

# Gastrointestinal and Liver Pathology at Rush

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## Case of the Month Answer – January 2008

*Contributed by Drs. Ajay Patel and Shriram Jakate*

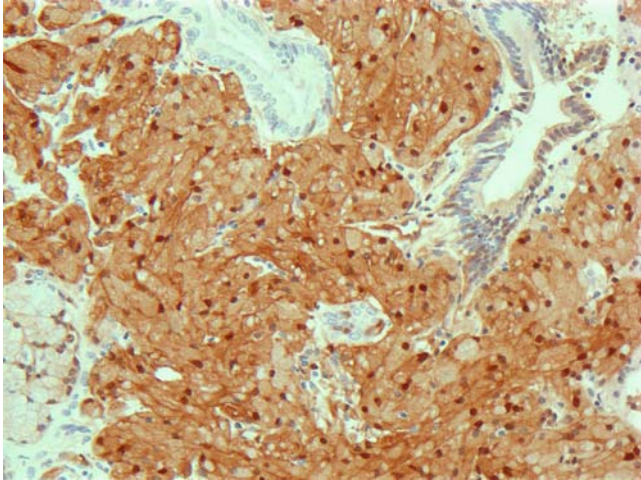


Figure 4. Microphotograph of the S-100 immunohistochemical stain positivity within the infiltrate in the bile duct.

### **Diagnosis: Granular cell tumor of the extrahepatic bile duct and secondary biliary cirrhosis of the liver.**

The monomorphic infiltrate within the lamina propria of the bile duct showed sheets and clusters of large, ovoid to round cells separated by thin fibrous connective tissue septa. The cells had abundant granular eosinophilic cytoplasm and small uniform hyperchromatic nuclei with minimal pleomorphism and no mitotic activity or necrosis. The cells were positive with periodic acid-Schiff stain followed by diastase digestion and were also strongly positive for S-100 immunohistochemical stain. Based on the morphology and ancillary studies, the diagnosis of granular cell tumor was rendered. The patient is doing well 19 months later with no graft failure, cholestasis, or evidence of recurrence.

Granular cell tumors are benign lesions usually occurring within the dermis and subcutaneous tissues. Approximately 5-9% of cases have been reported within the gastrointestinal tract with less than 1% of the cases occurring within the biliary tree. Within the biliary tree, the majority of the cases occur in black women with a median age of 31 years. Typically, patients present with abdominal pain and/or jaundice. Usually, due to the rarity of the disease, the patients are misdiagnosed with another entity such as a choledochal cyst due to characteristic radiologic findings of intra and/or extrahepatic dilatation. However, many conditions obstructing this narrow lumen structure can create the clinical picture of a choledochal cyst. The differential diagnosis of such obstructive lesions include cholangiocarcinoma, primary sclerosing cholangitis, biliary stricture, polyps, papillomas, adenomas, choledochal cysts, and infrequently granular cell tumors. Microscopically, these tumors are composed of polygonal cells with granular eosinophilic cytoplasm and small vesicular nuclei. These granules within the cytoplasm react with periodic-acid Schiff and S-100 staining. They appear as cluster or sheets and infiltrate diffusely within the surrounding tissue separated by thin fibroconnective tissue septa. Typically, mitoses are rare with no necrosis present. The accepted treatment for granular cell tumors in this location is surgical excision with tumor-free margins followed by

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hepaticojejunostomy. Only 1 transplant has been described thus far due to severe liver failure and secondary biliary cirrhosis in a patient over a 5 year period.

### References:

1. Te Boekhorst DS, Gerhards MF, Van Gulik TM, Gouma DJ. Granular cell tumor at the hepatic duct confluence mimicking Klatskin tumor: a report of two cases and a review of the literature. *Digestive Surgery*. 2000; 17: 299-303.
2. Fairchild RB, Freeman R, Salah Hammad EM, Rohrer R. Granular cell tumor with cirrhosis and transplantation. *Transplantation*. 1999; 68 (2): 315-317.