

Extramedullary Plasmacytoma of the Gall Bladder : A Case Report

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Abstract

Extramedullary plasmacytomas are uncommon. They have a better prognosis than disseminated myeloma. Prior to this report, only one case of a plasmacytoma involving the gall bladder had been reported. We describe a 76-year-old man with a plasmacytoma arising from the gall bladder and associated with an IgA monoclonal paraproteinemia. After 4 months of radiotherapy and chemotherapy, the tumor decreased markedly in size and the serum IgA value returned to normal. (J Intern Med Taiwan 2006; 17: 114-117)

Key Words : Plasmacytoma, Gallbladder

Introduction

Extramedullary plasmacytoma is a low grade malignant neoplasm with a relatively good prognosis. Isolated plasmacytomas are rare, comprising only 4% of all plasma cell malignancies. The first report of an extramedullary plasmacytoma was in 1905¹. Since then, many cases of this entity have been reported.

The most common site of extramedullary plasmacytoma is the upper respiratory tract, accounting

for 80% of all such tumors². Primary plasmacytomas of the liver and gall bladder are rare. We found only one case involving the gall bladder in the literature³. Here we report an unusual case of a locally advanced extramedullary plasmacytoma of the gall bladder.

Case Report

A 76-year-old man was admitted in October 2004 with a two-week history of epigastric pain and periodic vomiting. Physical examination revealed tenderness over the right upper quadrant. Abdominal

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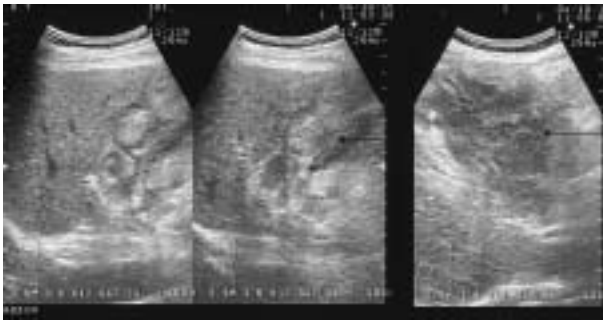


Fig.1. Abdominal echogram revealing a heterogenous mass in the gallbladder, and a hypoechoic mass just below the gallbladder.

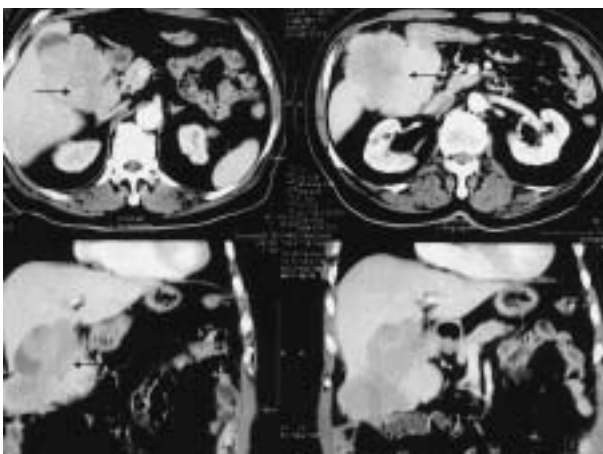


Fig.2. Computed tomography revealing a soft tissue mass seemed arising from the gallbladder with invasion of the adjacent duodenum, colon, and liver

echogram (Figure 1) and computed tomography (CT) (Figure 2) demonstrated a soft tissue mass probably arising from the gallbladder. There was peri-gallbladder infiltration leading to poor demarcation of the second portion of duodenum and the anterior aspect of the colonic hepatic flexure. This was interpreted as a probable gall bladder tumor invading the adjacent liver bed, second portion of the duodenum, and hepatic flexure. The hemoglobin was 12.4 g/dL. Liver function tests were within normal limits. Serum protein and albumin were 2.9 g/dL and 5.2 g/dL, the A/G ratio was 1.3. Serum CA199, serum AFP and CEA were 0.20 u/mL, < 2.76 ng/mL and 1.31 ng/mL.

A needle biopsy of the gallbladder tumor was performed. The biopsy specimens of the tumor revealed diffuse infiltration by uniform mature plasma cells with mild atypia, some with eccentric nuclei.

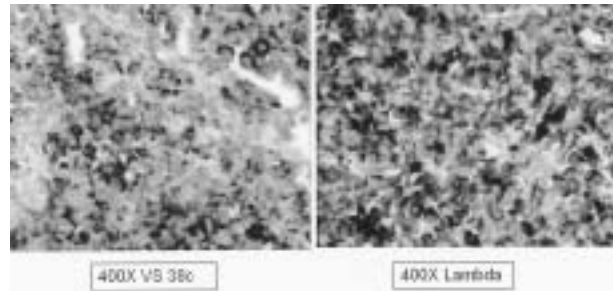


Fig.3. Positive VS38 (3A) and immunostaining for lambda chains (3B) VS38, which is a rough endoplasmic reticulum-associated protein of 64 kilodaltons present in normal and neoplastic plasma cells; it is reactive with an antigen expressed strongly on normal and neoplastic plasma cells but is absent from haemopoietic cells of other lineages. This picture showing positive staining of the malignant cells (brownish area), and the tumor is consist of plasma cells with lambda light chain restriction.

