

Letters to the Editor

Primary leiomyosarcoma of the gallbladder

Key words: Sarcoma. Leiomyosarcoma. Gallbladder neoplasm. Radical cholecystectomy.

Dear Editor,

Leiomyosarcoma is an uncommon subgroup of primary gallbladder sarcomas whose diagnosis requires immunohistochemical (IHC) studies. We report two cases of primary gallbladder leiomyosarcoma (GL).

Case report 1

A 79-year-old male patient presented with right upper quadrant abdominal pain radiating to the back, anorexia and weight loss of one month' duration. His past medical history included chronic atrial fibrillation and diabetes mellitus. He was diagnosed of urinary tract infection and treated with broad spectrum antibiotics. An abdominal computed tomography (CT) scan was performed because of the persistent symptomatology, which showed a distended gallbladder with perivesicular abscess and a CT-guided percutaneous drainage was performed. A few hours later, laparotomy was required due to the development of septic shock. Surgical findings were a gallbladder mass, enlarged lymph nodes in the liver hilum and bilobar liver metastases. He died 24 hours later due to multiple organ failure. Histopathology revealed a spindle cells tumour which, after ultrastructural and IHC studies, was diagnostic of GL.



Fig. 1. Abdominal CT scan: a 7 cm mass in IV-V liver segments that includes a calcium density imaging (red arrow). Intestinal obstruction development.

Case report 2

An 81-year-old-female patient presented with abdominal pain, bile vomiting and constipation of one week' duration. Her past medical history included cholelithiasis, hiatal hernia and depression. CT scan revealed a mass in the IV-V liver segments suspected of gallbladder carcinoma, with involvement of the duodenum and hepatic flexure of the colon and low-grade intestinal obstruction (Fig. 1). A laparotomy with double by-pass (gastrojejunostomy and ileotransversostomy) and biopsy of the mass were performed. She died one week later from multiple organ failure. Histopathology showed a spindle cells tumour with low mitotic count which, after ultrastructural and IHC studies, was diagnostic of GL.

Discussion

Primary sarcoma of the gallbladder is an uncommon entity (1.4/1,000 malignant gallbladder tumors and primary GL 7% of them). The first descriptions were reported by Griffon and Segall in 1897, and Landsteiner in 1904. Since then less than 200 cases, including our own, have been reported (1,2).

These are more common in women (ratio 5:1) in the 5th-6th decades of life; however, some cases such as ours, appear later.

Preoperative diagnosis is very difficult due to nonspecific symptoms (abdominal pain, anorexia, weight loss, palpable mass, fever, jaundice, intestinal obstruction) and radiological findings (distended gallbladder with thickened wall and liver mass) (3,4).

Hematoxylin-eosin staining (spindle cell population with oval nucleus and eosinophilic cytoplasm arranged in a fascicular pattern) and IHQ studies (positive staining to smooth muscle actin and vimentin and negative to cytokeratins AE1/A3, CD117, CD34 and S-100 protein) are required to establish the diagnosis and differentiate it from other mesenchymal tumors (1-3).

Radical cholecystectomy (cholecystectomy plus lymphadenectomy, liver segments IVb-V resection to right hepatectomy and/or infiltrated organs resection) seems to be the best approach. However, many authors recommended palliative surgical by-pass due to its poor prognosis, with liver involve-

ment in almost 75% of cases and a five year survival rate of less than 5%. Adjuvant chemotherapy (doxorubicin, mitomycin C) has prolonged the survival rate in some cases (3-5).

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