

Treated Rosai-Dorfman Disease: An Unusual Cause of Biliary Obstruction



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We report a case of a 50-year old woman with Rosai-Dorfman disease who presented five years prior to admission with vague abdominal pain secondary to bulky peripancreatic lymphadenopathy mimicking lymphoma on CT scan. She was treated successfully with chemotherapy. Five years later she presented with right upper quadrant pain, fevers and progressive jaundice. Imaging demonstrated intrahepatic and extrahepatic biliary ductal dilatation with an appearance of primary sclerosing cholangitis, cholelithiasis, and recurrent small upper abdominal lymph nodes. Abdominal MRI revealed discrete abnormal peripancreatic signal consistent with fibrosis which we believe reflected healed Rosai-Dorfman disease leading to intractable biliary obstruction. There are no cases to our knowledge of biliary obstruction related to this uncommon disease.

INTRODUCTION

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare idiopathic disorder of histiocytic proliferation. According to a review of the entity

by Foucar et al (1), the typical patient profile is a 20-year-old white or black male who presents with painless bilateral cervical lymphadenopathy. The clinical course is variable ranging from self-limited and indolent with spontaneous regression to, less commonly, aggressive and fulminant. A large number of patients

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have immunologic abnormalities. The disease has been reported to involve almost every organ of the body. Although it commonly involves cervical lymph nodes and not infrequently other nodal groups, it is uncommon in the abdomen. Biliary obstruction due to this disease has not been described. We encountered a patient who developed biliary obstruction five years after successful chemotherapy of extensive peripancreatic SHML.

CASE REPORT

A 50 year-old black woman was admitted to our hospital with right upper quadrant pain, vomiting, chills, and night sweats increasing over a two week period and progressive jaundice. Her past medical history included rheumatic mitral stenosis, atrial fibrillation, congestive heart failure and type II diabetes mellitus. She had a history of vague abdominal pain five years prior to admission, at which time bulky peripancreatic (Fig. 1A) and lesser omental lymphadenopathy was documented by CT scan. A laparoscopic biopsy specimen of the lesion at that time was diagnostic of Rosai-Dorfman disease. The patient improved after treatment with a combination of Cyclophosphamide, Vincristine and Prednisone, and later with Methotrexate and Prednisone. Seven years prior to admission, she had exci-

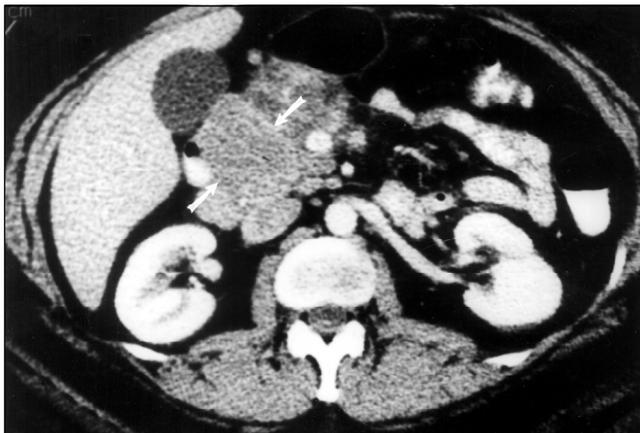


Figure 1A. Contrast enhanced CT scan of the abdomen five years prior to admission shows a dominant lobulated peripancreatic soft tissue mass (white arrows) and other enlarged peripancreatic lymph nodes. Duodenum is stretched laterally around the mass.



Figure 1B. Abdominal sonogram reveals an enlarged common bile duct measuring 1 cm in caliber (white arrow).

sion of a large painless right submandibular mass that was initially diagnosed as an inflammatory pseudotumor. Later review of the pathology confirmed the diagnosis of Rosai-Dorfman disease.

