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Hepatoblastoma of the adult with pericardial metastasis: A case report



Andrea Celotti*, Gian Luca Baiocchi, Marco Ceresoli, Michele Bartoli, Silvia Ulinici, Nazario Portolani

Department of Medical and Surgical Sciences, Surgical Clinic, University of Brescia, 25123 Brescia, Italy

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ABSTRACT

BACKGROUND: Hepatoblastoma is the most frequent liver tumor in children, but very rare in the adult and associated with an unfavorable prognosis. The diagnosis is always postoperative or post mortem and biopsy is not useful. Surgery is the only accepted treatment.

CASE PRESENTATION: Our patient underwent surgery in the suspect of liver metastasis from a previous gastric cancer. Surgery consisted in left lobectomy with partial diaphragm resection and partial pericardiectomy for a pericardial lesion, found after the opening of the thorax. The diaphragm defect was corrected with a biological mesh.

RESULTS: The histopathological examination indicated hepatoblastoma of the adult with pericardial metastases. The patient was asymptomatic and without recurrence after 21 months of follow up.

CONCLUSION: The hepatoblastoma of the adult is related to a poor prognosis with median survival time less than 5 months. Surgery is the only curative treatment, but in many cases tumor resection requires complex operations. Vascular and thoracic expertise could be useful in the management of hepatoblastoma.

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1. Introduction

Hepatoblastoma (HB) is a rare tumor, but the most frequent liver tumor in childhood.

Its usually occurs in infants under 3 years of age and the 5 years overall survival rate in children with non disseminated tumors is close to 70% [1].

Hepatoblastoma in adult is very rare, with an unfavorable prognosis [2]. In literature about sixty cases have been described. In 1958 Barnett et al. [3] reported the first case in a young man, who died after one month from the diagnosis due to a complete liver replace by the tumor spreading. The prognosis of HB in the adult is extremely poor, because most cases are unresectable at the diagnosis and non-surgical treatments have only a palliative role [4].

This report describes a case of a 68 year-old man who underwent total gastrectomy for gastric cancer. Subsequently, the patient underwent left liver lobectomy after 1 year for suspected liver metastases, but the definitive histological examination showed hepatoblastoma.

Abbreviations: HB, hepato blastoma; MRI, magnetic resonance imaging; HCV, hepatitis C virus; HBV, hepatitis B virus; GOT, glutamic oxaloacetic transaminase; GPT, glutamic pyruvic transaminase; AFP, alpha-fetoprotein; PO, post operative.

* Corresponding author at: Department of Medical and Surgical Sciences, Surgical Clinic, University of Brescia, III Chirurgia, Spedali Civili di Brescia, P.le Spedali Civili 1, 25123 Brescia, Italy. Fax: +39 030396011.

E-mail address: cei185@hotmail.com (A. Celotti).

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2. Case report

This is the case of a 68 year-old man who underwent, in January 2012 at another Institution, total gastrectomy with Roux-en-Y esophagojejunostomy with D2 lymphadenectomy for gastric cancer. Histological examination showed gastric adenocarcinoma, intestinal type, with low differentiation (G3), with metastasis to the regional nodes (pT3N3M0).

Patient received adjuvant chemotherapy with 10 cycles of FOLFOX. In December 2012, during the oncological follow-up, a suspected 2 cm liver metastases was discovered. For this reason, the patient started 8 cycles of FOLFIRI. In October 2013, new MRI revealed an increased liver metastases up to 5 cm (Fig. 1). No other cancer localizations were found. Patient was referred to our hospital. Blood examinations revealed negative serology for HCV and HBV, normal levels of GOT, GPT, bilirubin, albumin. AFP level was 1231 UI/mL. He had no history of alcohol intake. MRI was re-evaluated by hepato-biliary dedicated-radiologists and the malignant nature of the lesion, with the features of metastases was confirmed. In November 2013 the patient underwent surgery. Peritoneal exploration revealed a large solid lesion of the second liver segment, completely exophytic, with diaphragm infiltration (Fig. 2). Intraoperative liver ultrasound was negative for occult liver metastases and no peritoneal carcinomatosis or other cancer localizations were found. The operation consisted of left lobectomy and diaphragm resection.

