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Rhabdomyosarcoma of the Bile Ducts in Children Simulating Congenital Dilatation of the Biliary Tract

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Abstract **Case Report**

Introduction: Embryonal rhabdomyosarcoma of the biliary tree is extremely rare. It accounts for 1% of all rhabdomyosarcomas in children and 0.04% of paediatric cancers. It is often of the botryoid type and localised in the main bile duct or intrahepatic ducts. Localisation in the extrahepatic bile ducts is a rare cause of jaundice in the paediatric population. ***Observation:*** We report a case of embryonal rhabdomyosarcoma of the main bile duct managed in our hospital since 2022. ***Results:*** The patient was a 4-year-old male with cholestatic jaundice that had been progressing for 1 month and his general condition had deteriorated. Given this clinical picture, congenital dilatation of the main bile duct was suspected. Ultrasound followed by bili-MRI showed dilatation of the VBP and VBIH with echogenic material in the common hepatic duct, interpreted as a sludge. Surgical exploration revealed a distended gallbladder filled with serous fluid and endoluminal tumour-like material extending from the lower bile duct to the hepatic hilum, with no passage of bile and infiltration of the hepatic hilum, making bypass difficult. The tumour was biopsied. Histological examination was in favour of an embryonic rhabdomyosarcoma of the botryoid type. After a multidisciplinary consultation meeting in paediatric oncology, the patient received adjuvant chemotherapy and radiotherapy. The clinical course was favourable with complete regression of cholestatic jaundice. Follow-up imaging 6 months later showed total remission with disappearance of biliary tract dilatation. The patient has been followed up clinically and radiologically to date, with no sign of tumour relapse or recurrence ***Conclusion:*** Diagnostic challenges associated with Rhabdomyosarcoma of the biliary tract result from the diversity of the disease's forms and the ambiguous clinical presentation. The standard of care and recommended treatment strategy for rhabdomyosarcoma of the biliary tract requires multimodality treatment, including chemotherapy, surgical resection, and/or radiation therapy.

Keywords: rhabdomyosarcoma; biliary tract; liver; chemotherapy; radiotherapy; surgery.

INTRODUCTION

Embryonal rhabdomyosarcoma (RMS) of the biliary tree is extremely rare. It accounts for 1% of all rhabdomyosarcomas in children and 0.04% of paediatric cancers. It is often of the botryoid type and localised in the main bile duct or intrahepatic ducts [1-3].

Localisation in the extrahepatic bile ducts is a rare cause of jaundice in the paediatric population. It poses a significant challenge for both oncologists and surgeons. Key elements of treatment where there are no clear guidelines include surgical treatment of RMS of the bile ducts, chemotherapy and radiotherapy [4].

OBSERVATION

We review a case treated at our center, discuss treatment approaches and outcomes and compare results with the existing literature and guidelines. A 4-year-old male presented with cholestatic jaundice that had been evolving for 1 month.

Clinical examination revealed mucocutaneous jaundice and right hypochondrial pain. Everything was evolving in a febrile context with a deterioration in general condition.

Biological tests confirmed the cholestatic nature of the jaundice with an increase in conjugated bilirubin (106.8mg/L) and serum alkaline phosphatase (2339 IU/L). Given this clinical picture, congenital dilatation of the main bile duct was suspected.

Ultrasonography followed by bilio-MRI revealed dilatation of the main bile duct and intrahepatic bile duct, with echogenic material in the common hepatic duct, interpreted as sludge, as shown in the first figure 1.

