[症例報告]Abdominal Burkitt's lymphoma presenting with obstructive jaundice in a child : A case report

Abdominal Burkitt's lymphoma presenting with obstructive jaundice in a child : A case report

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ABSTRACT

A case of nonendemic Burkitt's lymphoma in a 4-year-old girl with obstructive jaundice as the initial symptom is reported. On admission, the total bilirubin was 7.3 mg/dl. Abdominal US and CT findings revealed a tumor mass in the porta hepatis and two masses in the pelvic cavity. Subsequently, she underwent an elective laparotomy for tissue diagnosis. A small mass of tissue was removed from the tumor in the hepatoduodenal ligament, whereas the right ovarian tumor was resected. Both tissue specimens showed B cell lymphoma (Burkitt's lymphoma) with bone marrow involvement. From these data, the patient was diagnosed as having stage Ⅳ lymphoma. Following the diagnosis, she underwent chemotherapy, and has been doing well two months after the chemotherapy. *Ryukyu Med. J., 16(3)123~126, 1996*

Key words: Burkitt's lymphoma, abdominal tumors, jaundice

INTRODUCTION

The abdomen is the most frequent site of involvement in nonendemic Burkitt's lymphoma. In addition, the majority of patients with primary abdominal disease, also tend to have intraabdominal disseminated disease[1-5]. The role of surgery in the management of lymphoma, however, remains unclear. In clinical practice, surgery may be one of the diagnostic tools used to delineate the diagnosis and may also be needed to either remove or debulk the primary tumor as well as help manage various complications, which may arise in the course of treatment.

Primary abdominal lymphoma has been found to present in various ways. However, an extrahepatic biliary obstruction with lymphoma involvement occurs very rarely, and results in the development of obstructive jaundice. In this report, we describe a case experience of Burkitt's lymphoma of the abdomen in a child with an unusual presentation, involving obstructive jaundice, and a brief literature review of the role of surgery in the management of such patients.

CASE REPORT

A 4-year-old girl initially presented with general malaise and appetite loss in October, 1995. She later developed jaundice and, was examined at a local medical clinic. She was referred to the Ryukyu University Hospital on October 26 for a further evaluation of her disease. On hospitalization, she appeared healthy but with mild jaundice. A physical examination revealed her tonsils to be swollen. There was no palpable superficial lymphadenopathy. The chest revealedd no abnormalities. The liver was palpable 2 finger breadths below the right costal margin with no evidence of any abnormal pathological findings. Laboratory examination disclosed normal reference ranges of the blood cell count, a total bilirubin level of 7.3 mg/dl, SGOT, SGPT, LDH, and γ-GPT levels of 205, 177, 1110 and 222 IU/L. Tumor markers including AFP, CEA and CA19-9, were all within the normal limits.

Abdominal ultrasonography (US) and CT:

Both revealed a localized hypoechoic or hypodensity mass measuring 5 cm × 4 cm in size below the porta hepatis. The intrahepatic bile ducts were dilated up to 1.0 cm in size and the proximal common bile duct measured 1.3 cm (Fig.1, 2).

Neither tumor masses nor enlarged lymph nodes were observed in the upper abdominal cavity, but two tumor masses measuring 3 cm × 3 cm in size were found in the location of the bilateral ovaries in the pelvic cavity. Ga scintigraphy revealed a high uptake in the tumors in the hepatoduodenal ligament and the pelvic cavity. An upper gastrointestinal series revealed a lateroposterior displacement of the second and third portion of the duodenum with evidence of luminal stenosis (Fig.3).

Percutaneous transhepatic cholangiography (PTC):

PTC under US guidance revealed a round, smooth stricture of the common bile duct just above the confluence

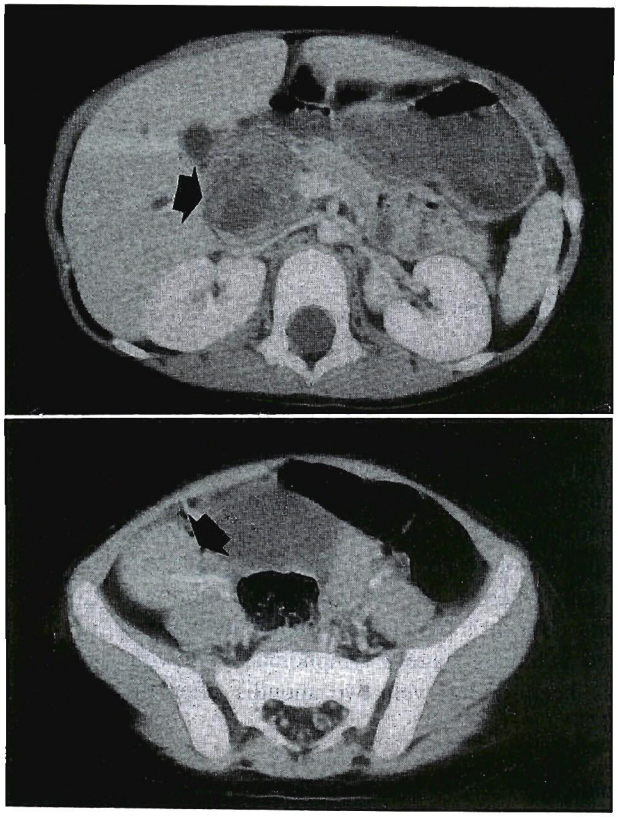

Fig.1 An enhanced abdominal CT scan showing a hypodensity mass in the porta hepatis (top; arrow) and a hyperdensity tumor in the pelvic cavity (bottom; arrow).