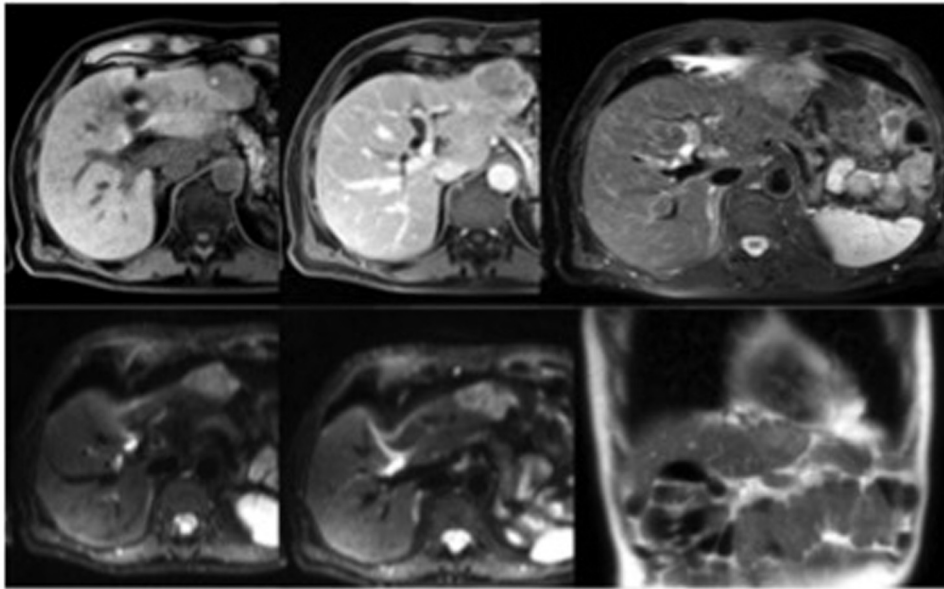


Fig. 1. RMN imaging: typical radiological aspect of a liver metastasis. No other localizations are evident in the RMN.

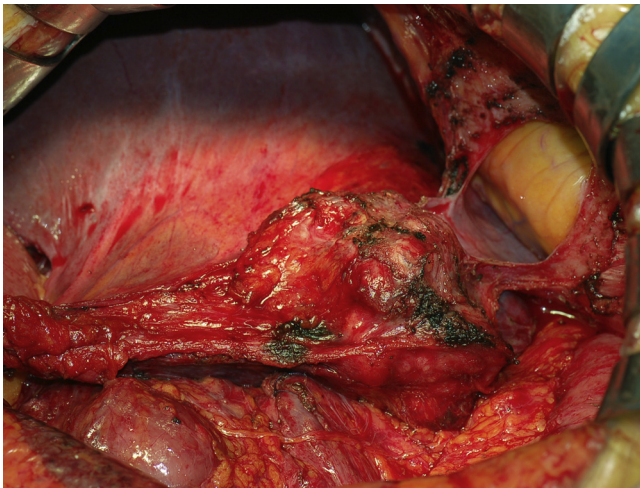


Fig. 2. Exophytic lesion of the II segment with diaphragm infiltration.