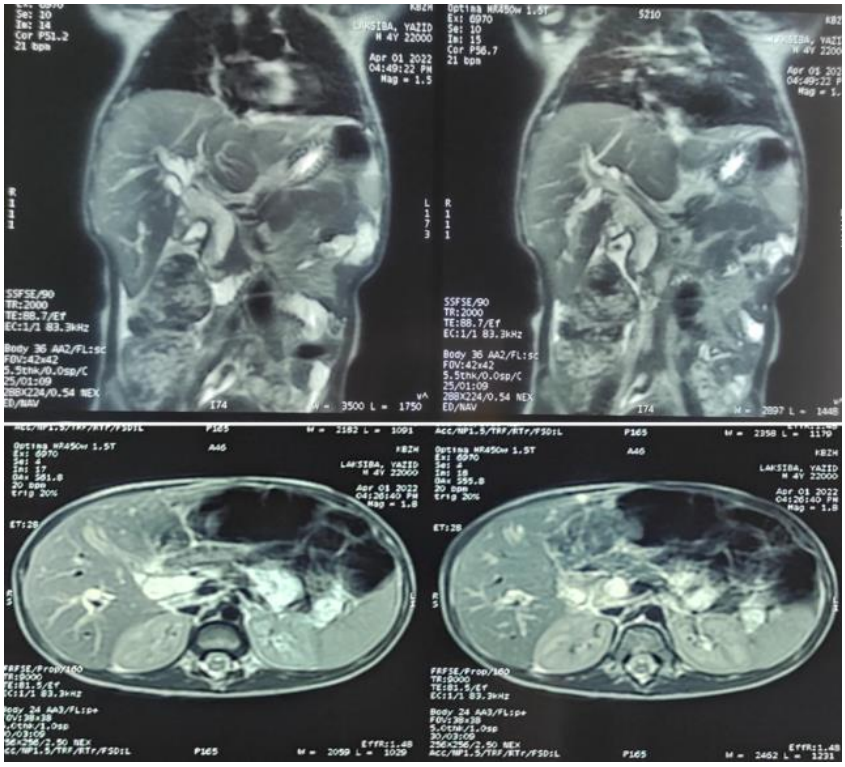

Figure 1: Initial MRI in T2 sequence after injection of gadolinium: Filling of the bile duct lumen by heterogeneous material enhanced by gadolinium with dilatation of the bile ducts

Surgical exploration revealed a distended gallbladder filled with a serous liquid with endoluminal tumour-like material extending from the lower bile duct to the hepatic hilum,as can be seen in the intraoperative photo in figure 2 and 3, with no passage of bile and biliary bypass was impossible due to invasion of the hepatic pedicle and hilum region. The tumour was biopsied.

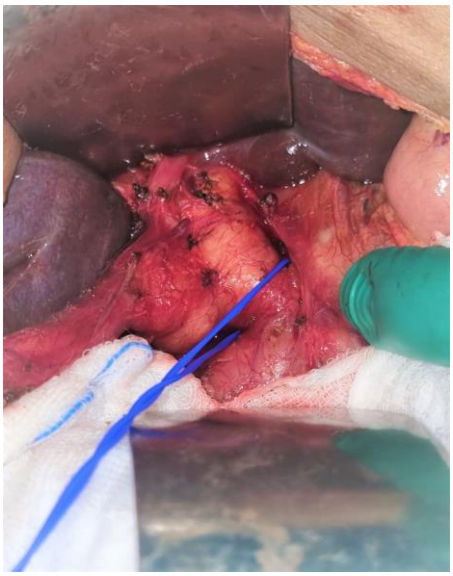

Figure 2: intra-operative appearance of the bile ducts: bile duct dilated and filled with tumour material with tumour infiltration