On physical exam, the patient had jaundice, low-grade fever, right upper quadrant tenderness, and a Murphy's sign. Pertinent laboratory data included: white blood cell count of 1600, hemoglobin of 9.4, hematocrit of 27, total bilirubin of 15.4, direct bilirubin

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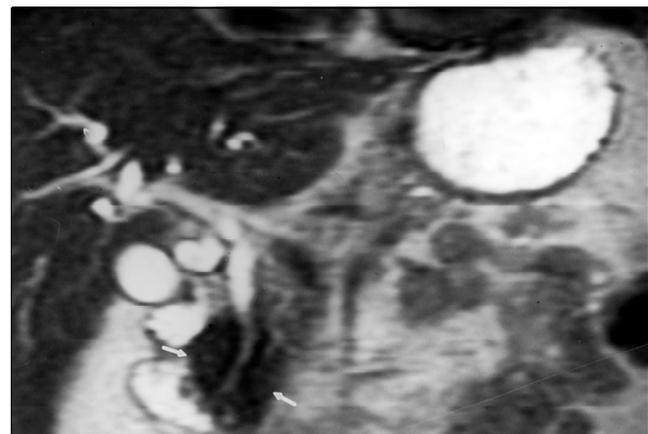


Figure 1C. MRI of the abdomen in coronal plane demonstrates markedly hypointense peripancreatic tissue (white arrows) on SSFSE sequence. Axial images (not shown) demonstrate normal signal in the pancreas proper.

(continued from page 27)

bin of 10.0, alkaline phosphatase of 649, AST of 66, and ALT of 120. Sequential serum multi-analysis-7 was normal.

Abdominal sonogram revealed intrahepatic and extrahepatic ductal dilatation (Fig. 1B) and cholelithiasis. Visualization of the pancreas was limited. CT scan of the chest, abdomen and pelvis demonstrated a minimal residual peripancreatic soft tissue density, much smaller than on previous CT scan five years earlier. There was, however, recurrent periportal and lesser omental lymphadenopathy and new mediastinal lymphadenopathy. MRI of the abdomen (Fig. 1C) demonstrated small peripancreatic tissue, which was isointense to pancreas on T1, markedly hypointense on T2 and nonenhancing with Gadolinium DTPA. The appearance was consistent with fibrosis. MR cholangiogram (Fig. 1D) revealed narrowing of the most distal common bile duct and proximal pancreatic duct, a double duct appearance. There was mild intrahepatic ductal dilatation with a beaded appearance due to multiple strictures. A dominant stricture was present in the common hepatic duct at the level of the cystic duct. ERCP (Fig. 1E) revealed multiple small stones (2–3 mm in size) in the common bile duct which were not discerned on MRCP, probably due to their small size. The stones were extracted and a biliary stent placed. Otherwise, the ERCP demonstrated similar findings as

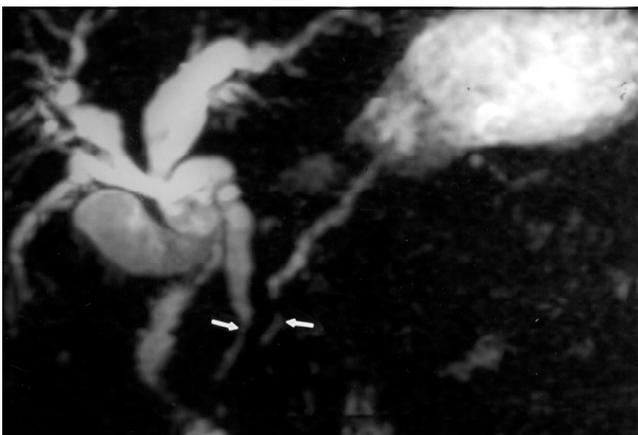


Figure 1D. MRCP shows focal narrowing (white arrows) of the distal common bile duct and proximal pancreatic duct (double duct appearance). Areas of stricturing and focal widening are present in the common hepatic duct and intrahepatic biliary tree.