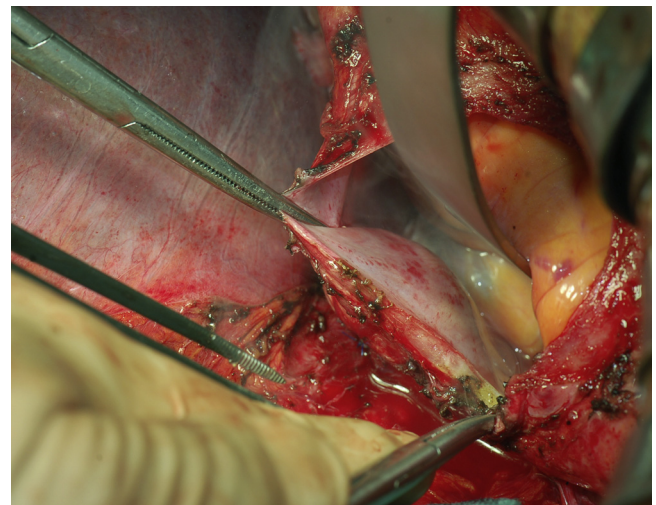


Fig. 3. Pericardial nodule. The Harrington retractor holds the heart to permit the exposure of the inferior vena cava.

The first step was the incision of the diaphragm surrounding the liver lesion, until a complete detachment of the tumor en-bloc with a piece of diaphragm. During the surgery, through the diaphragmatic defect, a pericardial nodule of about 4 cm was discovered along the infero-lateral cardiac margin (Fig. 3).

Although the presence of a second neoplastic localization, a R0 surgery was considered feasible. Inferior vena cava was isolated until right atrium and an extended pericardiectomy was performed, from the upper right atrium to the inferior cardiac margin. To prevent cardiac tamponade, the parietal pleura was sutured to the pericardial margin. Finally left liver lobectomy was completed. The defect of the diaphragm was repaired using a 10 × 10 cm of bovine pericardium (Fig. 4), sutured with Ethylene Terephthalate. Abdominal and thoracic drains were placed. Postoperative course was uneventful and drains were removed on 7th postoperative day (PO). The patient was discharged on 13th PO, without problems. AFP level was remarkably decreased at 290 UI/mL at the discharge.

The macroscopic pathology report showed a 6 × 4 cm liver mass and a 3.5 × 3 cm pericardial node with the same histological features. Both lesions were composed by double cell-populations: columnar and vacuolated cells and pleomorphic cells. Extensive

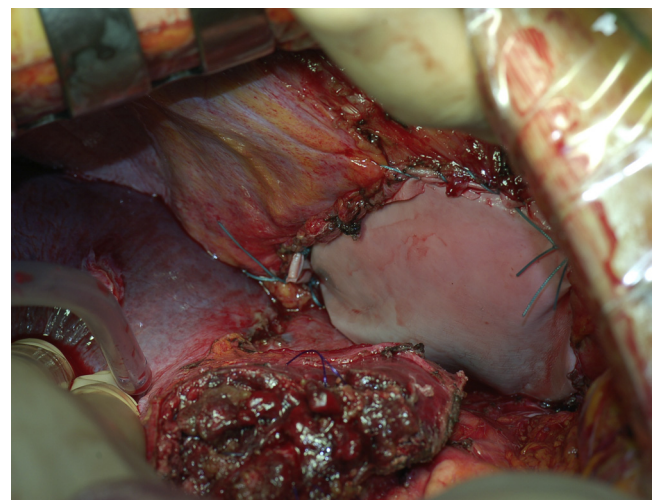


Fig. 4. Bovine pericardium patch to repair diaphragm defect.