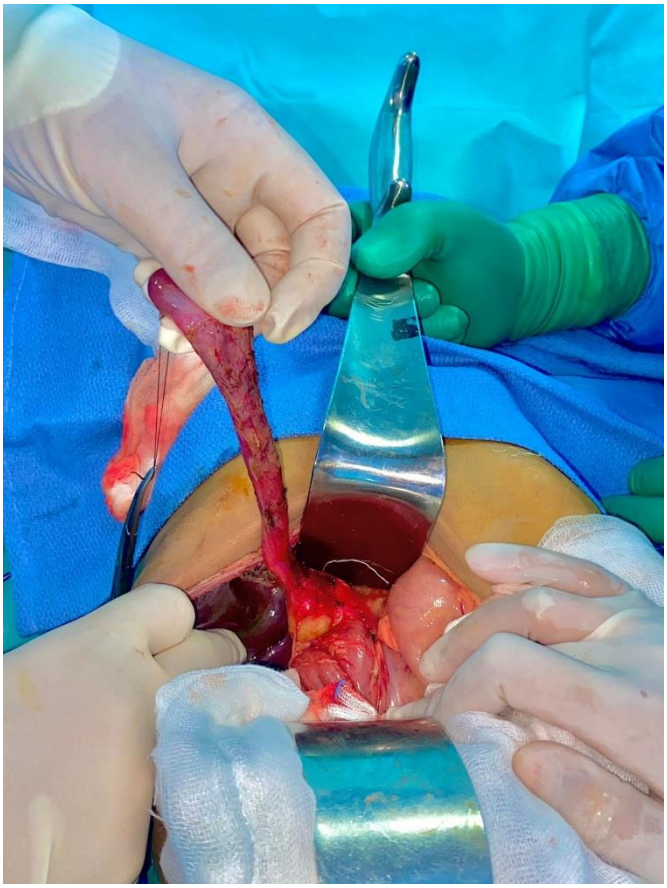

Fig 3: Histological examination was in favour of an embryonic rhabdomyosarcoma of the botryoid type

Following a multidisciplinary consultation meeting in paediatric oncology, the patient received adjuvant chemotherapy and radiotherapy.

The patient has received three courses of chemotherapy like the standard protocol, combine vincristine, actinomycin, and cyclophosphamide. The doses were adjusted according to biological cholestasis.

External beam radiation was implemented; The total radiation dose was 45 Gy to the tumor with margins with a Conformal 3-D radiation therapy without side effects, very well tolerated by patient.

The patient was monitored regularly, with a clinical examination and radiological check-up every three months. In the first year, we monitored with magnetic resonance imaging and from the second year with abdominal ultrasound. He is alive and disease-free four years from diagnosis.

DISCUSSION

The main limitation of research on RMS of the biliary tract in children is the small number of patients, even when considering multicenter analyses [1,2,3]. Therefore, any single center experience may add some information contributing to the discussions about the best treatment for children with this tumor.

It is a disease of young children aged between 1 and 9 years, with a slight male predominance [4,5]. Rhabdomyosarcoma of the bile ducts presents neoplasms of embryonal or botryoid histology. The embryonal variety is three times more common than the other varieties, but the botryoid subtype has the best prognosis because it is radio- and chemosensitive [5,6].

RMS of the bile ducts is generally manifested by obstructive jaundice in 60 to 80% of cases [7]. Other less frequent signs include abdominal pain, abdominal distension, anorexia, nausea, vomiting and fever [6].

Because of its rarity, biliary tract rhabdomyosarcoma is often misdiagnosed as a malformation of the common bile duct [1,3,7].

Magnetic resonance imaging (MRI) is the key examination for assessing the extent and characteristics of tumours at diagnosis, while ultrasound can be useful for evaluating therapeutic response and long-term follow-up [1]. The intricate anatomy of the biliary system complicates imaging interpretation and biopsy procedures, leading to delays in accurate diagnosis. Although the authors do not have experience in using positron emission tomography (PET) in the diagnosis of biliary tract RMS. for the future Studies conducted by the Paediatric Oncology Research Group aim to assess the effectiveness of PET scans in accurately differentiating

residual tumour post-therapy, a distinction that CT and MRI may sometimes fail to make [8,9].

Regional and distant metastases have been reported in 30-40% patients at diagnosis [1,2]. Histological diagnosis is established based on tissue samples obtained through primary tumor resection, open/laparoscopic biopsy or endoscopic biopsy.

From the published series, about 12% of patients had diagnostic Endoscopic retrograde cholangio-pancreatography [1]. In our opinion, it should be more widely used in establishing a diagnosis of biliary RMS.

Two main histological subtypes of RMS, embryonal and alveolar, are distinguished, embryonal being the most common. The alveolar subtype constitutes about 2% of RMS in this location [1].

The standard of care for rhabdomyosarcoma of the biliary tract requires multimodality treatment, including chemotherapy, surgical resection, and/or radiation therapy [1].

The standard chemotherapy protocols combine vincristine, actinomycin, and cyclophosphamide/ifosfamide. Surgery aims for complete tumor resection, which can be challenging due to the tumor's location in the biliary tract. Radiation therapy may be employed postoperatively to target residual disease, especially if complete surgical resection is not achievable. The protocol is often adapted based on the tumor's stage, histological subtype, and patient factors, with ongoing clinical trials helping refine these strategies. Multidisciplinary teams are crucial in tailoring treatment to optimize outcomes while minimizing complications.

