Elevated bilirubine in 9-months-old infants with chledocal cyst todani

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Abstract
Choledocus cyst is a rare medical condition. This disorder results from congenital cystic dilatation of the biliary duct, and often occurs in the main portion of the common bile duct. Choledocal cysts are extremely rare, with an incidence of about 1 in 100-150,000 in the western population. This disorder often occurs in infants, and children, but some case reports report this disorder can occur from birth to 80 years of age. However, in about 60% of cases, patients are diagnosed at the age of less than 10 years. And this disorder often affects women to men with a ratio of women to men 3-4:1, and often occurs in Asian races.
Keywords: Choledocal cyst Todani, CMV

Introduction

Choledocal cysts Todani is a congenital cystic dilatation of the bile ducts, often occurring in the common bile duct. This condition is relatively rare with an incidence of about 1 in 100-150,000 in western countries. Choledocus duct cysts are classified based on Todani's modification, there are Todani ductus choledocus cysts from type I to type V. Choledocus duct cysts are often a nuisance in infants and children, but can affect newborns up to 80 years of age. But about 60% of cases, this disorder affects children under the age of 10 years. And this disorder often occurs in women compared to men with a ratio of 3-4:1, and often affects Asian races.[1-2] In 1936 Yotsuyanagi et. al. suggested that choledocal cysts arise as a result of imbalanced vacuolization of the biliary ducts in the early embryonic period of life. While Babitt et al. in 1969 suggested that choledocal cysts arise from abnormalities of the pancreatobilliary junction.[3]

There are no specific laboratory tests for diagnosing choledocus duct cysts. However, tests such as liver function can be performed to differentiate from other disorders, and an increase in leukocyte levels accompanied by neutrophilia and immature granulocytes can indicate a cholangitis disorder. Only a proportion of patients present with the classic triad of symptoms, namely abdominal pain, jaundice, and an abdominal mass. The two most common complications that occur due to ductus choledocus cysts are the formation of stones and malignancy.[3]

In addition to laboratory tests, an abdominal ultrasound examination can be performed to help establish the diagnosis. Abdominal CT scan and MRI can also help to determine the anatomy of the lesion and surrounding structures. The accuracy of magnetic resonance cholangiopancreatography (MRCP) in diagnosing ductus choledocus cysts is 100%.[3]

Jaundice which is also known as hyperbilirubinemia is a yellow discoloration of body tissues resulting from the accumulation of excess bilirubin. Obstructive jaundice results from narrowing or blockage of the bile ducts due to stones, masses, inflammation, trauma, and anatomical abnormalities, parasites, and scarring of the liver. Symptoms of obstructive jaundice are putty-colored stools, dark thea-colored urine, yellowness of the eyes or body), itching, pain in the right upper abdominal side, nausea, vomiting, weight loss, and fever.[4]

Meanwhile, pathological jaundice occurs in the first 24 hours and is characterized by an increase in bilirubin levels of more than 0.2 mg/dl per hour or 5 mg/dl per day, or more than 2 weeks of age. Increased conjugated bilirubin is often a pathological condition, this is caused by a defect in the formation of bile salts or impaired transport. It can caused by viral infection such as Cytomegalovirus infection.

Cytomegalovirus infection is a double-stranded DNA virus, β human herpesvirus. Cytomegalovirus infection is one of the causes of infantile hepatitis and cholestasis. CMV infection can infect neonates from intrauterine transmission, from maternal exposure to the genital tract during delivery, or from drinking breast milk, blood transfusions, organ transplants, and transmission from daycare clinics. Postnatal CMV infection may appear 1 to 3 months after exposure and is usually milder than congenital infection. Manifestations of congenital CMV infection include jaundice, hepatosplenomegaly, microcephaly, lethargy, refusal to suckle, seizures, petechiae, purpura. And laboratory findings, namely the presence of anemia, thrombocytopenia, increased liver enzymes, hyperbilirubinemia, increased CSF fluid protein. And radiological findings in the form of pneumonia, neuroimaging such as calcifications (periventricular, thalamic, cortical), ventriculomegaly, congenital dysplasia.[6]

The diagnosis of CMV infection can be made by isolating the virus from urine, saliva, blood and biopsy samples. Specimens can be performed for viral culture, histopathological examination, tissue

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immunofluorescence, electron microscopy, and CMV DNA detection by PCR examination. And you can also go through ELISA or RIA antibody capture as a routine check.[7]

The purpose of writing this case report is to help establish the diagnosis of Choeldical cyst Todani and CMV infection if you find a patient with a similar case.

