

Gallbladder Involvement in Aggressive Systemic Mastocytosis

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To the editor,

A 48-year-old male patient with a 10-month history of systemic mastocytosis, confirmed positive for the KIT D816V mutation, presented with right upper quadrant abdominal pain. Due to hepatic involvement and resulting portal hypertension, he was being monitored under a diagnosis of aggressive systemic mastocytosis (ASM) and had been receiving cladribine therapy for the past three months. His medical history included previous episodes of cholecystitis and biliary pancreatitis. Abdominal ultrasonography revealed thickening of the gallbladder wall, multiple millimetric gallstones, and biliary sludge within the lumen. The common bile duct was measured at 7.5 mm in diameter, and the intrahepatic bile ducts were more prominent on the left side. Elective laparoscopic cholecystectomy was performed due to recurrent episodes of cholecystitis.

Intraoperative exploration revealed thickening of the gallbladder wall, cystic duct, and common bile duct. A relative narrowing of the cystic duct lumen was observed. The cystic duct could not be closed using endoclips or Hem-o-lok clips; therefore, it was successfully ligated with two interrupted sutures using polyglactin. The cholecystectomy specimen was opened along the lumen for examination. The lumen was markedly narrowed, and the mucosa appeared spongy, with a maximum wall thickness of 0.7 cm. Additionally, a 0.9 cm lymph node was identified near the neck region. Histopathological examination of the gallbladder revealed dense infiltration of atypical mast cells in the lamina propria and muscularis layers (Figure 1); these cells were diffusely positive for CD117. The lymph node also demonstrated mast cell infiltration (Figure 2).

In systemic mastocytosis, the primary pathological feature is the clonal infiltration of various internal organs, including the bone marrow, by mast cells. ASM is a rare but clinically severe subtype of systemic mastocytosis, defined by less than 20% mast cell infiltration in the bone marrow and the presence of at least one C finding. C findings indicate organ dysfunction caused by excessive mast cell infiltration. Mast cell accumulation can occur in the skin, liver, spleen, bone marrow, or gastrointestinal tract (1). Despite the relative frequency of gastrointestinal involvement in systemic mastocytosis (2,3), gallbladder infiltration remains exceedingly rare, with only one case available in the literature to date (4). We aimed to contribute this rare case of mast cell infiltration of the gallbladder observed in a patient diagnosed with aggressive systemic mastocytosis to the literature, along with its pathological images.

References

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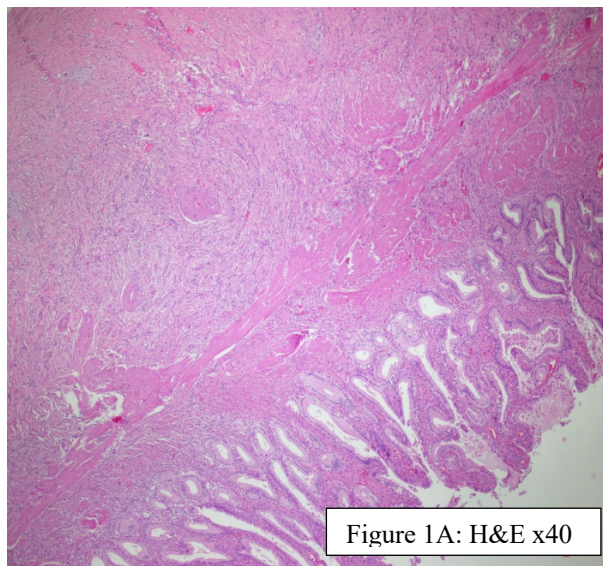


