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Hepatoblastoma: A success report using a combination therapy with preoperative arterial embolization☆

Hepatoblastoma: éxito de terapia combinada con embolización arterial preoperatoria

Hepatoblastoma is the most common hepatic tumour in children, accounting for 90% of all malignant liver tumours. It is usually diagnosed during the first 3 years of life.[1,2] Signs and symptoms are very subtle, with the main clinical manifestations including abdominal distension and a palpable abdominal mass. For diagnosis, alpha-fetoprotein (AFP) levels, compatible imaging studies and suggestive signs and symptoms are required; however, histological diagnosis of the tumour also plays an important role.[1] Survival rates of these patients have improved significantly over the last 3 decades, currently achieving rates of around 75–80%. This improvement is due to advances in therapy.[2]

Our case study involves a 2-year-old child with no personal or family history of disease, good overall health and no previous symptoms. The child visited the paediatric emergency department with symptoms of bronchiolitis. Upon physical examination, hepatomegaly was detected with no other findings. As a result, an abdominal ultrasound (hepatomegaly) and AFP tests (152,370 ng/ml) were performed. Since a liver tumour was suspected, the child was referred to the paediatric oncology clinic where he underwent imaging studies (Fig. 1: bulky tumour measuring 11 cm long × 6 cm thick, confined to the right lobe of the liver, without exceeding beyond the round ligament, with no portal vein thrombosis, but with compression/encasement of the hepatic hilar vessels) and a liver biopsy, which confirmed the diagnosis of foetal hepatoblastoma. Taking into consideration the *Pretreatment Extent of Disease* (PRETEXT) staging and risk stratification system proposed by the *Société Internationale d'Oncologie Pédiatrique-Epithelial Liver Tumor Study Group* (SIOPEL), the tumour was classified as PRETEXT III, considered inoperable, and therefore treatment was started with cisplatin.[3] The subsequent decision to start more aggressive chemotherapy with cisplatin, carboplatin and doxorubicin was due to suspected encasement of the hepatic hilar vessels. After 3 cycles of chemotherapy, an MR angiogram was performed which revealed overall

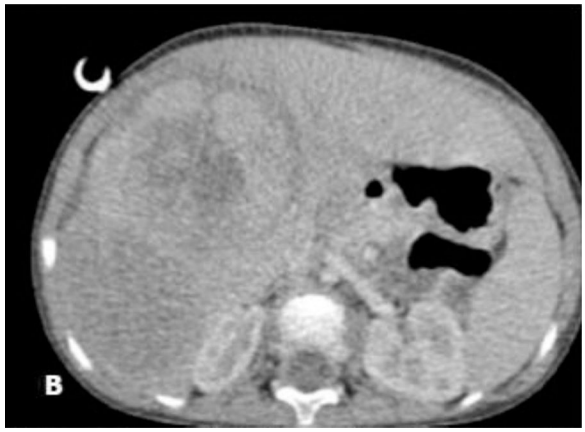

Figure 1 CT image of the abdomen at diagnosis.

