

Answers to Medical Quiz

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- A1.** Ultrasound of biliary system. Contracted gall bladder with single stone causing acoustic shadow.
- A2.** Gall bladder stone (cholelithiasis).
- A3.** Acute cholecystitis, acute pancreatitis and gall stone ileus.

DISCUSSION

Sickle cell disease (SCD) is an inherited disorder caused by the mutant sickle hemoglobin, HbS. At the sixth position of the hemoglobin beta-chain, valine is substituted for glutamine acid. This disorder leads to polymerization of the hemoglobin when oxygen saturation is lowered, resulting in chronic hemolysis, vaso-occlusion, ischemia and infarction¹.

The gallstones in SCD patients are pigmented because of chronic hemolysis of the sickle cells resulting in gastrointestinal and biliary complications include cholelithiasis, biliary sludge, colitis, and pancreatitis^{1,2}.

Studies reveal that about 70% of SCD patients will develop gallstones during their lives, though the incidence of gallstones is much lower in patients with HbSC and HbS-beta thalassaemia. The discovery of gallstones in children with SCD increased due to both regular use of the ultrasound and the long survival of these patients^{3,4}.

The development of pigmented gallstones in patients with SCD is age dependent: 15% under 10 years, 22% between 10 and 14 years, 36% between 15 and 18 years and 50% by the age of 22 years⁴.

Most cases of gallstones in pediatric SCD are asymptomatic. Symptomatic cases of gallstones commonly present with right upper quadrant or epigastric abdominal pain, fever, nausea and vomiting. These symptoms are common with wide differential diagnosis, including hepatic, intestinal and pulmonary crises⁵. Perez et al found that children with SCD develop clinical manifestation in 28% of cases with maximum delay of 2.5 years after the diagnosis⁶.

The mainstay diagnostic tool for gallstones in children with SCD is ultrasound⁷. Laparoscopic cholecystectomy is the treatment of choice in symptomatic and asymptomatic cholelithiasis to prevent major potential complications^{8,9}.

CONCLUSION

Gallstones are frequent complication in children with sickle cell disease. Children with chronic hemolysis in asymptomatic sickle cell disease are exposed to a higher risk of developing symptomatic gallstones. Elective laparoscopic cholecystectomy is the gold standard in children with sickle cell disease and cholelithiasis.

Potential Conflicts of Interest: No

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