Figure 1A: H&E x40

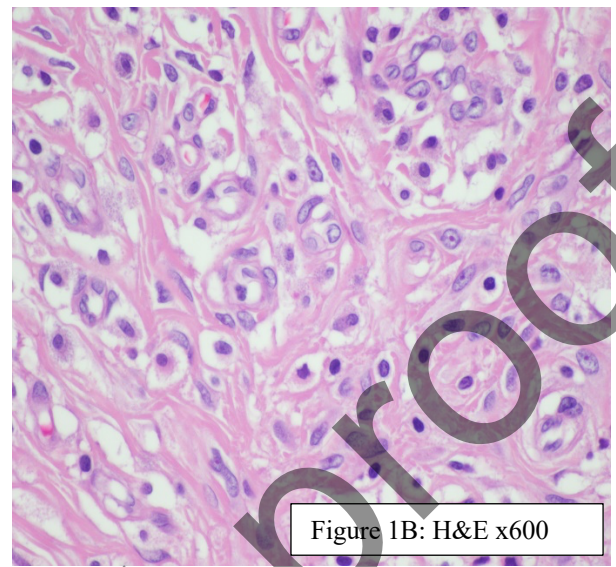


Figure 1B: H&E x600

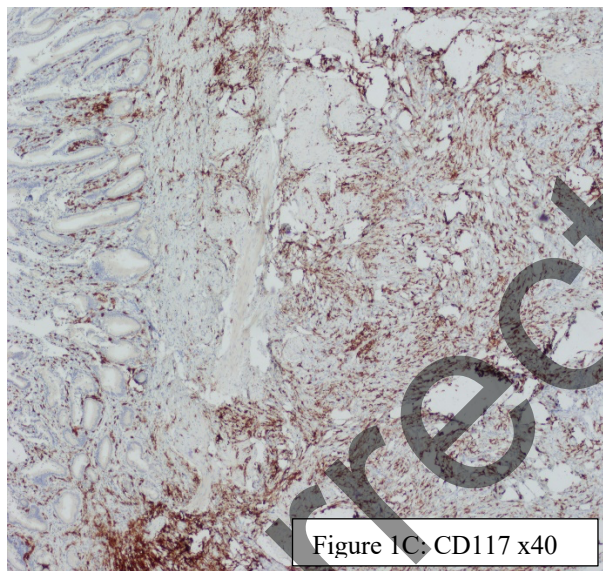


Figure 1C: CD117 x40

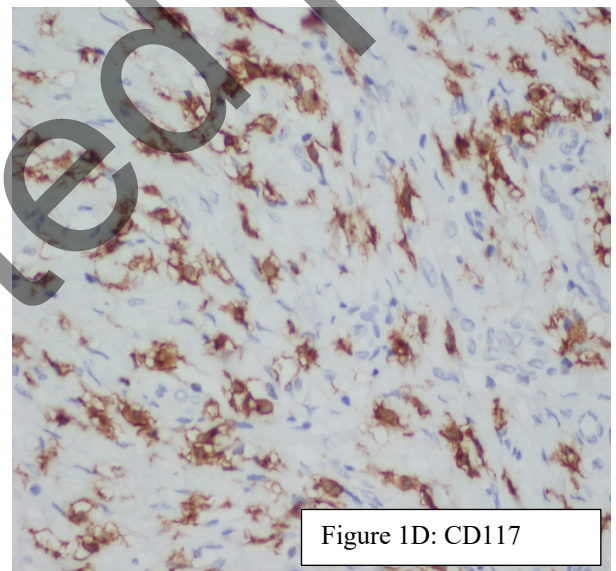


Figure 1D: CD117

Figure 1: Histopathological microscopic images of the patient's gallbladder. *A–B: H&E stain, x40 and x600; dense infiltration of atypical mast cells in the gallbladder wall. C–D: CD117, x40 and x600; diffuse membranous and cytoplasmic positivity in mast cells.*

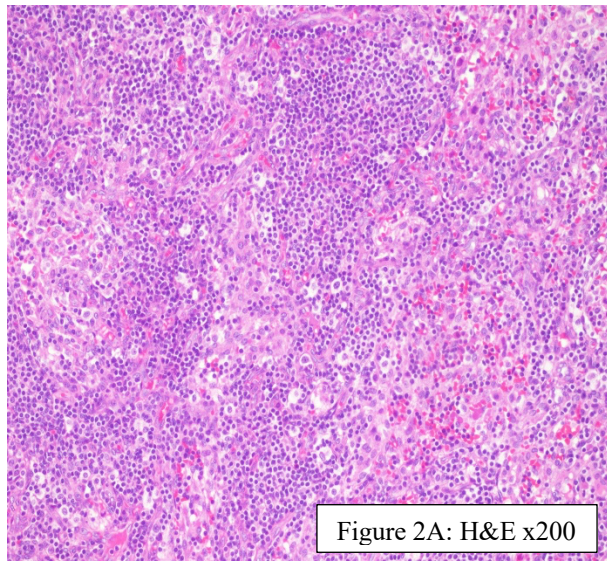


Figure 2A: H&E x200

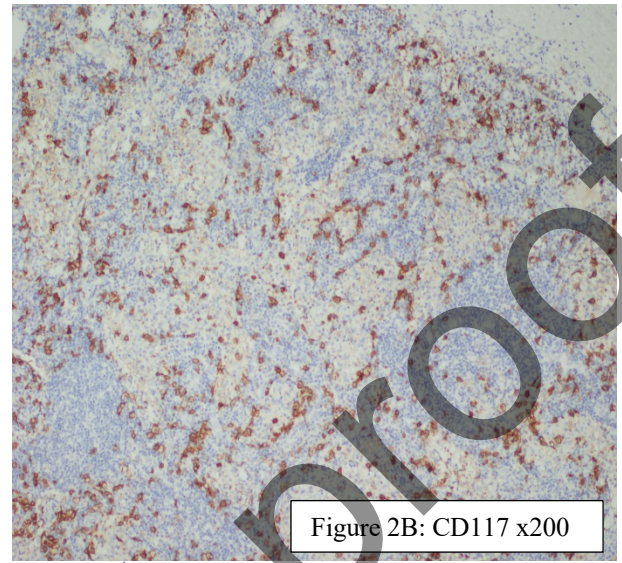


Figure 2B: CD117 x200

Figure 2: Histopathological images of the pericholecystic lymph node. *A:* Lymph node infiltration by mast cells (H&E stain, x200). *B:* CD117 staining (x200) shows mast cells with positive expression.

Uncorrected proof