Botyroid Rhabdomyosarcoma of the Common Bile Duct

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Abstract
Rhabdomyosarcoma is commonly seen in children and infants, the common sites are head and neck, genitourinary tract and retroperitoneum. Tumours of the extra hepatic biliary tract are rare in childhood. We report a case of one and half year old boy who presented with lump in right hypochondrium, pain in abdomen, jaundice and fever.
Clinical impression was hepatitis. Ultrasonography suggested a choledochal cyst, whereas computed tomography reported a neoplastic lesion. Histopathological examination diagnosed a botyroid rhabdomyosarcoma of the common bile duct which presented clinically as a choledochal cyst.

Introduction
Rhabdomyosarcoma of the biliary tree is an uncommon malignancy. This tumour can be clinically misdiagnosed as hepatitis or choledochal cyst. In our case, the child presented with symptoms suggestive of hepatitis and radiological investigations diagnosed it as a choledochal cyst. Rhabdomyosarcoma of the biliary tree should be kept as a differential diagnosis in children with symptoms suggestive of obstructive jaundice.

Case Report
A one and half year old boy presented with lump in right hypochondrium since one month. He also had pain in abdomen, jaundice and fever since 20 days. The initial clinical impression was hepatitis. Liver function test revealed marginally elevated transaminases with markedly elevated alkaline phosphatase of 2777 KA units.

Ultrasonography suggested a choledochal cyst, whereas computed tomography suggested a neoplastic lesion (Fig. 1). Intra operatively, on opening the choledochal cyst, multiple shiny white tissue bits were removed and sent for histopathological evaluation.

Roux –en – Y anastomosis was done for the patient.

Histopathological Examination
Multiple bits were received. Many bits showed necrosis. One bit showed a lining of cuboidal epithelium. Subepithelial tissue showed a tumour with densely cellular as well as hypocellular areas. Dense condensation of tumour cells was seen beneath the epithelium forming a cambium layer. Tumour cells were ovoid, spindle, stellate shaped with ovoid
hyperchromatic nuclei. Intermixed were few unipolar and bipolar spindle cells.

Occasional strap cells were identified (Fig. 2). Densely cellular areas revealed 2-3 mitosis/high power field. Hypocellular areas had abundant myxoid extracellular matrix. The histomorphology confirmed the diagnosis of botryoid rhabdomyosarcoma of common bile duct.

Discussion

Rhabdomyosarcoma is a malignant tumour arising from cells committed to skeletal muscle differentiation. Though this tumour is commonly seen in children and infants, the common sites are head and neck, genitourinary tract and retroperitoneum.1 Tumours of the extra hepatic biliary tract are rare in childhood but rhabdomyosarcoma is most common among these unusual tumours.2 The first case was reported by Wilks and Masson in 1875. Several case reports have been published since then which were clinically diagnosed as hepatitis or choledochal cyst.3-5 Age of presentation is usually 3-4 years. Luis A6 et al reported the youngest case in a 9 month old girl. Within the extra hepatic biliary tract the common sites are common bile duct (77%), hepatic duct (12%), liver, gall bladder and ampulla (4% each).2 Ultrasonography usually diagnoses the tumour as a choledochal cyst. Thus, CT scan complements the sonographic evaluation by displaying the solid nature of the tumour and also determines the operability of the mass.7

Intraoperative finding of soft white tissue bits also correlated well with the gross features described for rhabdomyosarcoma. Rhabdomyosarcoma has been divided into 4 clinical stages by intergroup rhabdomyosarcoma study based on the extent of disease and type of surgery performed.8

Stage I – Localised disease completely resectable.

Stage II – Microscopic residual following surgical excision.

Stage III – Gross residual tumour following surgical excision.

Stage IV – Metastatic disease.

Current modalities of treatment include surgical removal, radiation and chemotherapy.

However, spunt9 et al in his study reported a good outcome despite residual disease after surgery Fredrick and his colleagues10 in a study conducted over 10 year period suggest a bile culture for aerobic and anaerobic organisms as purulent cholangitis contributes significantly to postoperative morbidity and mortality. Our patient was given 4 cycles of chemotherapy, a CT scan done later showed marked reduction in the size of the tumour. Metastases of this tumour are initially seen in the regional lymph nodes, liver, retroperitoneum and lungs. Thus, an aggressive multidisciplinary approach is essential in treatment of this tumour at this unusual site.

References

POSSIBLE HARMS OF OSELTAMIVIR - A CALL FOR URGENT ACTION

Japanese Ministry of Health, Labour, and Welfare has funded two prospective cohort studies, and subsequently advised against oseltamivir's use in children and adolescents aged 10-19 years. This study found evidence of unusual behaviour in recipient children within the first day of infection.

A detailed independent review of eight serious cases concluded that three sudden deaths during sleep and two near-deaths, as well as two deaths from accidents resulting from abnormal behaviour in older children and adolescents shortly after taking oseltamivir, were probably related to the central depressant action of oseltamivir.

Despite these limitations, many serious harms were reported in people younger than 20 years. Abnormal behaviour, convulsion, delirium, and hallucinations were more common in the young.

Despite these inadequate data, the consistent reports of potential serious harms in adolescents, and the projected future heavy use of oseltamivir, make it imperative to quickly establish large multicentre studies to test any possible associations.