Case

A 9 month old baby girl with complaints of enlargement of the abdomen. And obtained a brown defecation. The patient also complained of a fever that had been going up and down since two days before he was admitted to the hospital, with a temperature of 38ºC when he had a fever. The fever improved with the administration of paracetamol, and now the patient's temperature is normal at 36.5ºC. The patient has been admitted to the Saiful Anwar Hospital since 2 months ago. The patient has complaints of jaundice in the body since the patient was 21 days old, and there is enlargement of the abdomen since the patient was 4 months old, and was treated at the Saiful Anwar Hospital since January 2022. The patient regularly took Valgancyclovir for 17 weeks. From physical examination, the patient appeared to be moderately ill, microcephaly, and malnourished, tachycardia, tachypnea, anemic conjunctiva, icteric sclera, pulmonary rhonchi, and minimal subcostal retraction, xylophone ribs, abdominal examination revealed distended abdomen, ascites, protruding collateral veins and umbilicus, thin subcutaneous fat, and baggy pants. Laboratory examination revealed normochromic anisocytosis, leukocytosis, hyperbilirubinemia (increased bilirubin T/D/I, and bilirubin D>I), increased procalcitonin, hyperchloremia, decreased urea, hypoalbuminemia, and immunoserological examination found anti-CMV IgM and reactive IgG.

Examination of the thoracoabdominal x-ray revealed a picture of pneumonia. Ultrasound examination of the abdomen showed the presence of contracted vesical fellea with intrahepatal cholangitis. And MRCP examination revealed multiple focal dilatation in CHD and IHBD, suspected choledocal cyst todani type IVA, contracted gall bladder, splenomegaly, and ascites.

Method

This patient obtained by direct observation and direct examination in baby ward Saiful Anwar Hospital, Malang.

Discussion

The presence complaints of jaundice and hypoalbuminemia in patients is caused by hyperbilirubinemia due to Choledocal cyst Todani type IVA but it can also be due to Cytomegalovirus infection. Cytomegalovirus infection in patients was characterized by the presence of positive anti-CMV IgM and IgG. The infection is thought to be caused by congenital infection or postnatal infection. And the cytomegalovirus infection resulted in cholestasis, and anemia in the patient, also characterized by the presence of hyperbilirubinemia in the patient. Pneumonia in patients is suspected to be caused by a viral infection, namely CMV infection or it can also be caused by a bacterial infection. In the patient, a chest X-ray showed an infection with pneumonia, and laboratory findings in the form of leukocytosis, CMV infection with anti-CMV IgM and positive CMV IgG, and increased procalcitonin.

CMV examination in this patient to confirm the presence of cytomegalovirus infection required CMV PCR examination of urine samples.

Conclusion

History, physical examination and investigations confirmed the diagnosis of Choledocal cyst Todani and CMV infection in this patient.