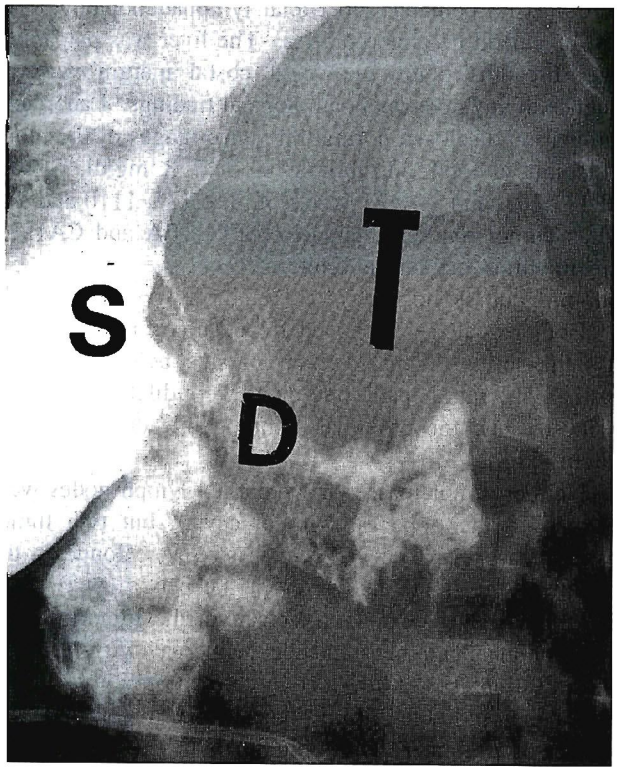

Fig.3 An upper gastrointestinal series (a right oblique view) demonstrating both a displacement and stenosis of the duodenum (T: tumor, S: stomach, D: duodenum).

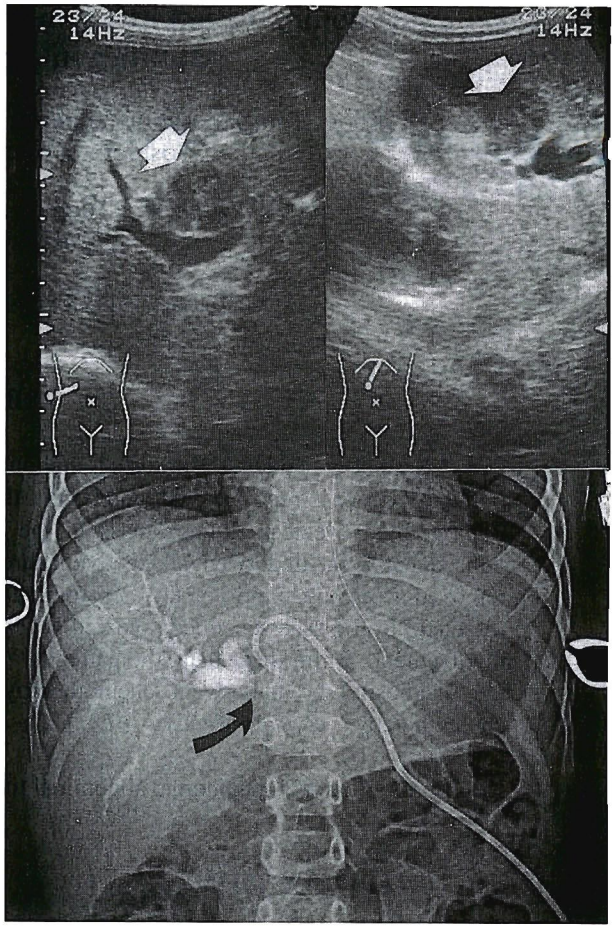

Fig.2 Abdominal ultrasonogaphy revealing a hypoechoic tumor in the porta hepatis (top; arrows) and PTC using a drainage tube showing a smooth occlusion of the hepatic duct (bottom; arrow).

of the cystic duct and the hepatic duct (Fig.2). A percutaneous transhepatic biliary drainage (PTBD) tube was placed in the left hepatic bile duct for biliary decompression.

No definite diagnosis could be made. Subsequently, she underwent an elective exploration for tissue diagnosis. A localized tumor was found within the hepatoduodenal ligament, and a small amount of tissue was removed with some associated bleeding. In addition, a resection of the right ovarian mass and bone marrow aspiration from the sternum were also carried out. The resected ovarian tumor weighed 42 g. The pathological findings indicated Burkitt's lymphoma (B cell lymphoma) with involvement of both the right ovary and bone marrow (Fig.4). She was therefore diagnosed as having stage Ⅳ lymphoma according to the Murphy staging system.

