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Extramedullary Plasmacytoma Imitating Neoplasm of the Gallbladder Fossa after Cholecystectomy

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A B S T R A C T

Extramedullary plasmacytomas are plasma cell tumors that arise outside of the bone marrow. They account for approximately 3% of plasma cell neoplasms and are most frequently located in the head and neck region. Five months after undergoing cholecystectomy, a 69-year-old patient presented with the pain under the right costal margin and a 12 kg weight loss. Computed tomography of the abdomen demonstrated irregular, vascular mass in the gallbladder fossa that dents towards the duodenum and the pylorus and lowers caudally to the hepatic flexure. His laboratory tests indicated normocytic anemia and showed elevated sedimentation rate. During operative procedure, a tumorous mass in the gallbladder fossa was found, inseparable of the peritoneum of the hepatoduodenal ligament and the IVb liver segment. Histopathological examination and immunohistochemical staining determined the diagnosis of the plasmacytoma. Total resection of the tumor was achieved and after 24-month follow-up patient showed no signs of local recurrence or dissemination of the disease.

Key words: extramedullary, plasmacytoma, gallbladder, cholecystectomy

Introduction

Plasma cell neoplasms, a group of entities characterized by the neoplastic proliferation of a single clone of plasma cells, can present as solitary plasmacytoma, multiple myeloma or plasma cell leukemias. Solitary plasmacytomas are most frequently found in the bones, but can also present as extramedullary plasmacytomas (EP), a extraosseous plasma cell tumors occurring in the absence of systemic signs^{1–2}. Eighty percent of EP are located in the head and neck region, mainly in the upper aerodigestive tract, but may also occur in the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, and skin. EP represent 3% of plasma cell neoplasms with the median age at diagnosis being 55 to 60 years. Men are more commonly affected than women³.

We present a rare case of EP arising from the gallbladder fossa, a localization that, to our knowledge, has not yet been described, in a patient who has underwent cholecystectomy five months prior to the diagnosis of the tumor.

Case Report

A 69-year-old man was admitted to the hospital in March 2008 for the elective cholecystectomy. Two months before admission, he came to our emergency department presenting with nausea and pain under the right costal margin. Laboratory tests revealed leukocytosis of $11 \times 10^9/L$, C-reactive protein 12 mg/L, hemoglobin level 126 g/L, while the rest of the tests were within the normal limits. Ultrasound revealed stones in the gallbladder. He was diagnosed with cholecystolithiasis. Antibiotic was introduced in the therapy. His medical history included hypertension, hyperlipidemia, chronic gastritis and pain in the lumbosacral region. Family history was negative for any type of malignancy.

Before performing a laparoscopic cholecystectomy, patient underwent colonoscopy, because of the 9 kg weight loss during a two month-period and a low hemoglobin level (Hg 100 g/L). Colonoscopy had shown no neoplasm. Tumor markers were normal (AFP, CEA, CA 19-9, tPSA).

A laparoscopic cholecystectomy was performed. In the gallbladder, six stones were found, up to 5 mm in size. The histopathological examination showed chronic inflammatory infiltration of the gallbladder wall.

On the third postoperative day, the patient complained of the abdominal pain under the right costal margin and later of the chest pain. Chest x-ray, abdominal ultrasound and lung scintigraphy detected no pathological condition. However, electrocardiogram detected atrial fibrillation and cardiologist was consulted. Patient was diagnosed with unstable angina pectoris. Nitroglycerine and low molecular weight heparin were introduced in the therapy. Chest pain disappeared, but the abdominal pain remained, especially under the right costal margin. On the 7th postoperative day, computed tomography (CT) of the abdomen and pelvis with the peroral contrast revealed small biloma (2×2cm) in the gallbladder fossa.

In the next few days, the abdominal pain disappeared; laboratory tests were within the normal limits. Biloma was reevaluated with the ultrasound showing the signs of regression.

On the 13th postoperative day, the patient was discharged from the hospital.

Five months later, he was readmitted, because of the pain under the right costal margin and a 12 kg weight loss. Sedimentation rate was high (45mm/h), red blood count decreased $3.6 \times 10^{12}/L$, hemoglobin level 74 g/L, MCV 84.4 fL, creatinine 92 $\mu\text{mol}/L$, total serum protein 64 g/L, Ca 2.24 mmol/L. Urine analysis as well as other laboratory tests were within the normal limits. Tumor markers were normal (AFP, CEA, CA 19-9, tPSA). CT scan of the abdomen demonstrated irregular, vascular mass in the gallbladder fossa that dents towards duodenum and pylorus and lowers caudally to the hepatic flexure.