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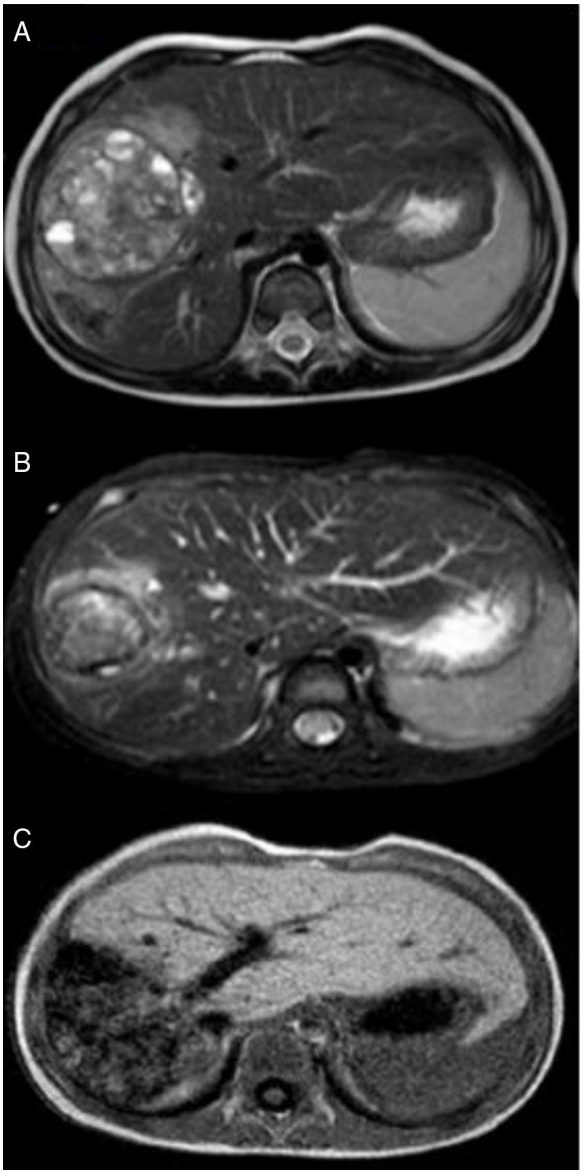

Figure 2 MR angiogram images of the abdomen after 3 cycles of chemotherapy (A), embolisation (B) and surgery (C).

reduction of the tumour mass, consisting of 3 adjacent nodules: the most central region had a transverse and anteroposterior diameter of 5.4 cm and a craniocaudal diameter of 6.7 cm; the posterior region showed a mass with a transverse diameter of 4.2 cm and the most anterior region showed a third tumour component measuring around 2 cm, with no evidence of portal vein thrombosis and with questionable vascular encasement (Fig. 2A Fig. 2). At this point, the decision was made to combine chemotherapy with embolisation, given the hypervascular nature of the tumour, in order to reduce the tumour mass and due to the risk of extensive surgery (trisegmentectomy). Therefore, selective embolisation was performed with polyvinyl alcohol (PVA) microspheres and metal coils. A hepatic angiography revealed a large, hypervascular mass measuring approximately 7–8 cm in liver segments V, VI and VIII. Microspheres were selectively released into the respective nutritive arterioles of the tumour and *microcoils* were released into the main nutritive arteries. Another MT angiogram was performed once month later, which revealed a significant reduction in tumour size, with reductions of 7 mm in the central component, 4 mm in the anterior component and 7 mm in the most posterior component (Fig. 2B). This reduction in tumour size allowed for right hepatectomy, with a resection plane running along the middle hepatic vein, corresponding to the usual limit for this type of resection (Fig. 2C). The anatomical pathology description describes a narrow hepatic resection margin of 0.5 cm for the resected specimen. The child completed the cycle of chemotherapy 2 months after surgery, obtaining a favourable response with a gradual decrease in AFP levels, which are currently normal.

The treatment for hepatoblastomas has advanced in recent years, resulting in better survival rates. Although treatment depends on the stage of the tumour, complete surgical resection (macroscopic and microscopic) is essential to achieve a cure.[1,2] However, surgery is not possible in 50% of patients due to the advanced stage of the tumour.[4] Since hepatoblastoma is more sensitive to chemotherapy, pre-operative chemotherapy helps reduce tumour volume, making resection easier, which results in a better prognosis.[1,2] Unresectable, high-risk hepatoblastomas can be made resectable with chemotherapy; however, this is not always effective.[4] Since they are usually hypervascular tumours, pre-operative embolisation may be effective in cases that do not respond to chemotherapy.[4,5] In this child, embolisation produced satisfactory results since a significant reduction in tumour size was achieved, allowing complete surgical resection. Although pre-operative embolisation is a viable and attractive outcome, it must be analysed with caution since it may produce some complications.[6]

Despite the promising results of embolisation in cases of hepatoblastoma, prospective studies must be conducted in order to evaluate the risk-benefit of this approach before it can be considered a potential first-line treatment.