The role of biliary RMS resection and its extent has been discussed by some authors, but this discussion seems to apply to patients who did not receive neoadjuvant chemotherapy [2,10]. In most reports, surgery was usually performed after preoperative chemotherapy, and often no disease on pathology was found. In most patients, thickening of the bile duct walls is still present on imaging after chemotherapy. Even if it is not, surgical excision should be performed whenever possible to achieve the highest degree of radicality. The risk of mortality associated with recurrence of RMS is higher than that of the surgery itself, even in the absence of tumor cells in the resected specimen [1].

As for primary surgery, it should only be performed to establish diagnosis and possibly determine the extent of the disease if it cannot be done by other methods (ERCP, MRCP, etc.).

Some reports demonstrate that chemotherapy alone leads to complete remission of RMS [1]. Consequently, the necessity of surgical treatment was questioned. From the available literature, there are reports which demonstrate that there are patients with biliary RMS cured with chemotherapy alone. Among seventeen patients described by Urla *et al.,* there were five patients who were not operated but received chemotherapy (according to CWS 96 protocol) alone. Among these patients, three were alive without evidence of disease at the time of the publication [2]. In Perrucio's *et al.,* study, there is also one patient treated with VAIA chemotherapy alone who is alive and disease-free 140 months from diagnosis [10].

In systematic review and meta-analysis of biliary rhabdomyosarcoma in children published in 2021, surgery after initial chemotherapy was recommended whenever possible [1]. It was shown that the lack of tumor resection is an independent risk factor for death and is significantly associated with relapse. Series published by Guerin and Urla confirm it [2,10].

Adjuvant chemotherapy after delayed surgery is recommended according to risk-adapted protocols, as in other RMS sites, and is not specific to the biliary tract location. The available literature does not depict nor suggest the number of chemotherapy courses given after delayed surgery [2,3,10]. Usually, it follows protocols using a risk classification scheme combining data about the clinical group and disease stage.

RMS are radiation-sensitive tumors. It is recommended in cases of incomplete tumor resection, alveolar histology and at relapse [1,2,11]. Aye *et al.,* report that 88% of patients who underwent radiotherapy, including those with micro and gross residual disease, did not experience recurrence, while patients who did not receive radiotherapy had local relapses [3]. In Guerin's study, four patients did not undergo surgery but instead received radiotherapy only as local treatment [10].

Among these patients, one experienced a relapse. They also observed that patients who underwent surgery and radiation had an 11% relapse rate, compared to 27% in those treated surgically (no statistical significance). Radiation therapy, according to the publications, contributes to reduced mortality in patients with biliary RMS [1].

However, radiotherapy carries a risk of early and late complications [1,2,8,11]. Complications related to radiotherapy can be significant, especially in the younger patient population. These complications may include veno-occlusive disease, nodular regenerative hyperplasia, hepatic dysfunction and secondary tumors.

For the future, new treatment strategies, such as introduction of molecular targeted drugs and immunotherapies, have shown superior efficacy and beneficial clinical outcomes as compared to standard treatments in selected childhood malignancies such as

neuroblastoma, non-Hodgkin lymphoma, low-grade gliomas and melanoma. However, benefits of new approaches in pediatric RMS are not yet available. Genomic profiling of childhood soft tissue sarcoma, which will eventually help to elucidate and discover clinically meaningful biomarkers, as of today is not standardly performed. Studies on the role of tumor mutational burden and its role as a biomarker in predicting response of soft tissue sarcomas to immune checkpoint inhibitors are inconclusive, since they have a low mutational tumor burden, and the results are based on small sample populations. Further studies of biomarkers predicting the response to treatment and identification of druggable molecular targets are needed [12].

CONCLUSION

Diagnostic challenges associated with RMS of the biliary tract result from the diversity of the disease's forms and the ambiguous clinical presentation.

Therapeutic difficulties often arise due to the unavailability of radical surgical treatment before implementing combined approaches.

The standard of care and recommended treatment strategy for rhabdomyosarcoma of the biliary tract requires multimodality treatment, including chemotherapy, surgical resection, and/or radiation therapy.

In the hope that targeted therapies and immunotherapy will do better, accurate and early diagnosis remains the best guarantee of a better prognosis.

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