
CASE REPORTS

Fatal Congenital Systemic Juvenile Xanthogranuloma with Liver Failure

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ABSTRACT

This is the second reported patient with systemic juvenile xanthogranuloma (JXG) to die with liver failure. The infant was born with multiple skin lesions and mild hepatomegaly. Direct hyperbilirubinemia was noted on the 2nd day of life, followed by progressive hepatomegaly, cholestasis, and death at 29 days of age. At autopsy, nodular tumor infiltrates of JXG were present throughout the liver, as well as in skin, abdominal lymph nodes, spleen, and pancreas.

Key words: juvenile xanthogranuloma, neonatal liver failure, histiocytosis

INTRODUCTION

Juvenile xanthogranuloma (JXG) is generally thought to be a benign cutaneous tumor of histiocytic cells, occurring in early childhood and regressing spontaneously. It is the most common “xanthomatous” lesion occurring in infancy. Though first described in 1905 as “congenital xanthoma multiplex,” it was in 1954 that Helwig and Hackney first proposed the term used today: “juvenile xanthogranuloma” when they found that there

were lipid-laden histiocytes and giant cells in the lesion [1]. Clinically, JXG most commonly presents as asymptomatic, solitary, reddish to yellowish, firm papules on the skin of the scalp, face, trunk, or extremities. However, in 5–10% of cases, there is systemic involvement of central nervous system (CNS), liver, spleen, lung, eye, oropharynx, or skeletal muscle [2–5].

Fatal cases of systemic JXG have been reported only rarely [5–7]. Systemic JXG leading to liver failure and a fatal outcome in a neonate is even more unusual.

CASE REPORT

Clinical course

A white female neonate presented with multiple brownish to reddish papules on the skin. She was born at 38 wk of gestation following an uncomplicated pregnancy. Family history was unremarkable. On physical exam, she was found to have intrauterine growth retardation (birth weight was 2462 g, 10–15% percentile) and hepatomegaly to 2 cm below the right costal margin. Skin examination revealed approximately 15 scattered brownish

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