Botryoid rhabdomyosarcoma of the biliary tract in children: a unique case report

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Rhabdomyosarcoma (RMS) is the most common tumour of the biliary tree in childhood. In children, it is a rare lesion, accounting for about 1% of all RMS. Hepatobiliary botryoid RMS is a disease affecting young children at a median age of about 3 years. In literature, the radiological findings of hepatobiliary RMS have been described in small series and some case reports. In this case report, we present a rare case of RMS of the extrahepatic biliary tree initially diagnosed as a choledochal cyst.

Keywords: rhabdomyosarcoma, biliary tree, botryoid.

INTRODUCTION

Rhabdomyosarcoma (RMS), a soft tissue malignant musculoskeletal tumour, accounts for approximately 1% of the cases of cancer among children aged 0–14 years and 2% of the cases among adolescents and young adults aged between 15 and 18 years [Iwai et al. 1990].

In most children, it develops as a localized disease curable with combined modality therapy, with a survival rate of >75% after 5 years from diagnosis.

Rhabdomyosarcoma is the most common tumour of the biliary tree in childhood, accounting for 1% of all paediatric RMS [Martinez et al. 1982].

Rhabdomyosarcoma occurs infrequently in the liver and biliary tree with only 45 cases reported since 1975. The management and prognosis of this tumour is constantly changing as new imaging techniques and chemotherapy as initial treatment become available [Maurer et al. 1988; Crist et al. 2001].

CASE REPORT

In March 2002, O.A.M., a 3-year-old Nigerian girl, was hospitalized for jaundice, anaemia and hepatomegaly, with WBC 17 000, AST 170, ALT 139 and total Bilirubin 16.54 mg/mL.

Objective examination showed a distended abdomen, with no pain during both superficial and deep palpation. There was no report of either hereditary diseases or previous episodes of abdominal pain.

An abdominal US scan showed an expansive formation with a diameter of about 6 cm and a mixed [solid and liquid] echostructure at the site of the hepatic hilum.
It also showed significant cystic duct ectasia, with small gallstones in the duct lumen.

Nuclear magnetic resonance showed a dilation of the common hepatic duct as well as a significant dilation of the choledochus duct up to its extra-pancreatic tract (Fig. 1).

The patient was then transferred to our Department with a diagnosis of choledochal cyst to undergo corrective surgery.

During super-umbilical transverse laparotomy, we found a major choledochal cystic formation which was difficult to isolate and remove (Fig. 2). The gallbladder appeared distended with tense neoformation surrounded by inflammatory tissue. Complete avulsion of the extra-hepatic duct and a Roux-en-Y hepatico-jejunostomy, without antireflux jejunum valve, was therefore performed (Fig. 3).

Histology revealed a botryoid RMS of the extra-hepatic bile duct [with a diameter of 5 cm] affecting both the choledochus and the common hepatic duct. There was also evidence of translucent polypoid formations both in the choledochus and in the common hepatic duct. The neoplasia developed near the margin of the proximal resection [hepatic duct], with a report of chronic cholecystitis.

Pericystic lymph nodes did not show significant structural changes.

Ten days after surgery, the patient was then transferred to the Paediatric Oncohaematology Unit and therefore discharged after 20 days.

The patient was included in to the IIa group of the Intergroup Rhabdomyosarcoma Clinical Groups Classification (Table 1). She received three chemotherapy cycles with
Vincristine, Actinomycin D, Ifosfamide and Adriamycin [V.A.I.A], in accordance with Intergroup Rhabdomyosarcoma Study Group IV. A CT scan carried out 8 and 18 months after surgery confirmed that distance metastasis did not develop. At present, the patient is being followed by our staff and is disease free.

DISCUSSION

Botryoid RMS of the biliary tree is a very rare tumour and the first case was described by Wilks and Moxon in 1875 [Wilks & Moxon 1875; Aggarwal et al. 2004].

It accounts for 1% of all RMSs, occurring predominantly in infants with a clear preponderance of male patients. About 2% of tumours are present at birth. Intermittent jaundice with or without abdominal distension, fever and anorexia is the typical presentation.

In the past few years, the approach to sarcoma botryoides of the biliary tree has apparently changed. Recently, all patients classified as stage I, including those treated at our Department, have undergone aggressive surgical resection and subsequent adjuvant therapy. After diagnosis, their mean survival rate is 5 years (80%) [Neville et al. 2000].

The Intergroup Rhabdomyosarcoma Study Group [IRSG], the National Wilms' Tumor Study Group and other two large cooperative paediatric cancer treatment groups have recently merged to form the Children's Oncology Group [COG]. Within COG, the Soft Tissue Sarcoma Committee [STS-COG] has developed new protocols for children with soft tissue sarcoma. In addition to IRSG, current STS-COG protocols also use a Tumour–Node–Metastasis (TNM)-based pre-treatment staging system. The disease stage is clinically determined by primary tumour site and size, regional lymph node status, with or without metastases [Table 2] [Lawrence et al. 1987, 1997].

The STS-COG includes patients into different treatment protocols using a risk classification scheme combining the information about clinical group and disease stage described in this report. For protocol purposes, patients are classified as low, intermediate, or high risk [Table 3] [Breneman et al. 2001; Raney et al. 2001].

The extent of surgical resection currently recommended depends on primary tumour site, and initial complete resection is generally recommended if it does not involve loss of organ function. Undetectable distant metastases at diagnosis, grossly complete surgical removal of localized tumour, botryoid histology, tumour size < 5 cm and patient's age < 10 years at the time of diagnosis are favourable prognostic factors. We also want to stress the importance of assessing regional lymph nodes since histological evaluations showed that more than 75% of patients with clinically negative regional nodes also developed microscopic tumour. This finding is important for staging and grouping purposes, affecting also chemotherapy regimen, follow-up and prognosis [Lack et al. 1981; Geoffray et al. 1987; Derek et al. 1998, Spunt et al. 2000].

Some authors recommend surgical exploration at the time of diagnosis only to confirm the diagnosis and to determine the extent of the regional disease. To this purpose, they suggest to perform a suitable incisional biopsy of the tumour and to explore the abdomen thoroughly to search for evidence of metastatic disease.

These authors also recommend minimally invasive biliary tract drainage procedures at diagnosis, like the place-
ment of an endoscopic stent. If stent placement is not possible, chemotherapy should be immediately started to try and decompress the biliary tract (Sanz et al. 1997; Pollono et al. 1998; Spunt et al. 2000).

We do agree with these authors, but we also believe that when treating children with the most favourable prognosis, like those under treatment at our Department, it is important to ensure high survival rates (>90%) and minimize long-term consequences of chemotherapy (Maurer et al. 1988, 1993; Crist et al. 1995).

CONCLUSION

In the last 20 years, the rate of disease-free survival for patients affected by RMS of the hepatic bile tree has significantly increased.

Oncologic surgery certainly plays a key role in the survival of these patients and disease staging. The introduction of chemotherapy and radiotherapy has certainly improved the treatment timing of RMS. At Stage 1, aggressive surgical therapies together with chemotherapy have increased the survival rate of patients after 5 years up to 80%.

REFERENCES


