Pediatr Surg Int (1997) 12: 200–201

CASE REPORT

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Rhabdomyosarcoma of the biliary tree

Accepted: 25 April 1996

Abstract Rhabdomyosarcoma (RMS) of the biliary tree is a rare tumor in children that has a very poor prognosis. Preoperatively, it is often mistaken for a choledochal cyst. We report a case of RMS of the biliary tree in a 4-year-old girl who presented with abdominal pain and obstructive jaundice. The RMS was diagnosed at laparotomy; excision was not possible due to its size and localization. Chemotherapy achieved complete regression of the tumor observed at second-look surgery. Preoperative chemotherapy can now avoid mutilating surgical procedures and improve survival.

Key words Biliary rhabdomyosarcoma · Bile-duct tumor · Preoperative chemotherapy

Introduction

Rhabdomyosarcoma (RMS) is the most frequent soft-tissue tumor in children. The most common primary sites are the genitourinary tract, extremities, and head and neck [4]. 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. We report a case recently treated in our hospital.

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Case report

A 4-year-old female was admitted with several months' history of abdominal pain. One month before admission she developed itching, brown urine, and jaundice. No further remarkable findings were seen on physical examination. Laboratory investigations showed an erythrocyte sedimentation rate of 17 mm/h, total serum bilirubin 6.7 mg% (direct 6.2), glutamic oxaloacetic transaminase 336 IU/l, glutamic pyruvic transaminase 723 IU/l, alkaline phosphatase 714 IU/l, γ-glutamyl transferase 1054 IU/l, lactic dehydrogenase 778 UI/l, cholesterol 318 mg%, α_1-antitrypsin 206 mg/dl (N = 5,000–85,000) and ceruloplasmin 46 mg/dl (N = 27–56). Hydatid, hepatitis, and cytomegalovirus serologies were negative. A barium meal study showed the second part of the duodenum compressed by an extrinsic tumor. Liver scintigraphy (HIDA) revealed hepatomegaly and total obstruction of bile flow. Sonography (US) showed a 4.5-cm, non-homogeneous, echogenic mass near the porta hepatis and markedly dilated intrahepatic ducts. Computed tomography (CT) showed hepatomegaly and a hypodense mass arising at the porta hepatis and extending down to the duodenum (Fig. 1). The clinical diagnosis before surgery was a choledochal cyst.

A non-resectable, firm, malignant mass was found at laparotomy involving the extrahepatic bile ducts (BD), between the porta hepatis and duodenum. It included the portal vein and hepatic artery and displaced the inferior vena cava posteriorly. There were enlarged regional lymph nodes. The intraoperative cholangiogram (Fig. 2) showed a large mass in the common bile duct (CBD), dilated intrahepatic ducts, and the entire afected biliary tree. A diagnosis of RMS of the bile duct (RMS/BD) was made by operative pathologic examination. After a liver biopsy, cholecystectomy, and T-tube insertion in the CBD, the abdomen was closed.

The tumor was designated as stage IV (pT3B pN2a M0). Chemotherapy with carboplatin, epirubicin, vincristine, ifosfamide, and VP-16 was given according to the International Society for Pediatric Oncology. Twenty-one weeks later, a T-tube cholangiogram and CT showed disappearance of the tumor. At second-look laparotomy the extrahepatic BD were resected and a Roux-en-Y hepaticojejunostomy was made. No tumor cells were seen on pathologic examination, which showed only granular tissue and fibrosis of the BD wall. Three months later, the patient died during the last phase of chemotherapy (massive chemotherapy and bone marrow transplantation) due to chemotherapy-related immunosuppression with acute renal failure.

Discussion

Rhabdomyosarcoma is the most frequent soft-tissue tumor in children. It can occur in almost every area of the body, but the most common primary sites are the genitourinary tract, extremities, and head and neck. RMS/BD constitutes less than 1% of all RMS [10], but is the most common malignancy of this site in children [2]. It was first reported in 1875 by Wilks and Moxon [12], and to date 58 cases have been reported in the literature.

RMS/BD often occurs in children aged 3–4 year [8], most frequently in girls (5/1). Obstructive jaundice and abdominal pain constitute the classic symptomatology. The best imaging