Attachment

Table 1. Hematology laboratory result

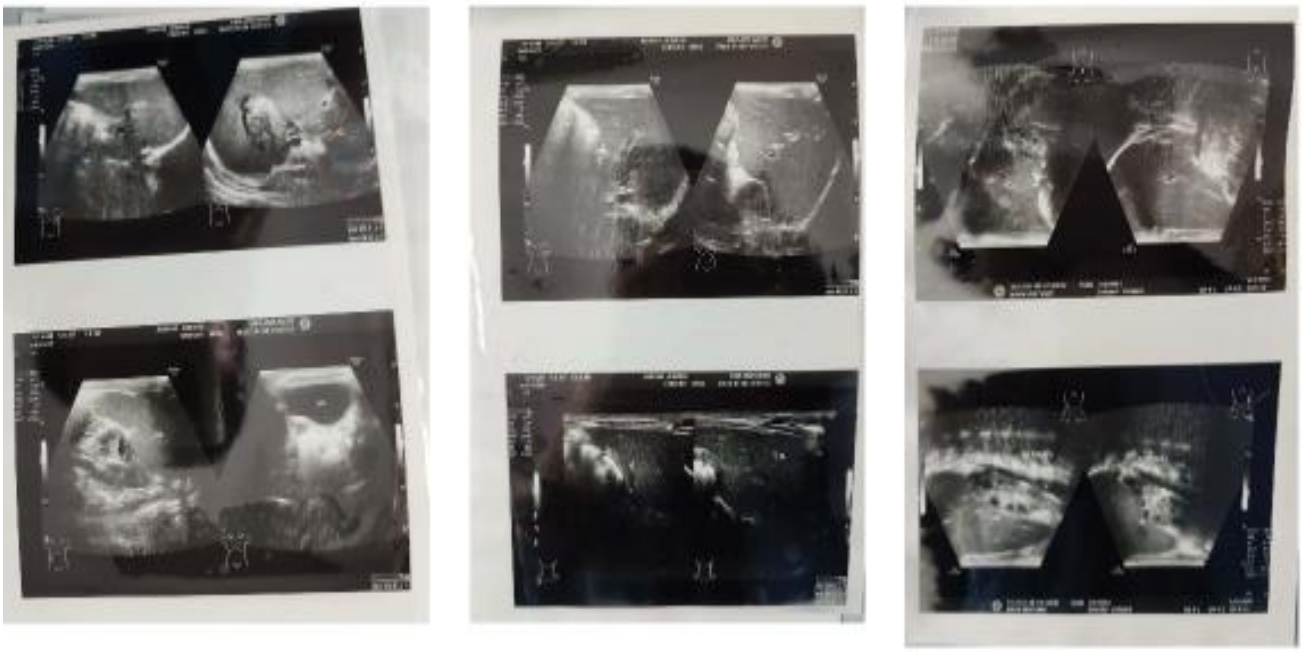
	8/06'22	11/06'22	15/06'22	16/06'22	Nilai normal
Hb	7,00	6,70	8,20	11,3	10,2 – 12,70 g/dL
Leukosit	20,37	22,48	15,82	19,52	6,48 – 13,04 $10^3/mm^3$
Hematokrit	23,30	22,4	26,2	35,7	30,60 – 38,1%
Trombosit	239	245	179	184	214 - 459
MCV	89,60	88,5	86,0	85,2	71,30 – 82,60
MCH	26,90	26,5	27,2	27,0	23,20 – 27,50
MCHC	30,00	29,9	31,3	31,70	28,00-32,00
RDW	19,80	19,00	17,50	17,80	12,7-15,10
Diff. count	1/0/0/58/32/10	1/0/0/69/23/9	1/0/57/33/9	1/0/57/33/9	1-3/0-1/3-6/50-70/20-40/2-8