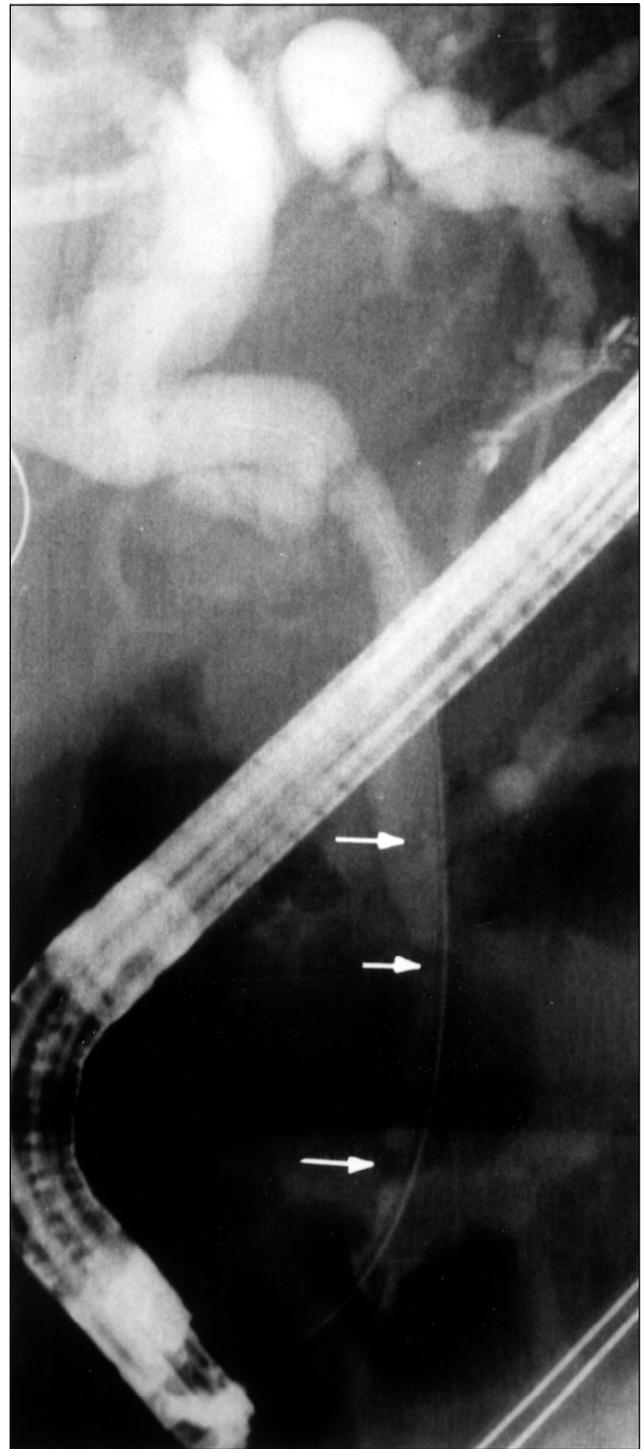


Figure 1E. ERCP shows similar findings as MRCP except that multiple small stones are visualized in the common bile duct (white arrows). The proximal pancreatic duct is narrowed and the remainder dilated.

Treated Rosai-Dorfman Disease: An Unusual Cause of Biliary Obstruction

A CASE TO REMEMBER

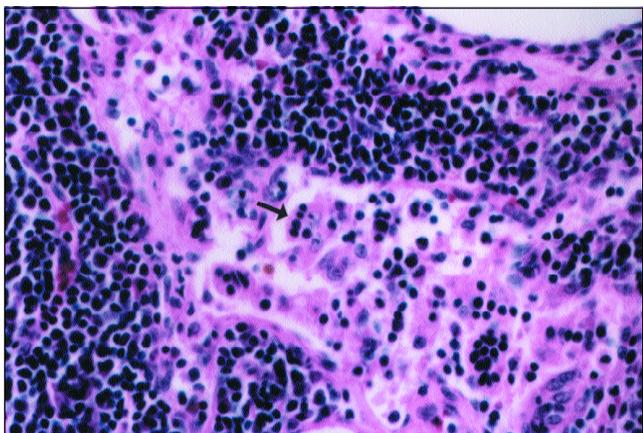


Figure 1F. Cystic duct lymph node contains dilated sinuses with histiocytosis. Black arrow marks a histiocyte nucleus (light stained) with numerous intracytoplasmic lymphocytes (dark stained nuclei), a process known as emperipolesis. (H and E; x200)

