predominantly by simple mastectomy or, occasionally, by wide excision biopsy encompassing the nipple and areola. This treatment, particularly in a female, is unnecessarily mutilating. With a limited wedge biopsy and subsequent radiotherapy our patient had a good clinical response, and an excellent cosmetic result.

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References

Congdon G H & Dockerty M B
(1956) *Surgery, Gynecology and Obstetrics* **103,** 185–192
Davis A B & Patchefsky A S
(1977) *Cancer* **40,** 1780–1781
Farrow J H
(1958) *Cancer* **8,** 16–19
Lupton G P & Goette K
(1978) *Archives of Dermatology* **114,** 1845
Robinson H B
(1893) *Transactions of the Pathological Society of London* **44,** 147–148
Wainwright J M
(1927) *Archives of Surgery* **14,** 836–859
Wyatt A P
(1965) *Proceedings of the Royal Society of Medicine* **58,** 509–510

Peutz-Jeghers syndrome causing obstructive jaundice due to polyp in common bile duct[1]

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A case is reported of a patient with Peutz-Jeghers syndrome with duodenal polyposis who developed obstructive jaundice due to a polyp in the common bile duct. Histology confirmed it to be typical of Peutz-Jeghers syndrome. The patient was treated by biliary diversion using a hepatodochojejunostomy. This appears to be the first documented case of obstructive jaundice due to a Peutz-Jeghers polyp of the common bile duct.

Case report

A 44-year-old Pole presented with a six-week history of epigastric pain and jaundice. Both he and two members of his family are known to have Peutz-Jeghers syndrome. In 1971 he had undergone surgery for partial intestinal obstruction at which two separate segments of jejunum containing multiple polyps were excised. He remained asymptomatic for eleven years when he again developed upper abdominal pain. A barium meal showed multiple polyps in the stomach, duodenum and small intestine. Oesophagogastroduodenoscopy confirmed the polyps and biopsy revealed histological proof of Peutz-Jehghers syndrome. At laparotomy a large mass of polyps was found in the duodenum and two further large polyps in the jejunum. The stomach was distended. The bile duct and pancreas were normal. A further segment of jejunum containing the two polyps was excised and a gastroenterostomy performed. The patient made a good recovery but three months later developed recurrent epigastric pain and obstructive jaundice. He was referred to the pancreato-biliary unit at St George's Hospital for further investigation and treatment.

Clinical examination at this time confirmed the jaundice and there were clear manifestations of Peutz-Jeghers syndrome, with pigmented lesions around the mouth and on the hands. There was also a smooth hepatomegaly but no other clinical abnormalities except for a well-healed surgical wound in the abdomen. Biochemical investigations confirmed the obstructive nature of the jaundice with a bilirubin of 270 μmol/l and alkaline phosphatase of 2050 IU/l, with a marginally raised aspartate transaminase of 114 IU/l. Haemoglobin was normal at 12.6 g/100 ml. The erythrocyte sedimentation rate was raised at 66 mm/h. The prothrombin time was slightly prolonged at 17 seconds with a control of 14 seconds. X-rays of the chest and abdomen were unremarkable. Faecal occult bloods were positive. He was treated with daily vitamin K 10 mg intramuscularly.

Oesphagogastroduodenoscopy revealed the oesophagus to be normal, but there was a small pedunculated polyp at the cardia of the stomach. The duodenum contained multiple small sessile polyps which were not causing complete obstruction to the duodenal lumen but were partially obscuring the ampulla of Vater. All the polyps appeared benign macroscopically. The gastroenterostomy stoma was inspected and was healthy but two small polyps were seen in the efferent jejunal loop 2 inches (5 cm) distal to the stoma. A barium meal confirmed the polyposis in the duodenum and unobstructed flow through the gastroenterostomy (Figure 1). Sigmoidoscopy confirmed multiple polypi above 15 cm, and a biopsy of one polyp confirmed it to be a hamartoma typical of Peutz-Jeghers disease. Ultrasound showed the whole biliary tree to be distended with an obstruction at the lower end of

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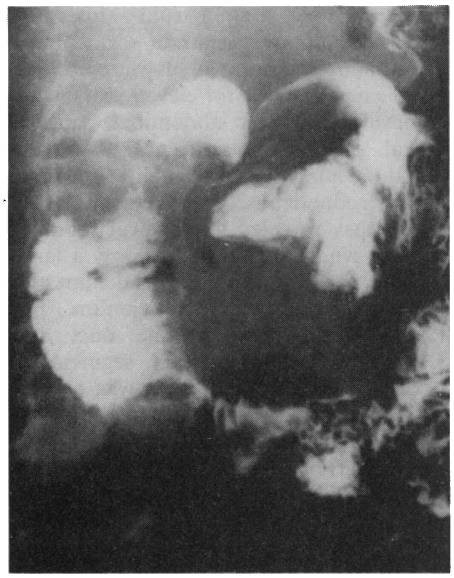

Figure 1. Barium meal demonstrating polyposis in the duodenum and flow-through gastroenterostomy

the common bile duct. This was confirmed by CT scan which was performed to exclude a carcinoma of the head of the pancreas. Multiple biopsies were taken from several polyps, all of which confirmed Peutz-Jeghers syndrome with no evidence of malignancy. Percutaneous transhepatic cholangiography showed an irregular single filling defect at the lower end of the bile duct and confirmed the dilatation of the biliary tree above (Figure 2).