Table 2. Chemistry laboratory result

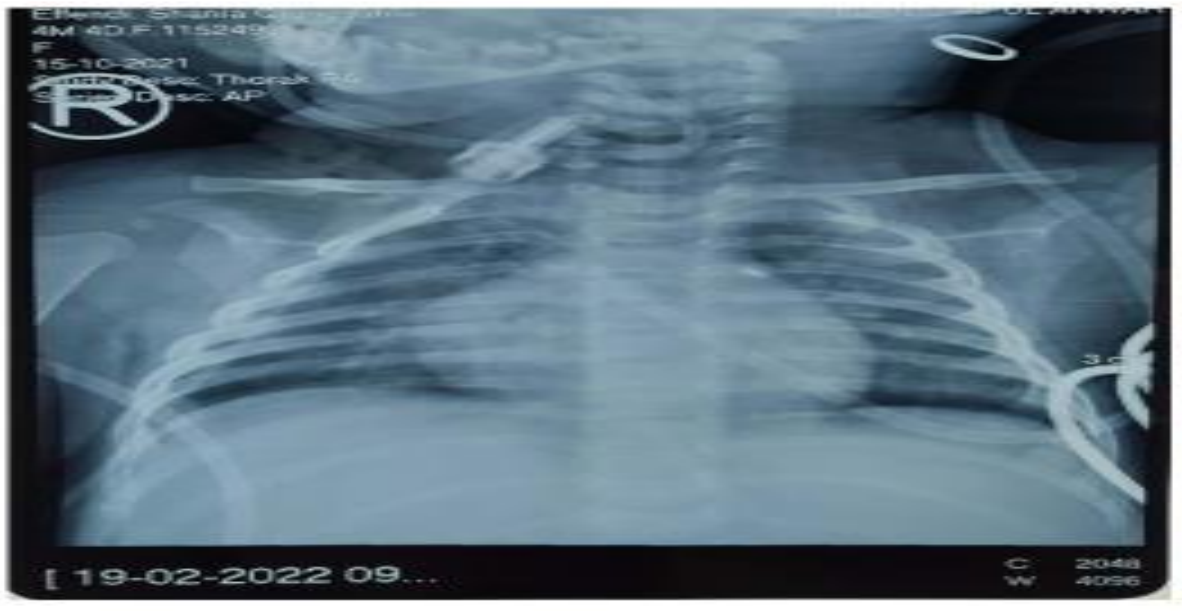
	8/06/2022	11/6/2022	16/06/2022	Nilai normal
AST	129	105	-	<89 U/L
ALT	61	52	-	<45 U/L
Albumin	-	2,56	3,41	3,8-5,4 g/dL
Total bilirubin	9,49	9,00	-	< 1,0 mg/dL
Bilirubin direct	8,38	7,97	-	< 0,25 mg/dL
Bilirubin indirect	1,11	1,03	-	< 0,75 mg/dL
Ureum	-	10,3	-	16,6 - 48,5 mg/dL
Creatinine	-	0,16	-	<0,39 mg/dL
Egfr (Schwartz)	-	160	-	mL/min/1.73 m^2
RBS	-	94	-	< 200 mg/dL

Table 3. Electrolite serum				
	8/06/2022	11/6/2022	16/06/2022	Nilai normal
Natrium	-	137	138	131– 140 mmol/L
Kalium	-	4,29	3,44	3,5 - 6,1 mmol/L
Chloride	-	107	102	97 – 106 mmol/L
Calcium	-	8,3	8,9	9,00-11,0 mg/dL
Phosphor	-	3,7	3,3	3,5 - 6,6 mg/dL

