



## Pediatric Xanthogranulomatus cholangitis

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### ABSTRACT

The Xanthogranulomatous cholangitis is rare entity, usually it happens as an extension of Xanthogranuloma of the gall bladder (XCC) [1], although they are case reports of isolated Xanthogranulomatous cholangitis in adult age group [2] [3], To best of our knowledge this the first case of isolated Xanthogranulomatous cholangitis in pediatric age group.

The Xanthogranuloma is chronic inflammation could affect any effect any part of the body, which characterized histologically by lipid-loaded macrophages, it could spread to the surrounding structures, which make it an important differential diagnosis of the malignancy especially in adult age group [1].

The Xanthogranulomatous cholangitis happen in the most case as an extension of Xanthogranulomatous cholecystitis which caused initially by chronic inflammation and/or obstruction [4,2].

In this case report we are presenting case with isolated Xanthogranulomatous cholangitis we had recently in our institution, it was boy 9 months old boy referred from secondary level hospital with typical symptoms of obstructive jaundice (deep jaundice, pale stool, dark urine) which started gradually over 4 months, clinically liver was palpable 4 cm below the costal margin, otherwise unremarkable clinical examination.

The infectious panel were all negative (HIV, HBV, HCV, and CMV).

The sonar showed Extra-hepatic ducts distension with cutoff at the hepatic hilum, which gave the impression of short segment biliary stricture at the confluence of the common hepatic and the cystic ducts, and normal gall bladder (Fig. 1).

MRCP confirmed the sonar's finding it demonstrated the right and the left hepatic ducts, and the cutoff at the confluence of the right and left hepatic ducts, (Fig. 2).

The radiological investigations showed the reason of the obstructive jaundice is cutoff at the hepatic hilum (? Stricture or/and mass).

The decision made to precede for intra-operative cholangiogram and intervene surgically depending on intra-operative finding.

The intra-operative finding was a fibrotic mass exactly at confluence point between the right and left hepatic ducts, no infiltration to surrounding structures, and was no enlarged lymph nodes. Intra-operative cholangiogram done through the gall bladder showed proximal obstruction, (Fig. 3 a,b).

The fibrotic mass resected, cholecystectomy, and hepatoduodenostomy were done.

Patient did well postoperatively the jaundice cleared dramatically, patient discharged day 5 post-operation.

Patient seen 1 month and 10 months after the discharge jaundice cleared completely, no hepatomegaly anymore, LFT improved significantly (Table 1).

### 1. Pathological evaluation

The common bile duct microscopically was completely obliterated by inflammation (Fig. 4) with proliferation of new bile ductules (Fig. 5) penetrating the mass of the inflammations, in more magnified picture

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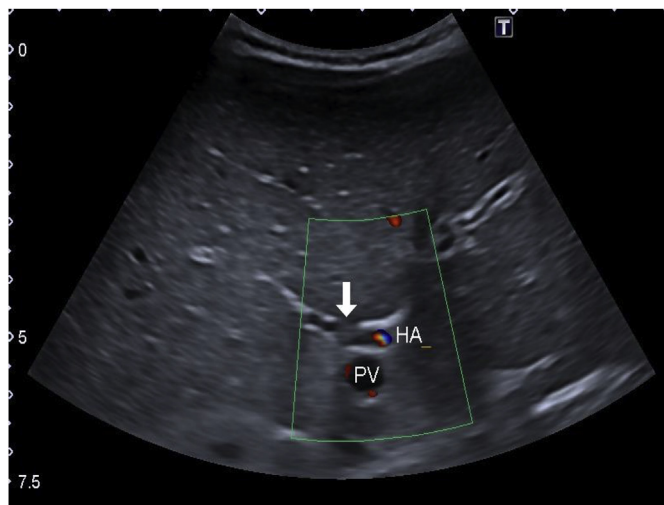


Fig. 1. Abdominal sonar, the arrow is pointing to cutoff point at the hepatic hilum.

we can appreciate the inflammation composed mainly from fat loaded macrophages (which typical for Xanthogranuloma) with scattered neutrophils (see Fig. 6).

The microscopic examination of gall bladder showed no signs of inflammation, but revealed signs of back flow pressure (flattened of papillae, Rokitansky-aschoff sinuses) (Fig. 7).

2. Discussion

Xanthogranuloma inflammation in children known as Juvenile Xanthogranuloma (JXG), it's non-Langerhans cell histiocytosis that is usually benign and self-limiting, the cause of this inflammation is not known [5].

It affects many parts of the body like skin, kidney, and gall bladder, it could spread to the surrounding structures [6].