Explorative laparotomy was performed. During operative procedure a tumorous mass in the gallbladder fossa was found, inseparable of the peritoneum of the hepatoduodenal ligament and IVb liver segment, 7.2×5.8×4 cm in size. We removed the tumorous mass, performed peritonectomy of the hepatoduodenal ligament, lymphadenectomy of the associated lymph nodes and a partial resection of IVb segment of the liver. Intraoperatively, a piece of the tumor was sent *ex tempore* for a histopathological analysis. Description was carcinomatous cells, but pathologist could not determine the origin of the malignant cells.

Postoperatively, histopathological finding was nodular tumor, consisting of small, medium and interspersed gigantic plasma cells. Tumor had positive immunostaining for kappa chain, CD138, MAM1 and CD43. It was negative for CD20, CD3, CD5, CD10, ALK, CD15, CD30, bcl2, bcl6 and CD79A. The diagnosis was plasmacytoma.

Bone biopsy revealed normal cellular components and after immunohistochemical staining, CD138 positive plasma cell counted for less than 5% of overall cells.

Bone marrow aspiration showed 0.15% cell with CD138 positive phenotype considering the total number of aspirated cells.

On the second postoperative day, patient was transferred to the intensive care unit, because of paroxysmal atrial fibrillation.

On repeated immunofixation no monoclonal gammopathy was found. Serum calcium was within the normal limits. Repeated urine analysis was negative for the free light chains. Radiological analysis had shown a small lytic lesion of the pelvis.

The patient showed no signs of local recurrence or dissemination of the disease after 24-month follow up and we continue to monitor our patient.

Discussion

The patient we presented had typical signs of cholelithiasis. Cholecystectomy was performed, several stones in the gallbladder were found and chronic inflammatory changes of the gallbladder wall were histopathologically confirmed.

Five months later he presented with the same symptoms, pain under the right costal margin and a considerable weight loss. His laboratory tests indicated normocytic anemia and elevated sedimentation rate. Tumor markers were within the normal limits. CT scan of the abdomen demonstrated irregular, vascular mass in the gallbladder fossa that dents towards duodenum and pylorus and lowers caudally to the hepatic flexure suggestive of the neoplastic disease^{4,5}.

The differential diagnosis of the tumorous mass in the gallbladder fossa includes hepatocellular carcinoma, gallbladder carcinoma, cholangiocarcinoma, lymphadenopathy and metastasis. After the surgical removal of the tumorous mass, histopathological examination and immunohistochemical staining, a diagnosis of plasmacytoma was determined^{6,7}.

After the tumor was biopsy-proven, subsequent workup confirmed the diagnosis – bone marrow aspirate and biopsy contained no clonal plasma cells, there was no hypercalcemia or renal insufficiency that could be attributed to a clonal plasma cell proliferative disorder. Although he was diagnosed with anemia and a small lytic lesion of the pelvis was found, they were not attributed to the underlying disease.

Eighty percent of extramedullary plasmacytomas occur in the head and neck region, mainly in the upper aerodigestive tract, while the involvement of the hepatobiliary system is rare^{8,9}.

To our knowledge, a localization in the gallbladder fossa has not yet been described.

For extramedullary plasmacytoma, if feasible, complete surgical removal should be considered^{2-3,9-10}.

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EKSTRAMEDULARNI PLAZMOCITOM KOJI IMITIRA TUMOR LOŽE ŽUČNJAKA NAKON UČINJENE KOLECISTEKTOMIJE

S A Ž E T A K

Ekstramedularni plazmocitomi su rijetki tumori plazma stanica, najčešće lokalizirani u regiji glave i vrata. Pet mjeseci nakon što je bio podvrgnutolecistektomiji, 69-godišnji pacijent se prezentirao sa bolovima pod desnim rebrenim lukom i gubitkom na težini od 12 kg. CT abdomena je pokazao nepravilnu, vaskulariziranu tvorbu u loži žučnog mjehura koja se protezala prema duodenumu i pylorusu te spuštala kaudalno do hepatalne fleksure. Njegovi laboratorijski nalazi su ukazivali na normocitnu anemiju i povišenu sedimentaciju eritrocita. Tijekom operativnog postupka nađena je tumorska tvorba u loži žučnog mjehura neodvojiva od peritoneuma hepatoduodenalnog ligamenta i IVb segmenta jetre. Histopatološkim nalazom i imunohistokemijskim bojenjem postavljena je dijagnoza plazmocitoma. Postignuta je potpuna resekcija tumorskog tkiva te nakon 24 mjeseci praćenja bolesti pacijent ne pokazuje znakove lokalnog recidiva ili diseminacije bolesti.