Embyronal rhabdomyosarcoma of the biliary tree mimicking a choledochal cyst

ABSTRACT

Embryonal rhabdomyosarcoma (ERMS) of biliary tree is a rare type of mesenchymal neoplasm diagnosed at surgery or by preoperative liver biopsy. We present a one year eight months old child who mimicked a choledochal cyst and was eventually treated with surgery, chemotherapy with IRS IV protocol and adjuvant postoperative radiotherapy to surgical bed with 6 MV photons to a dose of 5040cGy in 28 fractions.

KEY WORDS: Choledochal cyst, embryonal rhabdomyosarcoma

Embyronal rhabdomyosarcoma (ERMS) of biliary tree is a rare mesenchymal neoplasm (0.5%) which affects infants and children. The most common presentations are of obstructive jaundice and/or pruritus. Diagnosis is usually made at surgery or by preoperative liver biopsy. Biliary RMS remained incurable for nearly 100 years until the report of a child cured by partial surgical resection, radiotherapy and vincristine and dactinomycin chemotherapy in 1971. It is crucial to identify it, differentiate it from choledochal cyst and start treatment at the earliest.

CASE REPORT

A one year eight months old child presented with jaundice, clay colored stool, dark urine, occasional pain in the abdomen at another hospital. Examination revealed icterus, with mild hepatomegaly. On investigation the counts were normal and bilirubin and transaminases were raised (Sr. bilirubin - 8.6 direct 6.6 indirect 2 mg%, SGOT 148, SGPT 134, Sr. Alk Phos 484). His renal function tests and chest X-ray were normal.

A CT scan of abdomen revealed multiple cystic lesion in biliary radicles of both lobes of liver (L>R) with dilatation of common hepatic and common bile duct suggestive of choledochal cyst. MRI of abdomen contributed to the same findings. MRCP revealed tubular branching with gross dilatation of both lobes and biliary radicles with dilated common bile duct reaffirming the diagnosis made on CT/MRI imaging.

Patient underwent choledochojejunostomy with cholecystectomy with Roux - en - y end to side jejunojjejunostomy in November 2005. Intraoperatively there was probable infiltration to the surrounding hepatic parenchymal tissue. Postoperative imaging revealed minimal soft tissue mass left behind suggestive of R + resection.
Histopathology report with Immunohistochemistry showed round spindle stellate cells with abundant mitosis with inflammatory reaction suggestive of ERMS, positive for desmin.

After establishing the diagnosis, the patient was started on IRS IV protocol after complete metastatic workup (bone marrow, bone scan Normal). Patient completed IRS IV protocol with adjuvant postoperative radiotherapy to surgical bed with 6 MV photons to a dose of 5040cGy in 28 fractions with shrinking portals. The boost was planned with three dimensional conformal radiotherapy technique. Pt tolerated the treatment well and was disease free for one year till his last follow up.

DISCUSSION

Embryonal RMS is a rare tumor in children that can present in the bladder/prostate, extremities, parameningeal area, retroperitoneum, head and neck region and biliary tract. 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. 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. Metastatic disease is apparent on radiographic studies in up to 30% of patients.

The common presentation is features of obstructive jaundice. Pain, nausea and vomiting and fever are less common symptoms.

The differential diagnosis of a mass in the porta hepatis in a child includes hepatoblastoma or undifferentiated embryonal sarcoma, hepatocellular carcinoma, pancreatic neoplasms such as pancreaticoblastoma and papillary cystic tumor of the pancreas, metastatic lesions of the liver such as neuroblastoma and Wilm’s tumor, lymphoma and granulomatous disease with retroperitoneal lymphadenopathy, lung nodules or liver masses. However, in children, there are no neoplasms that arise from the bile ducts other than embryonal RMS.

Histopathologically, embryonal RMS is characterized by immunohistochemical staining for desmin and muscle-specific actin.

However, neither of these stains is specific for this type of tumor. Recently, myogenin has been shown to be a specific immunohistochemical marker for embryonal RMS.

It is usually diagnosed at surgery or by liver biopsy and may be suspected on radiographic studies such as CT, MRI or percutaneous transhepatic cholangiography. 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. Three-year failure-free survival with use of these regimens ranges from 78% to 92%, depending on tumor site and stage. 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. It is well-established that in addition to chemotherapy, the majority of children will also need surgery or radiotherapy or occasionally a combination of both, to achieve local tumor control. The decision regarding resection or irradiation should take into account the stage of the lesion, age of the patient, site of the primary tumor and the likely sequelae of an aggressive surgical procedure or intensive radiation therapy.

REFERENCES

Nemade, et al.: Embryonal rhabdomyosarcoma


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