Following the diagnosis, the patient receieved chemotherapy. The treatment consisted of five cycles repeated consecutively at 13-day intervals using cyclophosphamide, vincristine, predonisone, adriamycin and methotrexate. Two months after the first cycle of chemotherapy, an upper gastrointestinal series showed an improvement in the duodenal stenosis. PTC using a drainage tube revealed the

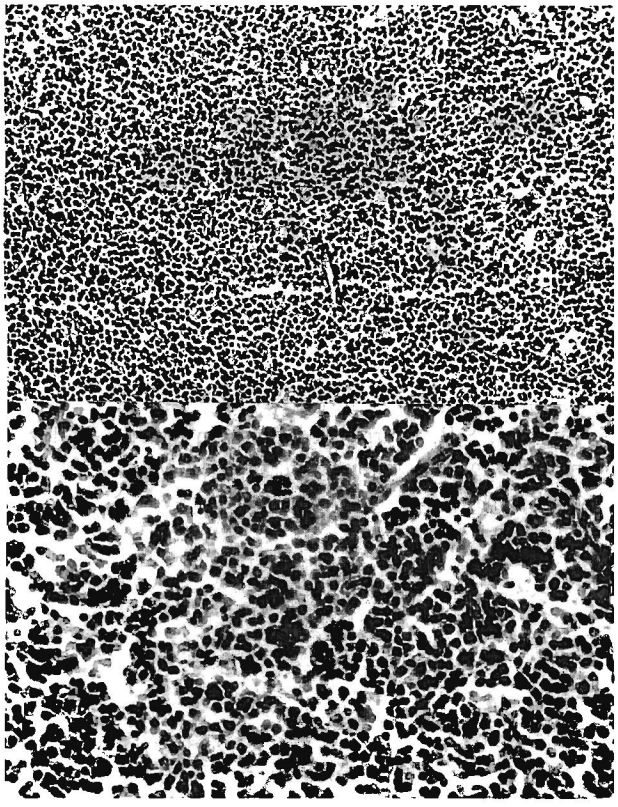

Fig.4 Microphotographs showing B cell lymphoma (medium sized, diffuse type) (top: HE, ×25) (bottom: HE, ×50).

obstructed bile duct to have been reopened. She is currently in her second cycle of chemotherapy.

DISCUSSION

Nonendemic Burkitt's lymphoma has been detected with greater frequency in pediatric population[6-8]. This lymphoma comprises approximately 15% of all cases of non-Hodgkin's lymphoma. Forty-five per cent of Burkitt's lymphoma cases occurs in the abdomen, whiles 36% is observed in the head and neck[11]. Only a few cases (1.4-3.5%) of primary abdominal Burkitt's lymphoma[9-11] have been reported involving obstructive jaundice as the initial presentation. Rosenberg et al. reported that non-Hodgkin's lymphoma frequently developed obstructive jaundice either in the advanced or late stage of the disease, thus accounting for 12.5% of all cases[1]. Our patient was initially presented with jaundice. The abdomen had a localized tumor mass within the hepatoduodenal ligament with involvement of the ovaries and bone marrow, and thus the patient was diagnosed as having stage Ⅳ lymphoma[12]. Some authors have reported that patients with primary abdominal lymphoma demonstrates poor prognosis[6-8], primarily due to really evidence of metastases along with the involvement of either their bone marrow or central nervous system. Most such patients die due to recurrent disease despite an initial positive response to chemotherapy with median survival time of 6 months. Compared to the findings of the same disease located in the head and neck, this poor survival could possibly be attributed to the fact that primary abdominal Hodgkin's lymphoma may either represent a different biological entity, or a more advanced stage of this disease.

The role of surgery in the treatment of Burkitt's lymphoma, still remains to be clarified. Some reports have suggested that an increased survival was noted in patients who underwent either a complete or near complete resection of the tumor[3-5]. However, such findings may only reflect the fact that localized, less progressive disease is more easily completely resected. In patients with disseminated disease, surgery should be only limited to a tissue biopsy. However, even if a complete surgical resection is not possible, most patients will require surgical intervention at some time during their treatment. The types of surgical intervention include either an emergent laparotomy, an early biopsy to make a diagnosis, or for the management of the complications arising due to chemotherapy. Such surgical intervention may therefore play a role in the increased survival of these patients.

Unfortunately, for such patients, the procedures other than a laparotomy and tumor biopsy tend to also carry a high risk. Some reports have suggested that the most rational approach for an unresectable abdominal Burkitt's tumor is to perform a laparotomy for a biopsy only when the diagnosis can not possibly be made by any other methods and to shrink the tumor using chemotherapy[6-8].

Taking these reports into consideration, we thus conclude that it is desirable to resect a localized Burkitt's tumor as early as possible and immediately follow with induction chemotherapy. The debulking of large tumors is not considered to contribute to survival, and particularly in this group of patients, as well as those in whom the tumor is disseminated, chemotherapy should thus be the treatment of choice. Surgery should be limited to only a tissue biopsy.

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