



PAEDIATRIC DERMATOLOGY

CUTANEOUS XANTHOMATOSIS RELATED TO PRIMARY SCLEROSING CHOLANGITIS: A RARE CASE REPORT

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Background: Primary sclerosing cholangitis (PSC) is a chronic progressive cholestatic disease, characterized by patchy inflammation of the biliary tree, resulting in obliterative fibrosis. Secondary hyperlipidemia is common in cholestatic liver disease, however marked elevation of serum cholesterol with widespread cutaneous lipid deposition is rare. In this report, we present a case of cutaneous xanthomatosis secondary to PSC.

Observation: A 14-year-old boy presented with a 2-month history of pruritic yellow papules on the hips and flexor surfaces of the arms. Coincident with the enlargement of the papules, he had noted yellow streaks on the palmar and plantar creases. He was diagnosed with PSC 3 years ago and asthma since his childhood. The diagnosis of PSC was made by liver biopsy and endoscopic retrograde cholangiopancreatography. Over the past 3 years the patient's condition and liver biochemistry remained stable. Medication at the time of presentation included ursodeoxycholic acid 600 mg daily and salbutamol twice daily. There were no family history of hyperlipidemia or premature atherosclerosis. Examination revealed xanthoma striatum palmare, intertriginous xanthomas were noted on the creases of the wrists and antecubital fossae bilaterally. Lipid investigations revealed: fasting total cholesterol 580mg/dl, low-density lipoprotein cholesterol (LDL-c) 518mg/dl, high-density lipoprotein cholesterol (HDL-c) 39mg/dl, triglycerides 115mg/dl. The patient was referred to the endocrinologist who suggested the introduction of cholestyramine.

Key message: New-onset of xanthomas in a child should prompt investigation for primary or secondary hypercholesterolemia, which can signal more severe hyperlipidemias. In this case, the xanthomas were secondary to high LDL-c from cholestasis due to primary sclerosing cholangitis. Xanthomas secondary to hypercholesterolemia is common in adults, however, primary sclerosing cholangitis and cutaneous xanthomatosis in a child is a very rare condition.

