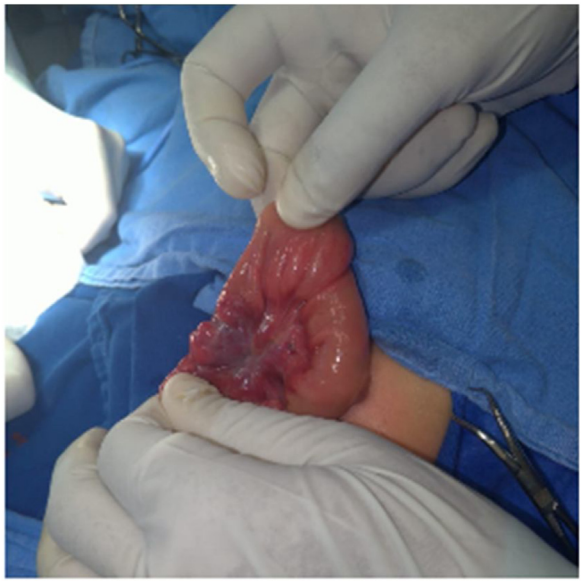

Fig. 3. Intraoperative finding of a stenotic area with a purplish hue.

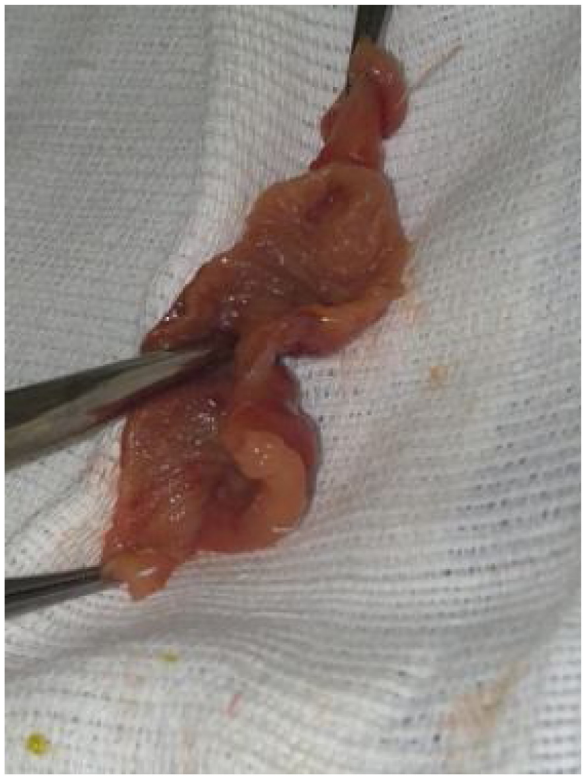

Fig. 4. Picture of the resected area.

herein, in which the patient had abdominal distension and bilious vomiting. However, at 1 month old and presenting a vascular mass in the inoperative, it is the fifth reported case and the youngest patient so far [4]. Other forms of rare presentation include intussusception, obstruction and perforation [8]. Ileal perforation is a rare complication in neonates and, to date, only one case is related in the literature, by McGaughey et al., who discovered evidences of ileal perforation due to intestinal hemangioma while operating on a newborn with intestinal obstruction [8].

After the report made by Zuckemberg et al., about 183 cases of kaposiform hemangioendothelioma were described, mostly in the

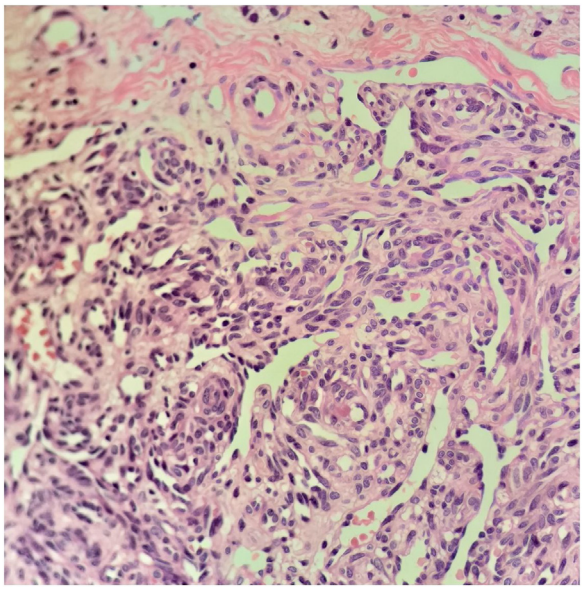

Fig. 5. Presence of kaposiform hemangioma.