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Ischemic enteritis with unusual presentation

Enteritis isquémica con presentación inusual

The authors present the case of an 83-year-old woman submitted a month before to a total gastrectomy for gastric adenocarcinoma who admitted to the emergency department with hematemesis. After resuscitation and hemodynamic stabilization an upper endoscopy was performed that showed a regular esophagojejunal anastomosis in healing process; however, in the efferent jejunal loop the mucosa appeared edematous, of purplish appearance, with superficial ulceration and exudate, and friability with spontaneous bleeding (Fig. 1). Biopsies were performed. With the suspicion of ischemic enteritis a CT angiography was performed that revealed a conglomerate of jejunal loops with wall thickening and increased uptake of the mucosa, with tightness and twist of the mesenteric vessels (Fig. 2). The arterial vessels seemed patent, however the mesenteric veins had become globular with aspects suggestive of an ischemic process by venous congestion. The patient then underwent laparotomy where was noted migration/herniation of entero-enteric anastomosis of the ''Y-en-Roux'' to the supramesocolic floor through the mesocolon, confirming the presence of mesenteric torsion and venous congestion conditioning edema and endoluminal bleeding. The ischemic segment was resected and new anastomosis were performed. The patient was discharged after 14 days under oral nutrition. Later, histology evaluation confirmed the diagnosis of ischemic enteritis (Fig. 3).

In the literature there are rare cases of ischemic enteritis, particularly non-duodenal ischemia, documented by endoscopy, since most of the cases are diagnosed by imaging, and especially surgery.[1] Ischemic enteritis can be classified as either occlusive or nonocclusive. Embolism and

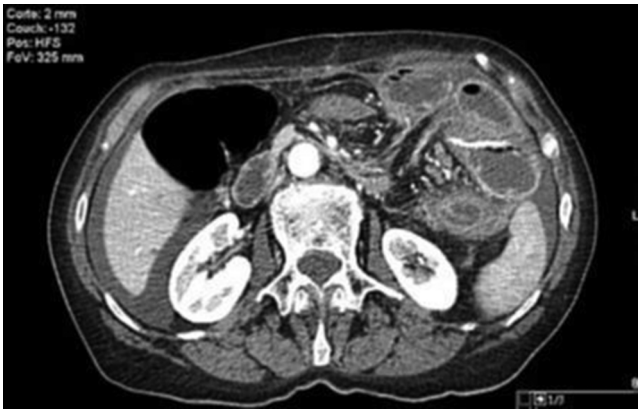

Figure 2 CT angiography with globular mesenteric veins with aspects suggestive of an ischemic process by venous congestion.

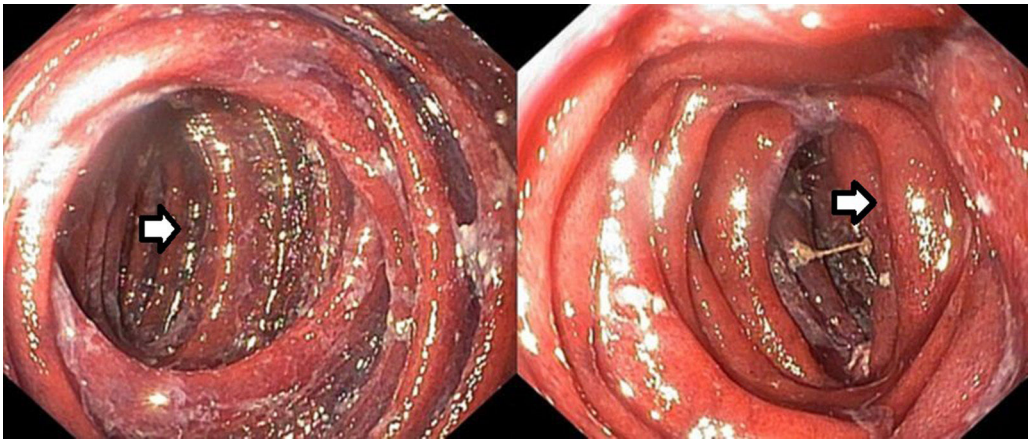

Figure 1 Efferent jejunal loop with edematous and congestive mucosa, friability and purplish appearance, findings suggestive of an ischemic process.