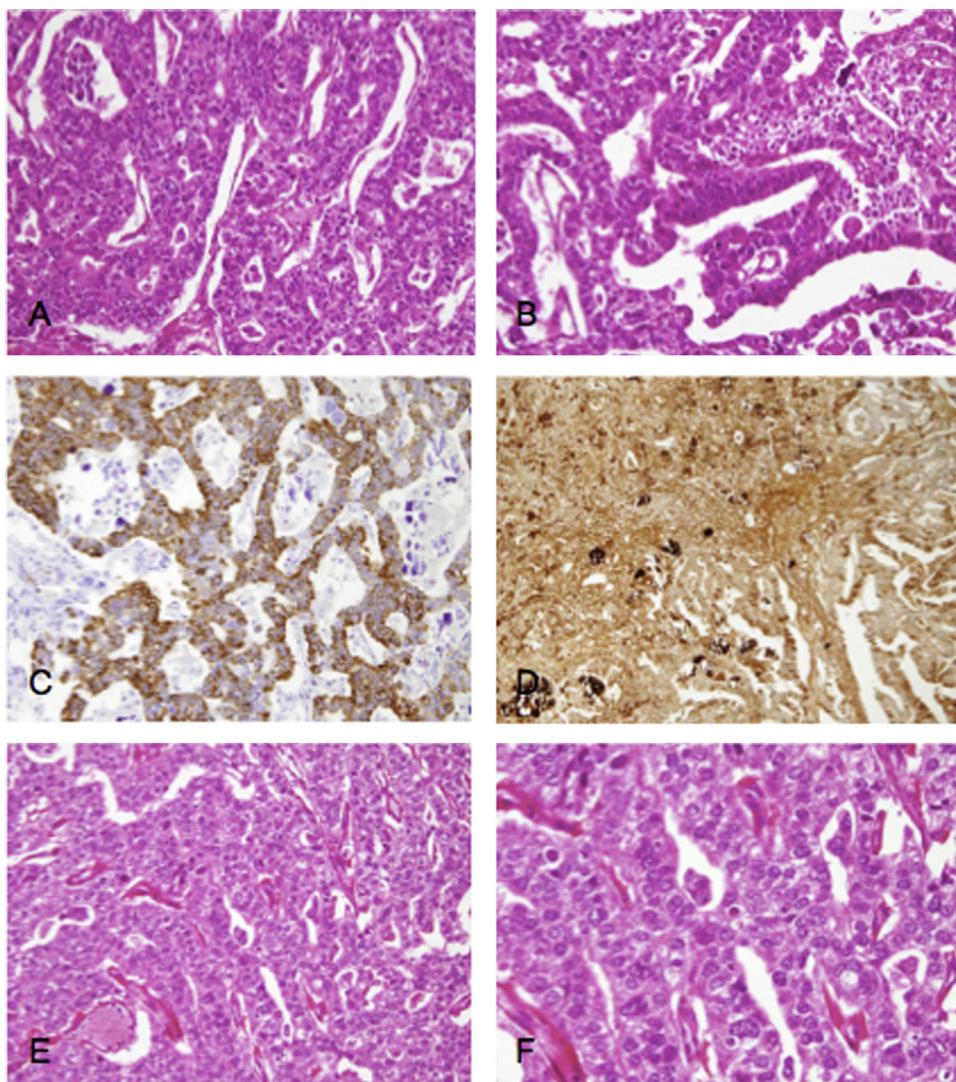


Fig. 5. (A, B) Hematoxylin eosin of the liver tumor; (C, D) anti-hepatocytic antigen and alphafetoprotein positivity respectively of the liver tumor; (E, F) hematoxylin eosin pericardial metastasis.

necrosis and calcific areas were found. Immunohistochemistry was positive for: anti-hepatocytic antigen, alphafetoprotein, vimentin, CD10 and cytokeratin 8 e 19 and negative for cytokeratin 7, cytokeratin 20, CDX-2, PLAP, beta HCG, OCT4, TCL1, CD30, calretinin. Final diagnosis was adult hepatoblastoma with pericardial metastasis (Fig. 5). We discussed the case with the oncologists and no adjuvant therapy was started. After 21 months patient is disease-free and asymptomatic.

3. Discussion

Hepatoblastoma is the most common liver tumor in children, and occurs especially in patients under 5 years of age [1].

Nowadays in children, the advances in chemotherapy, especially to allow complete surgical resection even in advanced tumors, have increased the overall survival from 30% to over 80% [5]. In adult, HB is very rare and in literature only 62 cases are described and presents an extremely poor prognosis.

Patients with HB usually have right upper quadrant pain, often in presence of abdominal mass. In some cases acute presentation with hemoperitoneum is reported [6–8].

Imaging is not useful for the diagnosis, because HB presents similar characteristics to others common liver tumors such as hepatocarcinoma or metastases [8].

