

An Unusual Case of Acute-on-Chronic Xanthogranulomatous Cholecystitis

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Citation

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Abstract

Xanthogranulomatous cholecystitis (XGC) is a rare but severe form of cholecystitis. In this pathology, the gall bladder is thickened and irregular with extension of yellow xanthogranulomatous inflammation to adjacent organs. This condition is extremely difficult to suspect preoperatively as it macroscopically resembles carcinoma of the gall bladder. The acute variant of this condition also is unusual. We would like to report such an unusual case at our institute.

CASE HISTORY

A 40-year-old Hindu male railway employee came with complaints of pain in the abdomen mainly the in right upper quadrant for 20 days along with fever with chills at night. The pain was colicky in nature and was not associated with vomiting. There was no history of trauma. There was a history of constipation, loss of weight and appetite. He had a history of passing high-colored urine but no history of passing clay-colored stools. There was no history of any major medical illnesses or any surgery done in past. He had no addiction or any contributory family health record.

On examination, his vital parameters were within normal limits. Per-abdominal examination showed right hypochondriac guarding and tenderness and mild hepatomegaly. The rest of the abdomen was soft. Bowel sounds were present normally. Per-rectal examination was normal.

On investigating, his total bilirubin was 4.7 (direct 3.7), AST 85 and ALT 105, Alkaline phosphatase 364 and Prothrombin time 18/14. The remaining reports all were within normal limits. Ultrasonography of abdomen and pelvis showed cholelithiasis and choledocholithiasis with a CBD of 13 mm. A decision to perform an ERCP (Endoscopic Retrograde Cholangio-Pancreatography) was taken. ERCP showed normal-sized CBD, cholangitis with sludge and no stone within. A 7-Fr stent was kept for free drainage of bile after papillotomy. Post ERCP, after a one-week course of antibiotics, the patient settled completely. Repeat investigations came back to normal range. A decision to

perform cholecystectomy was taken.

Operative findings: 1) small contracted intrahepatic gall bladder, 2) multiple yellow gall stones, 3) thickened and indurated wall, 4) minimal sludge within CBD with stent in situ and no stones.

On gross pathology, the external surface showed marked congestion and hemorrhage. The cut section showed a thickened wall and yellowish plaque-like areas, and the lumen contained yellowish stones with debris.

On microscopy, the thickened wall showed fibrosis, hemorrhage, transmural inflammation and edema. The inflammatory infiltrate showed sheets of foamy histiocytes, granulomas, foreign body-type giant cells and occasional neutrophil infiltrates. There was no evidence of dysplasia, malignancy or tuberculosis. The impression was acute-on-chronic xanthogranulomatous calculous cholecystitis.

DISCUSSION

Christensen and Ishak first coined the pathological diagnosis of XGC, describing seven patients with pseudotumor of gall bladder (1). Many other pseudonyms such as ceroid granulomas, ceroid-like histiocytosis, and fibroxanthogranulomatous inflammation existed before the Armed Forces Institute of Pathology formally characterized XGC (2).

The reported incidence of XGC varies from 0.66% to 1.8%. Pre-operative diagnosis is rarely made. Frequently, these patients are misdiagnosed as either having only cholelithiasis

or gall bladder cancer.

XGC was initially thought to be a malignant disease process, but many studies now show that this disease behaves in a similar fashion to another benign disorder, xanthogranulomatous pyelonephritis (XGPN) (3). Both of these disease processes are associated with chronic infection and calculi that induce histiocytosis to begin the inflammatory cascade.

Sjodahl first speculated that gall bladder calculi creating mechanical trauma along the mucosa incite this inflammatory response. The inflammation causes the lecithin in the bile to react with free fatty acids, thus producing lysolecithin. This results in further damage to gall-bladder mucosa, initiating a destructive cascade. This inflammatory process progresses from cholecystitis to XGC when the Rokintansky-Aschoff sinuses become occluded and inspissated with bile and mucin.

When the tension of the distended gall bladder exceeds its normal compliance, these sinuses rupture, spreading inflammation to the adjacent tissues. The degradation of bile results in phagocytosis of insoluble cholesterol and bile salts by histiocytes, giving the characteristic XGC appearance of macrophage-laden foamy histiocytes (4).

XGC has many similarities to its original counterpart (4), but it is usually more severe. Similar pathogens are found in both XGC and cholecystitis includes *Klebsiella*, *E. coli*, *Proteus mirabilis*, *Enterobacter* and *Citrobacter*. Neither XGC nor its sequelae are usually suspected pre-operatively.

Complications such as gall-bladder perforation, hepatic abscess, biliary stricture, biliary obstruction, ascending cholangitis and biliary fistulas are associated more commonly with XGC (5).

Although imaging modalities such as CT scan, Ultrasound, ERCP and percutaneous transhepatic cholangiography (PTC) aid in diagnosing the complications pre-operatively, only histological examination of a surgical specimen will accurately give the exact diagnosis of XGC.

In addition, there is increased association of underlying carcinoma in patients with XGC as compared to those with simple cholecystitis (6). Due to the high prevalence of primary fistulas, cholecystectomy along with excision of all the surrounding xanthogranulomatous tissue is required to assure complete prevention of fistulas and recurrence of XGC in the future.

Since dissection at Calot's triangle is extremely difficult due to dense adhesions, laparoscopic surgery is rarely successful in XGC. Some have advocated subtotal cholecystectomy with drainage of the subhepatic space to decrease the likelihood of iatrogenic injury at the CBD level. Also due to the high incidence of associated fistulas and malignancies, many advocate intra-operative cholangiography.

SUMMARY

XGC is an uncommon variant of cholecystitis that is rarely diagnosed pre-operatively. It is generally more virulent than simple cholecystitis, but the same pathogens are isolated from both, implicating a primarily histiocytic-driven inflammatory mechanism as the cause of this complicated sequel.

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References

1. Howard TJ, Bennion RS, Thompson JE. Xanthogranulomatous cholecystitis: A chronic inflammatory pseudotumor of the gallbladder. *Am Surg* 1991;57:821-824.
2. Reed A, Ryan C, Schwartz S. Xanthogranulomatous cholecystitis. *J Am Coll Surg* 1994;179:249-252.
3. Malek RS, Elder JS. Xanthogranulomatous pyelonephritis: A critical analysis of 26 cases and of the literature. *J Urol* 1978;119:589-592.
4. Goodman ZD, Ishak KG. Xanthogranulomatous cholecystitis. *Am J Surg Pathol* 1981;5:653-659.
5. Parna JA, Acinas O, Bueno J, et al. Xanthogranulomatous cholecystitis: Clinical, sonographic, and CT findings in 26 patients. *AJR Am J Roentgenol* 2000;174:979-983.
6. Yoshida J, Chijiwa K, Shimura H, et al. Xanthogranulomatous cholecystitis versus gallbladder cancer: Clinical differentiating factors. *Am Surg* 1997;63:367-371.

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