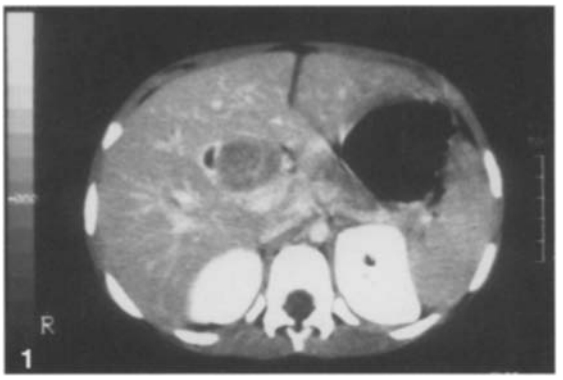

Fig. 1 Computed tomography scan showing size and localization of tumor

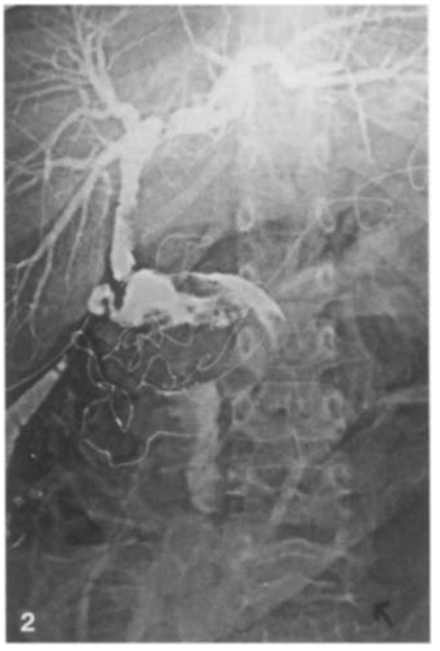

Fig. 2 Intraoperative cholangiogram

procedures are US, CT, and magnetic resonance imaging, which reveal the lesion as a hypodense intraductal mass with intrahepatic BD dilatation [6]. However, the definite diagnosis often is not possible until laparotomy: the usual diagnosis before surgery is choledochal cyst [1, 6, 8]. The most frequent histologic diagnosis is embryonal RMS or sarcoma botryoides. It is characterized by a high risk of local recurrence and a low risk of remote metastases.

Radical surgery was traditionally considered the treatment of choice for RMS/BD, and long-term survival after resection alone has been reported [9]. Nevertheless, improvement of outcome has only been obtained by the combination of surgery, radiation, and chemotherapy, and the longest known survivals were related to the use of multidisciplinary therapy [8], which might be due to the characteristics of these tumors (multilocular growth) that make complete surgical resection almost impossible. Several surgical procedures have been employed, from BD aspiration and curetage [5] to more aggressive technics such as partial resection of the liver or pancreaticoduodenectomy [1, 3, 9]. The most frequently used chemotherapeutic agents are cyclophosphamide, vincristine, and actinomycin D. Adriamycin, cisplatin, and etoposide have also been used. In this case, primary resection of the tumor was not possible due to its local extension. Surgery consisted only of biopsies of the tumor and liver, cholecystectomy, and insertion of a T-tube in the CBD. We consider it essential to ensure adequate bile drainage before chemotherapy.

It is commonly accepted that chemotherapy is the first therapeutic step when excision is not possible, the aim being devitalization of the tumor cells and reduction of its mass. Although preoperative chemotherapy has proved to be effective in the treatment of RMS situated elsewhere, results are not so clear in RMS/BD. While some authors have reported cases where preoperative chemotherapy did not reduce the size of the tumor [9], others have established its usefulness to facilitate surgery and avoid mutilating procedures [6, 8, 11]. Mulet et al. reported two cases treated by BD curetage and chemotherapy, and we think that their patients' long survival were probably due to chemotherapy [5]. Perisic et al. [7] observed tumor disappearance from the CBD with chemotherapy and obtained histologic findings similar to ours.

In our case chemotherapy proved to be effective achieving complete disappearance of the tumor at second-look surgery and histologically. It would be the second case in the literature of proven total tumor regression after chemotherapy. In conclusion, the prognosis of RMS/BD is still poor, but we believe that surgery with pre- and postoperative chemotherapy can improve survival.

References

1. Caty MG, Oldham KT, Prochownik EV (1990) Embryonal rhabdomyosarcoma of the ampulla of Vater with long-term survival following pancreaticoduodenectomy. J Pediatr Surg 25: 1256–1258
2. Isaacson C (1978) Embryonal rhabdomyosarcoma of ampulla of Vater. Cancer 41: 365–368
3. Martínez F LA, Haase GM, Koep LJ, Akers DR (1982) Rhabdomyosarcoma of the biliary tree: the case for aggressive surgery. J Pediatr Surg 17: 508–511
4. Maurer HM, Belgantady M, Gehan EA, et al. (1981) The Intergroup Rhabdomyosarcoma Study-I. A final report. Cancer 61: 209–220
5. Mulet JF, Illa J, Mainou C, Ros J, Claret I (1981) Rabdomiosarcoma de vías biliares. An Esp Pediatr 17: 65–70
6. Oelsnitz GVD, Spar HJ, Lieber Th, Münchow B, Booß D (1991) Embryonal rhabdomyosarcoma of the common bile duct. Eur J Pediatr Surg 1: 161–165
7. Perisic VN, Howard ER, Mihailovic T, Vujanic G, Milovanovic D, Ivanovsky P (1991) Cholestasis caused by biliary botryoid sarcoma. Eur J Pediatr Surg 1: 242–243
8. Ruymann FB, Raney RB, Crist WM et al. (1985) Rhabdomyosarcoma of the biliary tree in childhood. A report from the Intergroup Rhabdomyosarcoma Study. Cancer 56: 575–581
9. Schweizer P, Schweizer M, Wehrmann M (1994) Major resection for embryonal rhabdomyosarcoma of the biliary tree. Pediatr Surg Int 9: 268–273
10. Sharma AK, Lahoti BK, Aggarwal R, et al. (1995) Obstructive jaundice may be due to a malignant tumour of the common bile duct. Pediatr Surg Int 10: 496–497
11. Treuner J, Kaatsch P, Anger Y, et al. (1986) Ergebnisse der Behandlung von Rhabdomyosarkomen bei Kindern. Ein Bericht der Cooperativen Weichteilsarkomstudie (CWS-81). Pädiatr Onkol 10: 208–217
12. Wilks S, Moxon W (1875) Lectures on pathological anatomy. Longman, Brown, Green, Longmans, Roberts & Co, London, pp 465–466