In this case MRI was typical for a malignant lesion with the features of a secondary liver tumor, especially the presence of the T1-hyperintensity peripheral ring was strongly suspected for secondarism. Blood examination of the patient revealed an AFP level of 1231 UI/mL, without liver disease and he presented a history of gastric cancer. This data could be related to the previous oncologic history, because in literature are reported cases of gastric cancer alpha-feto producing [9–11]. Anyway Yamazaki et al. [12] declared that AFP levels were often elevated in HB, so it could be used as a serological marker like for the hepatocarcinoma.

Considering its rarity, the lack of particular radiological features and the uncertain role of AFP, the preoperative diagnosis of HB in the adult is quite impossible without surgery or biopsy.

Anyway the use of the biopsy is debatable in literature, because in the presence of a potentially resectable liver malignancy, surgery should be performed and the postoperative diagnosis considered adequate. Mondragon et al. [13] reported a case of a patient died after percutaneous biopsy for multifocal HB.

The core treatment of this tumor is surgery as in pediatric patients. In children surgery is usually performed after pre

operative chemotherapy, but in the adult there is no standardized management of HB. In many cases, in order to achieve an R0 resection, major hepatectomies or en-bloc resections of adjacent organs such as diaphragm, stomach, esophagus, pericardium, or spleen are needed. Anderson et al. [14] reported a case of an extensive resection of the left liver lobe, with demolition of left diaphragm, partial pericardiectomy, distal esophagectomy, partial gastrectomy, splenectomy and omentectomy. Regarding the chemotherapy treatment, there is no agreement in literature on its use. In the review of Yamazaki et al. [12], no response to chemotherapy was observed in any case and the author concluded that an excision of the tumor should be considered, without preoperative chemotherapy and tumor puncture.

Instead Nakamura et al. [4] reported a case of a patient with a 4 years survival after multimodal treatment with neoadjuvant chemotherapy, liver resection and postoperative chemotherapy.

HB presents a poor prognosis in the adult due to its metastatic behavior or the local aggressiveness. In the review of Wang et al. the median survival time for 27 patients with available follow up was 4 months, with 1-year survival of 29.6% [2]. Rougemont et al. [6] reported 34 patients with a median survival time of 2 months and with a 1-year survival of 24%. Like in our case, in literature are reported just two cases of diaphragm resection due to the invasion of the tumor, but no informations about reconstruction are discussed [14,15].

In this case the diaphragm defect was too bigger to allow a direct closure, so a biological mesh of bovine pericardium was used. This method is generally adopted, in our Division, in the cases of large hepatocarcinomas of the posterior segments with good results.

In literature the use of bovine pericardial mesh seem to have good outcome in terms of biocompatibility, high-tensile strength and easy use, so it could be recommended in many cases of diaphragm reconstruction [16–18].

In the case of our patient the use of this material was particularly appropriate because after pericardiectomy, the heart would be directly into contact of a soft biological mesh, but with a high-tensile strength. In this case, the pericardial metastasectomy allowed a curative surgery. The patient, after 21 months of follow up, was asymptomatic and without recurrence. This is the case of metastatic hepatoblastoma with the longer disease-free survival in the literature.

4. Conclusion

The prognosis of the hepatoblastoma in the adult is unfavorable with median survival time less than 5 months. Many HB are unresectables at diagnosis, because of local aggressive behavior or metastatic spread. Surgery represents the gold standard treatment, but in many cases tumor resection requires complex operations. Vascular and thoracic expertise could be useful in the management of hepatoblastoma.

Conflict of interest

The authors declare no potential conflicts of interest relevant to this article.

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Ethical approval

This is not a research study.

Consent

We obtained a written and signed consent to publish this case report.

Author contribution

All the Authors made substantial contributions to conception and design of the study, Celotti A. and Ceresoli M. conceived the study and wrote the paper; Bartoli M. and Ulinici S. performed Literature search; Baiocchi G.L. and Portolani N. reviewed the paper.

Guarantor

Andrea Celotti.

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