

concurrent symptoms although primary biliary cirrhosis is often diagnosed after an asymptomatic period (Tian et al, 2009). This would suggest that the link between the two conditions might be associated with the natural history of disease progression in primary biliary cirrhosis as well as the underlying autoimmune predisposition. The significance of the latter is highlighted by cases of autoimmune haemolytic anaemia in patients with primary biliary cirrhosis a number of years after liver transplant (Retana et al, 2007). Some cases have been successfully treated with ursodeoxycholic acid alone (Fuller et al, 2003; Gentile et al, 2009). The current case is an example of where ursodeoxycholic acid alone has failed and high-dose immunosuppression was required.

This case adds to the small number of reported cases. It is important to evaluate the rising bilirubin level in conjunction with other biochemical markers. Anaemia

in patients with chronic liver disease often leads to endoscopy as first-line investigation to rule gastrointestinal bleeding. However, a significant rise in bilirubin levels out of proportion to the rise in alkaline phosphatase levels, anaemia with no active bleeding and a macrocytosis with a high reticulocyte count and lactate dehydrogenase can help expedite the diagnosis in these patients. **BJHM**

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Fuller SJ, Kumar P, Weltman M, Wiley JS (2003) Autoimmune hemolysis associated with primary biliary cirrhosis responding to ursodeoxycholic acid as sole treatment. *Am J Hematol* **72**: 31–3 (doi: 10.1002/ajh.10252)

Gentile M, Verta M, Vigna E et al (2009) Autoimmune hemolytic anaemia concomitant with sequential autoimmune hepatitis-primary biliary cirrhosis overlap syndrome and hashimoto's thyroiditis: a new entity of autoimmune polyendocrine syndrome. *J Endocrinol Invest* **32**: 287–8 (doi: 10.1007/BF03346469)

Medeiros CRD, Setubal DC (2004) Autoimmune hemolytic anemia as a complication of primary biliary cirrhosis. *Hematology and Bone Marrow*

## LEARNING POINTS

- Autoimmune liver disease is associated with other autoimmune phenomena, including haemolysis.
- Significant anaemia and rising bilirubin levels out of proportion to a rise in alkaline phosphatase levels should prompt investigation for haemolysis to expedite diagnosis.

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Retana A, Kaplan M, Erban J (2007) Autoimmune hemolytic anemia in patients with liver transplants for primary biliary cirrhosis: Three case reports and review of the literature. *Am J Gastroenterol* **102**: 197–200 (doi: 10.1111/j.1572-0241.2006.00810.x)

Tian Y, Wang C, Liu J-X, Wang H-H (2009) Primary biliary cirrhosis-related autoimmune hemolytic anemia: three case reports and review of the literature. *Case Rep Gastroenterol* **3**: 240–7 (doi: 10.1159/000229189)

## Images in Medicine

# Skin lesions in calciphylaxis

**A** 68-year-old woman with end-stage renal failure on haemodialysis underwent revascularization of the foot. A few days after admission, she developed painful necrotic skin lesions on the breast (*Figure 1*), abdomen and arms (*Figure 2*). The patient had been treated with high dose vitamin D for hypocalcaemia on admission, resulting in a rapid shift in serum calcium. The diagnosis of calciphylaxis was established clinically and incisional skin biopsy was non-specific.

The painful skin lesions developed suddenly and progressed rapidly, starting as purple discoloration (retiform purpura). Lesions generally develop on the lower extremities in areas of adiposity, but may occur in any area (Nigwekar et al, 2015). Ulcerated lesions typically form black eschar and secondary infection may occur; ulceration increases the associated mortality to 80% (Weenig et al, 2007).

**Figure 1. Right breast.**



This patient was treated with analgesia, wound care, sodium thiosulphate and daily dialysis. She unfortunately deteriorated and died 6 weeks after the presentation of the skin lesions. **BJHM**

Nigwekar SU, Kroshinsky D, Nazarian RM et al (2015) Calciphylaxis: risk factors, diagnosis, and treatment. *Am J Kidney Dis* **66**(1): 133–46 (doi: 10.1053/j.ajkd.2015.01.034)

Weenig RH, Sewell LD, Davis MD, McCarthy JT, Pittelkow MR (2007) Calciphylaxis: natural history, risk factor analysis, and outcome. *J Am Acad Dermatol* **56**(4): 569–79 (doi: 10.1016/j.jaad.2006.08.065)

**Figure 2. Right arm.**



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