Xanthogranuloma is an intense inflammation believed that caused

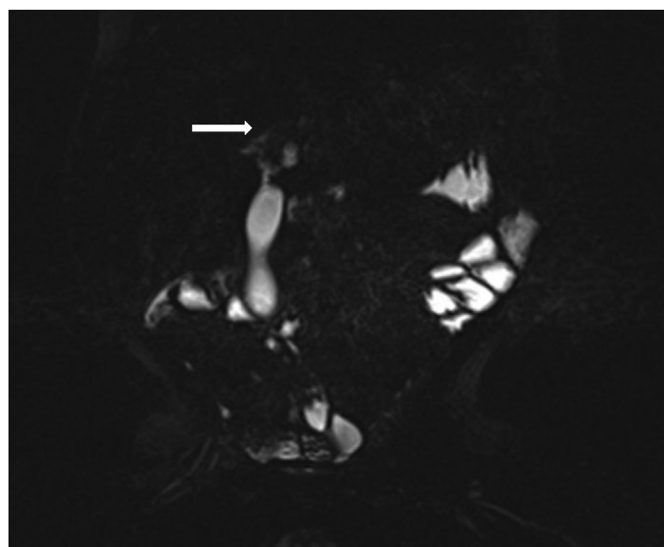


Fig. 2. MRCP demonstrated elongated gall bladder, and cutoff at the hepatic hilum-the arrow is pointing toward the level of proximal obstruction.

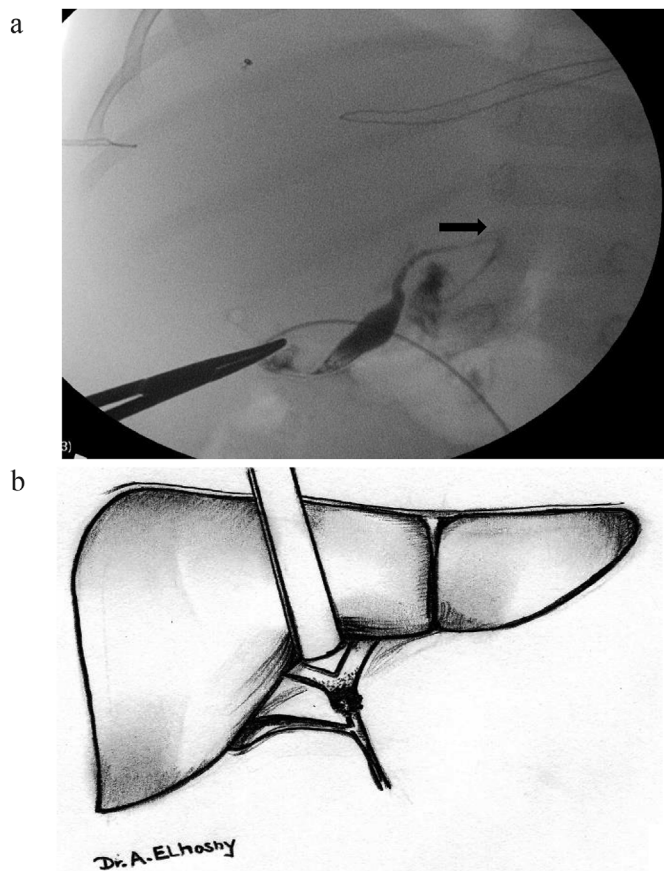


Fig. 3. A: The intra-operative cholangiogram demonstrate the proximal obstruction-the arrow points to proximal obstruction in CHD, distally the contrast drained to the duodenum. b: Sketch explain the intra-operative finding.

by obstruction and/or chronic infection, and its known ability to invade the surrounding structure, and it can mimic the malignancy especially in adult age group [1].

Xanthogranulomatus inflammatory lesions classically stains with some macrophage markers including CD68 or Ki-M1P, anti-FXIIIa, vimentin, and often anti-CD4. They are distinguished from Langerhans Cell Histiocytosis lesions by being negative for S-100 and CD1a [7].

There are few case reports of Xanthogranulomatus cholecystitis spreading to extra-hepatic biliary system in adult age group [1], and one case of infant [8].

Also there few reports of primary Xanthogranulomatous cholechochitiss whether it confined to the extra-hepatic biliary system [2,3], or invading the surrounding structure [6], To best of our knowledge this case the first case of isolated Xanthogranulomatus choledochitiss in pediatric age group.

Patient consent

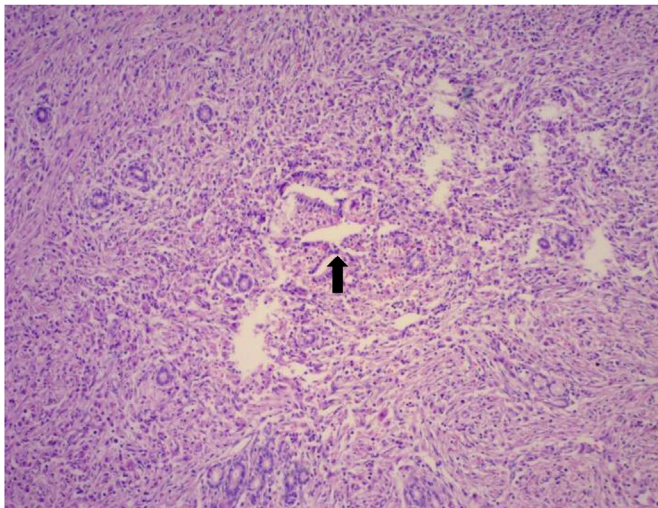
The consent to publish the case report was not obtained, this does not contain any personal information that could lead to the identification of the patient.

