Biliary peritonitis, due to rupture of intrahepatic duct

Sir,

We would like to report our experience of a patient with biliary peritonitis due to rupture of intrahepatic biliary ducts.

A 65 year old male patient presented with the complaints of pain and distension of abdomen for 3 days. Following examination, a diagnosis of peritonitis with shock was made. Resuscitative measures were started and with improvement in his general condition, an exploratory laparotomy was performed. Approximately 2.5 litres of bile stained fluid were sucked out from the peritoneal cavity. A growth occupying the proximal 1/3rd of the stomach, causing complete occlusion of the stomach lumen, was found. During the search for possible metastasis, two ulcers were found on the superior surface of the left lobe of the liver. Bile was seen oozing from the floor of the ulcers. The extrahepatic biliary ducts, gall bladder and duodenum were normal. A biopsy was taken from the ulcer margins, but revealed no evidence of malignancy. The patient expired on the second post-operative day.

Nauwerk¹ in 1905 first recorded 2 fatal cases of bile peritonitis, each of which at autopsy, proved to have a minute perforation on the surface of the liver. Since then 17 more cases of intrahepatic biliary duct perforation have been added.^{2,3}

Most intrahepatic duct perforations have been seen in elderly female patients. Pre-operative diagnosis of spontaneous perforation of biliary ducts is exceedingly difficult.³ Biliary peritonitis has a very high mortality rate (30–50%).⁴ The present case differs from the earlier cases reported in that spontaneous perforation of intrahepatic ducts occurred in the absence of any apparent common bile duct obstruction, and there were two perforations (ulcers) from which the bile leak was taking place. The cause of the perforation was probably secondary metastasis from carcinoma of the stomach infiltrating the intrahepatic ducts.

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Hyperimmunoglobulinaemia D and periodic fever mimicking familial mediterranean fever in the Mediterranean

Sir,

We have seen two Turkish patients in the paediatric age group with hyperimmunoglobulinaemia D and periodic fever syndrome resembling familial mediterranean fever (FMF). This syndrome was first reported by Van der Meer¹ and, subsequently, by Reeves² in 1984. In 1988 Ostuni reported another case.³

Our first patient was a 10 year old girl who initially had recurrent fevers preceded by chills, transient erythematous rashes, arthralgia, diarrhoea, which sometimes coincided with the fever attacks, since 5 years of age. On physical examination she had multiple cervical micro-lymphadenopathy. Her blood serology was as follows: C₃, C₄ levels were normal, rheumatoid factor, ANA, anti-DNA, LE cell were negative. A skin biopsy revealed capillaritis. Immunoelectrophoresis showed high IgD level (220 IU/l) (normal adult < 150 IU/l). She was put on colchicine, with a diagnosis of hyperimmunoglobulinaemia D and periodic fever syndrome, with little benefit.

Our second patient was a 9 year old girl. She presented with recurrent fevers and abdominal pain, transient erythematous rashes, arthralgia and diarrhoea attacks since age 5. She had postauricular lymphadenopathy. Her C₃ and C₄ levels were slightly elevated, CH₅₀, cryoglobulins, ANA, and fibrinogen were normal. Bone marrow aspiration showed erythroid hyperplasia. Skin biopsy was consistent with nonspecific dermatitis. Her IgD level was elevated at 22 mg/dl (normal adult < 14 mg/dl). A diagnosis of hyperimmunoglobulinaemia D and periodic fever syndrome was made and a course of colchicine was tried, without much improvement.

IgD is a membrane-bound immunoglobulin as an antigen receptor regulation of B cell development. IgD neither crosses the placenta nor is synthesized in significant quantities by the newborn. It reaches adult levels by age 15.^{2,3} Elevated IgD levels in disease are only rarely observed.⁴ Increased IgD levels have been reported in Hodgkin's disease, ataxia telangiectasia, infection with human immune deficiency viruses and recurrent bacterial infections.^{2,5–7} Both our patients were under 15 years of age. They had elevated levels of IgD with the symptoms mimicking FMF but differing from it in several respects, such as predominance of fever with chills rather than abdominal pain. Also lymphadenopathy and diarrhoea occasionally coincided with attacks as reported before.^{1,2}

All reported hyperimmunoglobulinaemia D and periodic fever syndrome patients are of non-Mediterranean origin.^{1,2} Our Turkish cases show that this disease should be kept within the differential diagnosis of FMF even in parts of the world where FMF is more common. Also, the benefit of colchicine regimen in these patients is unclarified.^{1,2}

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