the MRCP. After biliary stent placement, the patient had some initial improvement with decrease in right upper quadrant pain and bilirubin levels. Three days after biliary stent placement laboratories were: total bilirubin of 8.2; direct bilirubin of 4.5; alkaline phosphatase of 446; AST of 43; and ALT of 43. However, the patient's symptoms were not completely resolved and she suffered from continued abdominal pain which led to surgical therapy. She ultimately underwent cholecystectomy, choledochoduodenostomy and intraoperative periportal lymph node biopsy. Six days after surgery laboratories were: total bilirubin of 2.6; direct bilirubin of 1.4; alkaline phosphatase of 101; AST of 12; and ALT of 18. Pathology demonstrated changes of SHML in excised lymph nodes from the porta-hepatic space and peripancreatic region (Fig. 1F and G). The residual small soft tissue contiguous with the pancreatic head was not sampled. There was evidence of chronic cholecystitis and cholelithiasis with mild acute and chronic inflammatory changes of the common bile duct wall. There was no pathologic evidence of SHML in the distal common bile duct wall.

DISCUSSION

According to a review of a registry of 423 patients with SHML by Foucar et al (1), the disease can involve

almost any organ, however, the majority of cases (85.3%) present with cervical lymphadenopathy. Yet, other nodal groups, including axillary, inguinal, mediastinal and miscellaneous areas are affected in a significant number of patients. Forty-three percent of patients are reported to have at least one site of extranodal involvement. The most common extranodal sites include skin, nasal cavity or paranasal sinuses, soft tissues, eyelids and orbit, bone, salivary gland and central nervous system. Occasional cases of abdominal involvement, specifically liver, intestine, adrenal, kidney and pancreas are reported. The majority of patients with extranodal disease have one to three extranodal sites involved. The presence of immunologic abnormalities, involvement of multiple extranodal organ systems and involvement of a larger number of node groups tend to correlate with an unfavorable prognosis.

We could find no references in the literature of biliary obstruction related to Rosai-Dorfman disease. Although there are many etiologies of biliary obstruction, including postinflammatory, postsurgical, sclerosing cholangitis, ascending cholangitis, extrinsic processes such as metastatic disease, lymphoma, cholangiocarcinoma, ampullary/periampullary processes and pancreatic masses (2), this case appears unique. There are only a few case reports of SHML involving the peripancreatic region (1,3,4) and only one case involving the pancreas proper (5). Esquivel et al (5), illustrate abdominal CT scan findings of a solid mass in the body of the pancreas of a 48 year old black woman. Other reports in the literature (1,3,4) describe retroperitoneal disease without detailed description of imaging findings.

Our case is therefore unique in that our patient presented initially with a CT appearance mimicking lymphoma characterized by a dominant bulky lymph node mass adjacent to the pancreatic head with associated mediastinal, hilar, axillary, lesser omental and peripancreatic lymphadenopathy. Biliary obstruction was not present. The patient's clinical course, although characterized by recurrent vague abdominal pain, was somewhat indolent. The dominant peripancreatic mass and upper abdominal lymphadenopathy present five years prior to this admission had regressed after chemotherapy. Our patient then presented with new obstructive jaundice which we believe was due to chronic extrin-

(continued on page 34)

Treated Rosai-Dorfman Disease: An Unusual Cause of Biliary Obstruction

A CASE TO REMEMBER

(continued from page 32)

sic mass effect on the distal common bile duct and not due to choledocholithiasis alone. This longstanding process probably caused stasis leading to stone formation and intrahepatic and extrahepatic biliary strictures, accounting for the appearance of sclerosing cholangitis seen on ERCP and MRCP.

MRI uncovered a distinct region of peripancreatic tissue which was markedly decreased in signal on T2 weighted images. This tissue caused the distal common duct and proximal pancreatic duct narrowing (double duct appearance) seen on both ERCP and MRCP. It probably represents healed mass replaced by fibrosis after chemotherapy. The underlying contributor to biliary obstruction in this patient probably lies in the focal peripancreatic soft tissue that remained after successful therapy for the earlier peripancreatic disease. Recall that our patient initially presented five years prior to admission with extensive peripancreatic disease but no biliary obstruction. We propose that healed SHML accounts for the abnormal dark signal peripancreatic soft tissue and led to extrinsic mass effect on the common bile duct and pancreatic duct,

mimicking a double duct sign. It is documented in the literature that as SHML regresses, fibrotic tissue replaces the affected lymph nodes (6). Therefore, even though there was no evidence of SHML involving the common duct wall pathologically, replacement of peripancreatic tissue by fibrosis probably accounted for the intractable biliary obstruction in this patient. ■

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