Laparoscopic Cholecystectomy in a Patient with Erythropoietic Protoporphyria

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ABSTRACT

Erythropoietic protoporphyria (EPP) is an inherited defect in haem synthesis causing dangerous phototoxic reactions following exposure to wavelengths of light around 400nm. It can cause catastrophic post-operative complications following open surgery, in which environment various safety measures are now routinely employed. The dangers at laparoscopy have never been discussed in the literature, and nor have any specific precautions been recommended.

We describe a 35 year old woman with gallstones undergoing prophylactic laparoscopic cholecystectomy to prevent future cholestasis precipitating porphyric liver failure. A pre-operative trial of the cutaneous effects of the laparoscopic light source was performed to assess the potential risk of use within the peritoneal cavity. The procedure was uneventful and the patient suffered no adverse reaction.

We suggest that a trial of the effects of the laparoscopic light source on the skin of EPP patients provides valid reassurance regarding the safety of the laparoscopy for short surgical procedures.

INTRODUCTION

Erythropoietic protoporphyria (EPP) is an autosomal dominant defect in haem synthesis caused by defective ferrochelatase; the terminal enzyme. Excessive plasma protoporphyrin is deposited in the skin causing photosensitivity triggered by light wavelengths around 400nm (1). This corresponds to the absorption spectra of porphyrins, which are stimulated to produce free radicals and oxygen atoms (2).

EPP is usually detected before the age of 2, often following a history of screaming and pain when taken outside. Phototoxic burning may occur, although extreme blistering and oedema is more a feature of G?nther's disease - the rarer autosomal recessive form (3). Hepatic protoporphyrin accumulation may lead to liver failure (4).

Although the dangers associated with exposure to theatre lights at laparotomy have been reported (5,6), the possibility of phototoxicity during laparoscopy has never been explored.

CASE REPORT

A 35 year old woman presented for consideration of elective laparoscopic cholecystectomy. EPP was diagnosed after post-natal testing prompted by paternal carriage. Although her brother was severely affected, having undergone liver transplantation following hepatic failure, her own symptoms were purely cutaneous, marked photosensitivity causing pain and blistering.

Examination revealed mild scarring of the face and dorsal aspects of both hands, characteristic of EPP(3), but no other abnormal signs.

Abdominal ultrasonography (a routine prognostic tool in EPP) demonstrated a distended gallbladder containing multiple stones. The liver measured 14cm and exhibited normal echotexture. The CBD and biliary tree were of normal dimensions. Blood tests showed an isolated rise in ALT to 59u/L (0-50), and free protoporphyrin levels of 1.53?mol/L (