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Botryoid Rhabdomyosarcoma of BiliaryTree:A Diagnostic Puzzle

Sanjay Raina [1], Nilay Kumar[2], Beena Jad[3] and A. N. Gangopadhyay[4]

Abstract: *Rhabdomyosarcoma (RMS) is a soft tissue malignant tumor which arises from primitive mesenchymal cells and is diagnosed in first two decades of life.It is a rare tumor in children but the most common cause of malignant obstructive jaundice in them. We report a second case of rhabdomyosarcoma of the extrahepatic bile ducts in a 6-year-old boy who presented with abdominal pain and obstructive jaundice. He was diagnosed as choledochal cyst on imaging studies. The RMS was diagnosed at laparotomy and confirmed on histopathology . Choledochotomy with tissue biopsy with T-tube insertion was done. Post operatively the patient was put on chemotherapy. Although this being an uncommon entity, second case report from our hospital within three years.*

Keywords: *Biliary rhabdomyosarcoma, Choledochal cyst, Choledochotomy.*

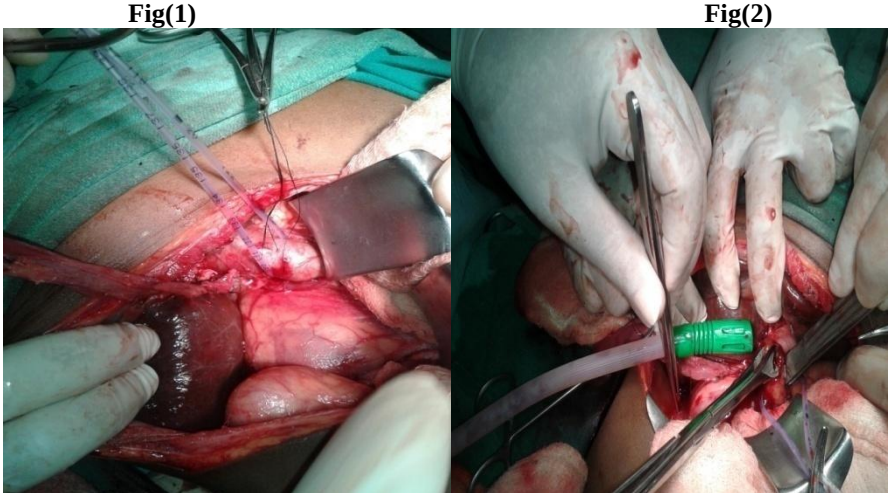
I. Introduction

RMS is a malignant tumor arising from skeletal muscle. The common sites seen in children and infants are head and neck, genitourinary tract and retroperitoneum (1) The biliary tree, however, is an extremely rare site for the primary lesion. Its prognosis is still poor, but now chemotherapy can improve the outcome. (2) Hepatobiliary RMS is a disease affecting young children at a median age of about 3 years. It is a rare tumor; however, it is the most common cause of obstructive jaundice due to neoplastic biliary obstruction in children (3). We treated a case of RMS of the extrahepatic biliary tree, which was initially diagnosed to be a choledochal cyst.

II. Case report

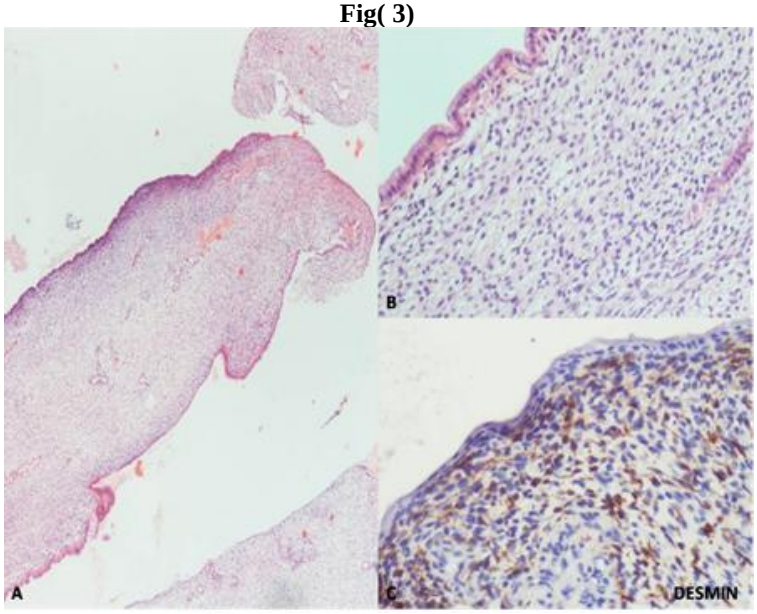
A 6-year-old boy was admitted for progressive jaundice, pruritis, vomiting, loss of appetite and fatigue which had developed over three month duration. On examination, the child was alert and in discomfort. Jaundice and pallor were noted. There was mild hepatomegaly and liver could be palpated 8 cm below the costal margin. No splenomegaly or ascites was noted. Laboratory investigations showed Hemoglobin 8 gm%, White blood cell count 16,000 with 56% polymorphs, platelets 2.6 lakhs, INR 1.6. Total serum bilirubin 7.8 mg%, Direct 5 mg%, SGOT 80 IU, SGPT 125 IU/L and Alkaline phosphatase 2311 IU/L. Urine positive for bile salts and pigments. Renal function, prothombin time, and partial thromboplastin time were within normal limits. Ultrasonography (USG) of the abdomen showed dilated common bile duct (CBD) (20mm in diameter), and dilated intrahepatic biliary channels. MRCP showed fusiform dilation of CBD,the right and left hepatic ducts and intrahepatic biliary radical were dilated, suggestive of type 1 choledochal cyst. An exploratory laparotomy revealed hepatomegaly. CBD as well as right and left hepatic ducts were hugely dilated with firm consistency and irregular surface Fig (1). Choledochotomy revealed, bunch of grapes like gelatinous material mixed with bile Fig(2)