Funding

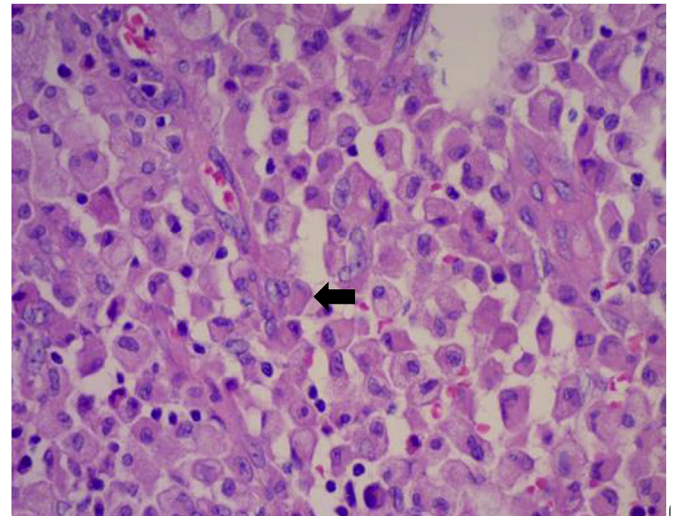
No funding or grant support.

**Table 1**  
Blood investigations showed the obstructive figure of LFT.

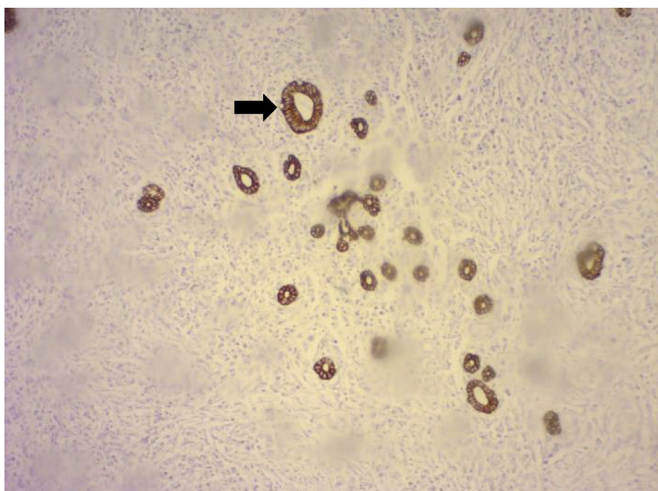
LFT Values	Total bilirubin (5–21 μmol/l)	Conjugated Bilirubin (0–5 μmol/l)	ALP (82–383 μmol/l)	GGT (1–39 μmol/l)	AST (0–65 μmol/l)	ALT (4–35 μmol/l)
Pre-surgery x 1 month	61	42	502	461	85	79
Pre-surgery x 1 day	63	54	1178	1549	128	138
post-surgery × 10 month	3	2	378	173	57	44



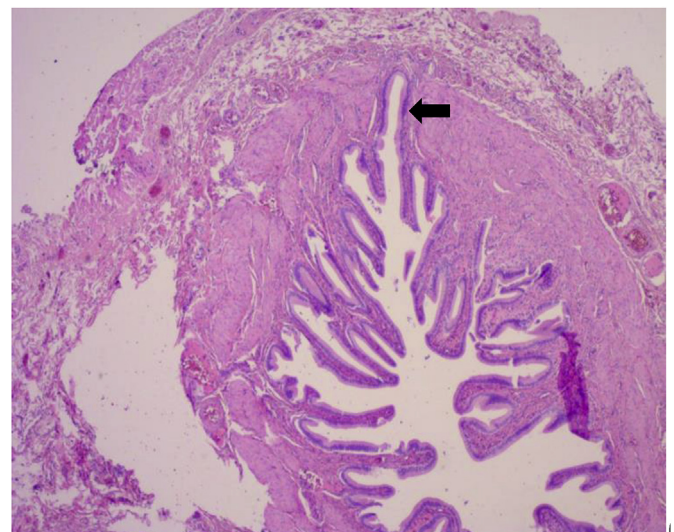
**Fig. 4.** Slide magnified ×100, showing the near total obliteration of CBD – The arrow.



**Fig. 6.** Slide magnified ×400, demonstrates fat loaded Macrophages in CBD.



**Fig. 5.** Slide of CBD stained with MFN-117 stain, which demonstrates the proliferation of small bile ductules.



**Fig. 7.** Slide of gall bladder (×100), showing signs of back flow pressure, Rokitsansky-Aschoff sinus -the arrow.

**Authorship**

All the authors attest that they meet the current ICMJE criteria for Authorship.

**Conflict of interest**

The following Authors have no financial disclosure.

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**Appendix A. Supplementary data**

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.epsc.2018.06.018>.

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