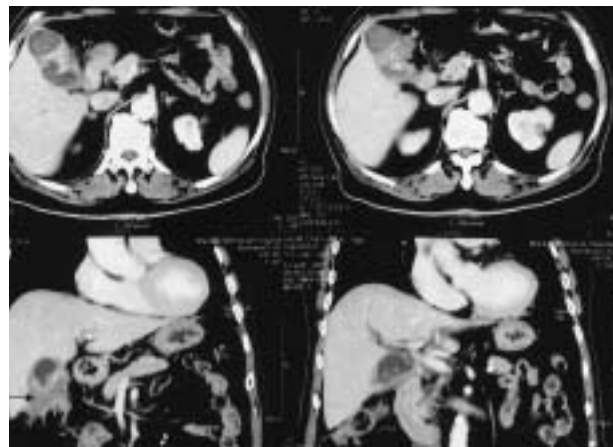


Fig.4. Marked regression of the tumor on CT one month after radiotherapy

Benign liver tissue was not found. The tumor had positive immunostaining for lambda chains (Figure 3B), vimentin, CD79a and VS38, characteristic of plasma cells (Figure 3A).. It was negative for kappa chains, CK, MPO, and CD20. No centrocyte-like cells or follicular structures were observed, nor were lymphoepithelial cells found on careful examination of cytokeratin-stained slides. There was some amyloid accumulation. Serum protein electrophoresis showed increased beta-globulins at 28.7%. Serum immunoelectrophoresis revealed an IgA lambda monoclonal gammopathy. Urinary excretion of lambda light chains was within normal limits. Skeletal radiographs (including skull, spine, pelvis, and long bone) were

normal. Bone marrow biopsy was performed which revealed normal cellular components with no increase in the number of plasma cells. In light of the monoclonal plasma cell infiltrate in the tumor and the absence of bone marrow involvement, the patient was diagnosed with a primary plasmacytoma of the gall bladder with invasion of the adjacent liver bed, second portion of the duodenum and the hepatic flexure.

As the size and location of the tumor did not permit complete resection, the patient underwent local radiotherapy and oral chemotherapy (melphalan and prednisolone). The patient had one episode of neutropenic fever, with the low white count attributed to melphalan⁴. The white count returned to normal after the chemotherapy was stopped. CT one month after radiotherapy was completed revealed marked regression of the tumor (Figure 4). The serum IgA levels returned to normal.

Discussion

Gallbladder carcinomas, accounting for 98% of all gallbladder malignancies, are epithelial in origin, with 90% being adenocarcinomas. The remainder are sarcomas, lymphomas, carcinoid tumors, metastases, and other unusual malignancies⁵. The differential diagnosis for a mass in the gallbladder fossa includes hepatocellular carcinoma, cholangiocarcinoma, and metastases. The radiologic findings in early-stage malignancies are subtle and may mimic those of acute or chronic cholecystitis. Direct extension to the liver and biliary tree, biliary obstruction, lymph node metastases, and hematogenous metastases are characteristic findings of advanced malignancies⁶. There was nothing in the radiologic appearance of our patient's tumor that suggested plasmacytoma. Biopsy is the only way to make this diagnosis⁷⁻⁸, as we did in our patient. Biopsy is still necessary in the diagnosis of gall-bladder masses in combination with normal tumor marker, especially in the differentiation of adenocarcinoma from curable lymphomas and carcinoid tumors. Myeloma may affect the hepatobiliary sys-

tem through biliary obstruction by plasmacytomas present in the pancreas or main biliary duct⁷⁻⁸ or by amyloid deposition⁹⁻¹⁰. The histological diagnosis of solitary plasmacytoma of the liver may be relatively difficult. The lack of spindle cells, histiocytes, and lymphocytes between the plasma cells is suggestive of plasmacytoma. Immunohistochemistry and molecular studies may be needed to demonstrate the monoclonal nature of the tumor, so that an unequivocal diagnosis can be made.

The only other reported case of an extramedullary plasmacytoma in the gall bladder was in a patient who had had a solitary plasmacytoma of the maxillary sinus. This had been treated with radiotherapy, but two years later, the patient developed a plasmacytoma in the gall bladder². Our patient's tumor, however, apparently arose *de novo* in the gall bladder, as we found no other evidence of disease elsewhere.

About 15% of patients with an extramedullary plasmacytoma eventually develop multiple myeloma³. Locally directed treatment, such as resection or radiotherapy with further long term monitoring, is appropriate for these patients. It will be important to follow carefully the patient discussed in this report, even though he had a good response to treatment.

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膽囊之骨髓外漿細胞瘤：一病例報告

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摘 要

骨髓外漿細胞瘤並不常見，相對於瀰漫性個骨髓瘤而言，骨髓外漿細胞瘤的愈後較佳。在本報告之前，僅有一例報告過侵犯膽囊的骨髓外漿細胞瘤。本報告提出一名76歲的男性，自膽囊生出骨髓外漿細胞瘤並合併IgA單株副蛋白血症（monoclonal paraproteinemia）。經過四個月的放射線及化學治療，腫瘤明顯變小且血清中IgA值也恢復正常。