Fig(1) **Fig(2)**



Tissue biopsy with T-tube insertion and external drainage procedure was done; Cholecystectomy was performed along with it. There was no evidence of metastasis or lymph node involvement. The histopathology was suggestive of botryoid RMS Fig (3)

Fig(3)



A- Tumor showed polypoid nodules, with an abundant loose, myxoid stroma embedded in which are small undifferentiated cells and showing distinctive hypercellular zone (cambium layer) just below the columnar mucosal epithelium (Hematoxylin & eosin,40x). **B**- High power view showed round to oval , spindle and stellate shaped primitive mysenchymal cells embedded in a loose myxoid matrix. (Hematoxylin & eosin stain,400X) **C**- Several tumor cells show immunoreactivity for desmin (Desmin, 400X)

Postoperatively, chemotherapy was given including vincristine, dactinomycin and cyclophosphamide (VAC Regime).T-tube cholangiogram was done which showed no features of hepatobiliary obstruction and thus T-tube was removed.On follow up patient received six cycles of VAC regime. Patient recovered and follow up MRCP revealed full regression of the residual mass.

III. Discussion

Embryonal rhabdomyosarcoma of the biliary tree, sometimes referred to as boytroid rhabdomyosarcoma, only accounts for 0.04% of childhood neoplasms.(4) It accounts for 1% of all embryonal rhabdomyosarcomas.(4) It was first described by Wilks and Moxon in 1875 on the basis of the typical location and gross description of the tumor. Median age at presentation is 3 ½ years, with a slight male predominance. (5,6) Histopathologically the tumor arises as an intraluminal biliary mass or cluster of grape-like masses and is similar to sarcoma botryoides of the bladder or vagina in children. The tumor has a yellow, shiny, gelatinous appearance.(7) The tumor often exceeds 8 centimeters at the time of discovery and can invade the duodenum 8,9) It can arise from almost anywhere along the biliary tree including liver, intrahepatic and extrahepatic. biliary ducts, gallbladder, or ampulla It has also been reported to arise from hepatic and choledochal cysts.(10)

The most common clinical features are jaundice and abdominal distention with pain, vomiting and fever being less frequent. (8,11) Elevation in liver transaminases and bilirubin is often present. A tumor arising from the biliary tree discovered in children over one year of age is most commonly an embryonal rhabdomyosarcoma, however other considerations include: choledochal cysts, inflammatory pseudotumor, and cholangiocarcinoma arising within a choledochal cyst. (10)

Ultrasonography generally demonstrates biliary ductal dilatation, an intraductal mass, or fluid filled mass if the tumor has a cystic component. (4) Because of this, it can have a radiologic appearance similar to a choledochal cyst, especially if there is no local invasion. (9) CT may show a heterogeneous or hypoattenuating mass with biliary ductal dilatation. (10) MRI has advantages over other modalities because of its ability to define the extent of disease and relationship to hepatic vasculature.(4) It is usually diagnosed at surgery or by liver biopsy.(5) The management of embryonal RMS has improved greatly over the past 30 years, with a 3 fold increase in the cure rate from 25% in 1970 to greater than 75% currently. It is believed that more intensive chemotherapy is mainly responsible for result. The treatment of embryonal RMS currently recommended by the

Intergroup Rhabdomyosarcoma Study IV is a combination of vincristine, actinomycin- D and cyclophosphamide, ifosfamide or etoposide.(12) In our case same regimen was followed. Although systemic multiagent chemotherapy is the mainstay of treatment, controversy still exists as to the appropriate local regional management of the primary tumor. Many advocate surgery as the initial approach while others assert that RMS is uniquely radiosensitive and possibly curable with radiation therapy. (13)

A review of the literature has reported several different surgical approaches depending on the extent of the tumor burden. These include pylorus sparing pancreaticoduodenectomy(14), Roux-en-Y hepaticojejunostomy (15), and choledochojejunostomy with cholecystectomy and end to side jejunojejunostomy (16) to name a few. In some cases, chemotherapy and radiation therapy may be the initial treatment to shrink tumor burden prior to surgical consideration.(4) Because of the use of adjuvant chemotherapy, positive margins after surgical resection are tolerable and do not seem to increase mortality. With the combination of therapies, survival has increased significantly and a recent study reported a survival rate of greater than 75%, compared to 25% in 1970.(16) However, other studies recommend long term follow up because hepatobiliary rhabdomyosarcoma has been reported to recur up to 9 years after initial therapy.(10)

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