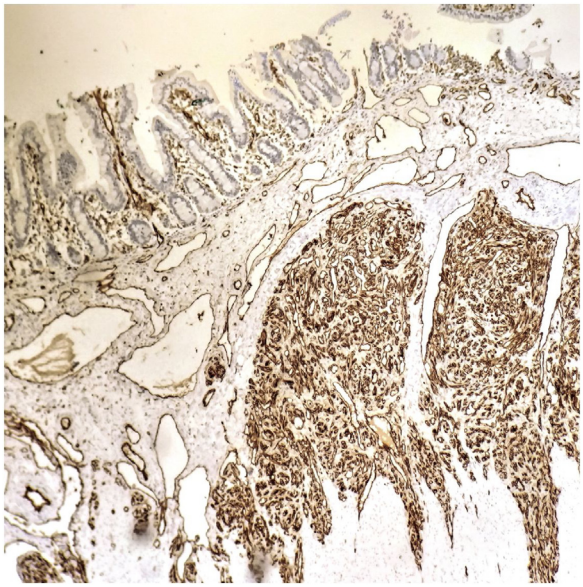

Fig. 6. The CD34 was positive.

cutaneous form, followed by retroperitoneal and mediastinal types. The main symptom presented was vomiting and abdominal distention, just as in the case reported herein, differing from other types of intestinal hemangiomas. Despite the aggressiveness described, all the reported cases presented survival after the complete resection of the lesion. In one of the selected cases, where there was a diffused involvement of the gastrointestinal tract, the use of vincristine and propranolol associated with surgical treatment caused the remission of the symptoms of anemia and abdominal pain [2,6].

Although several causes of neonatal intestinal perforation – including the rare intestinal hemangioma – have been reported, the diagnosis can be difficult and an exploratory laparotomy has often been used as the final diagnostic tool [8]. Thus, the best therapy is a surgical excision with injury margins, if possible. The prognosis of the lesion takes into account its location and the possibility of resection and association with Kasabach-Merrit syndrome. No case described in the literature has presented an association with such syndrome [4,9].

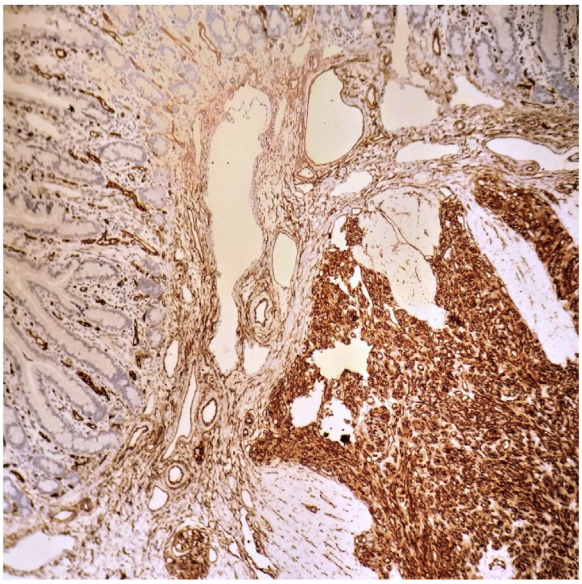

Fig. 7. The CD34 was positive.

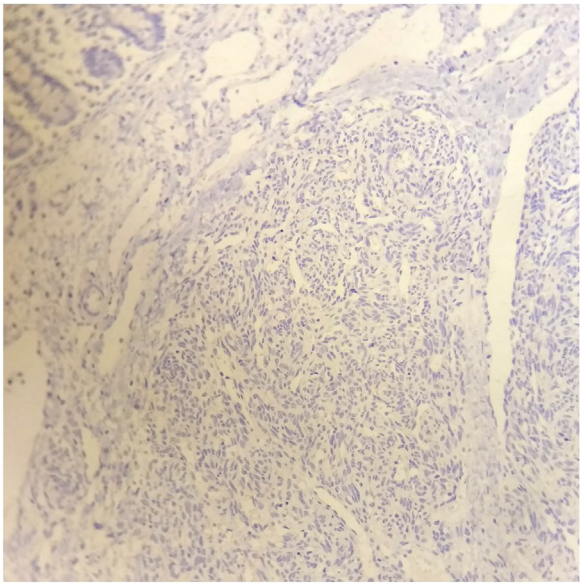

Fig. 8. The HHV-8 was negative.

4. Conclusion

Therefore, we can conclude that the kaposiform intestinal hemangioma can affect neonates, and this case was described in the literature for the first time. Also, it is possible to make a differential diagnosis with congenital intestinal stenosis and membrane intestinal atresia. More cases should be reported when seeking an excellence in diagnosis and therapy; however, the resection with free margins is associated with a better prognosis.

Declaration of Competing Interest

None of the authors have a conflict of interest.

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Ethical approval

The study was accepted by the ethics committee at the hospital (Clinical Hospital, Curitiba, Brazil) where the patient had the surgery and has been following up since then. The reference number is 36943220.3.0000.009. We wrote in the article that it was accepted by ethics commitee.

Any images/figures/photos are suitably anonymised with no patient information or means of identifying the patient.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Isabela Picolotto Moraes: study concept and writing the paper.

Oona Tomiê Daronch (corresponding author): writing the paper.

Camila Girardi Fachin: correcting the paper, follow-up of the patient.

Luiz Paulo Junqueira Rigolon: study concept and writing the paper.

André Ivan Bradley dos Santos: correcting the paper, chief of surgery of the case.

Registration of research studies

1. Name of the registry: KAPOSIFORM HEMANGIOMA OF JEJUNO IN NEWBORN: A CASE REPORT AND REVIEW OF LITERATURE.
2. Unique identifying number or registration ID: not a “first in man” case report.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not a “first in man” case report.

Guarantor

Oona Tomiê Daronch (corresponding author and the Guarantor).

Provenance and peer review

Not commissioned, externally peer-reviewed.

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