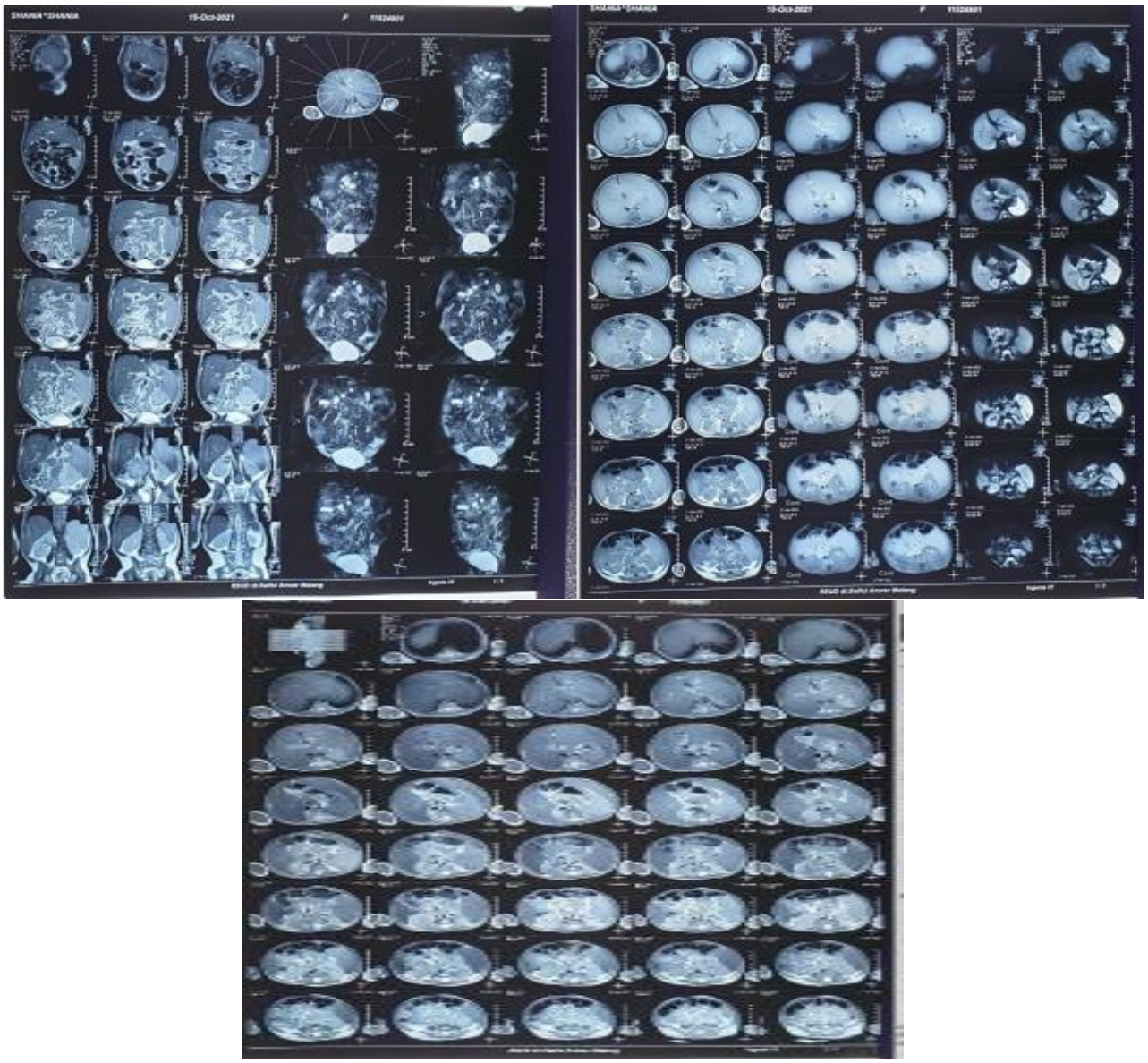
Table 4. Immunology lab result				
	8/06/2022	11/6/2022	16/06/2022	Nilai normal
Anti CMV IgM	PositiveIndex: 1,78	-	-	COI negative < 0,7COI Indeterminate ≥ 0,7 - < 1,0COI Positive≥ 1,0
Anti CMV IgG	215,7	-	-	Negative < 0,5 U/mLIndeterminate 0,5 - < 1,0 U/mLPositive ≥ 1,0 U/mL
Procalcitonin	0,51	-	-	< 0,5 ng/mL low risk for severe sepsis or septic shock>2 ng/mL low risk for severe sepsis or septic shock



Picture 1. USG abdomen result (21/12'22), conclusion contracted vesical fellea with intrahepatal cholangitis



Picture 2. Thoracoabdominal radiographic result (19/02'22), the conclusion is pneumonia



Picture 3. MRCP & MRI lumbal (22/04'22). Kesimpulan focal multiple dilatation on CHD & left IHBD suspect choledocal cysts todani classification type IVA, contracted gall bladder, splenomegaly, and ascites.

Picture 4. Clinical picture of patient

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