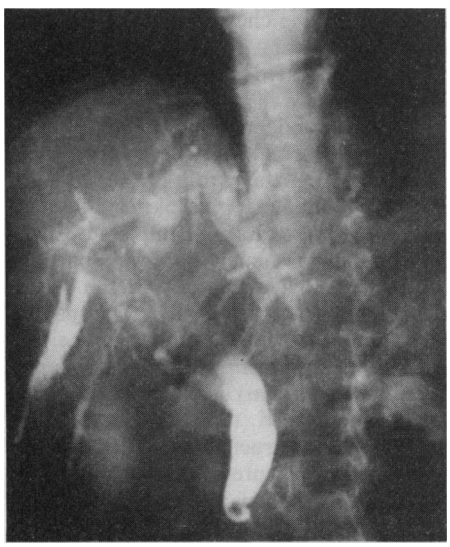

Figure 2. Percutaneous transhepatic cholangiogram showing irregular single filling defect at lower end of dilated bile duct

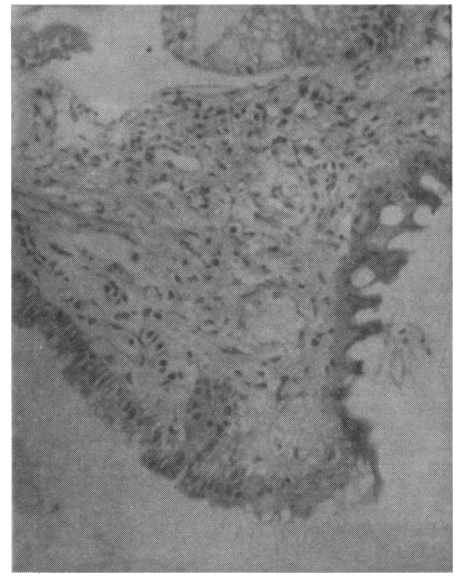

Figure 3. Section of lesion in bile duct demonstrating features of Peutz-Jeghers polyp. (H&E × 230)

At laparatomy the duodenum was found to be full of polyps and several further polyps were palpated in the remainder of the jejunum. The gallbladder and common bile duct were grossly dilated and the liver congested and enlarged. The pancreas was normal. The bile duct was explored and choledochoscopy revealed a lesion at the lower end of the duct which was thickly covered with inspissated bile. Frozen section biopsy showed it to be benign and it was later demonstrated to be typical of a Peutz-Jeghers hamartoma (Figure 3). A sutured hepatodocho-jejunostomy was performed with an antecolic Braun loop of jejunum distal to the previous gastrojejunostomy.

Postoperatively, he made an uneventful recovery and was discharged home two weeks later, by which time the bilirubin had dropped to 70 μmol/l and the alkaline phosphatase to 990 IU/l. He was seen in the outpatient clinic two months later and was fit and well. Liver function tests had returned to normal by this time, and have remained normal at one year.

Discussion

Peutz-Jeghers (P-J) syndrome is an autosomal dominant inherited disease with a high degree of penetration which is characterized by polyposis of the gastrointestinal tract and mucocutaneous pigmentation (Erbe 1976, Bartholomew *et al.* 1957, 1962, Gannon *et al.* 1962, Dormandy 1957). The majority of the polyps occur in the jejunum and ileum but are also seen in the oesophagus, stomach, duodenum and colon (Bartholomew *et*

al. 1957, 1962, Andre *et al.* 1966). The polyps are usually hamartomas, although adenomas are also seen.

One case of gallbladder polyps in P-J syndrome has been described (Foster & Foster 1980) and one case of a papillary (villous) adenoma of the pancreatic duct associated with bile duct carcinoma and tonisillar carcinoma (Bolwell & James 1979).

The relationship of P-J syndrome to gastrointestinal carcinoma remains controversial. In 1957 Bartholomew *et al.* reviewed the literature on the P-J syndrome and concluded that those reports documenting carcinoma arising in P-J syndrome had been misinterpreted. Reid (1974) estimated that 2–3% of patients with the P-J syndrome develop gastrointestinal carcinoma. This figure is probably not significantly different from the normal population. On the other hand, many of those who did develop carcinoma, did so before the age of 40 (Reid 1974, Perzin & Bridge 1982, Bussey 1970). More recently, Linos *et al.* (1981) reported no evidence of decreased survival among 21 patients with P-J syndrome followed for up to 24 years compared to a matched general population. This was contrary to the Japanese survey by Utsunomiya *et al.* (1975). In the most recent review by Perzin & Bridge (1982) their conclusion is that carcinomas may arise within hamartomatous polyps but are possibly preceded by adenomatous change and dysplasia. In our patient it was important to ascertain that there was no evidence of malignancy in the polyps otherwise more radical surgery would have been necessary. In fact, as this was not the case, simple biliary bypass was considered appropriate.

In conclusion, obstructive jaundice occurring in a patient suffering from P-J syndrome may be due to polyposis of the common bile duct and the authors would recommend percutaneous transhepatic cholangiography as a means of preoperative diagnosis. In the absence of malignant change and in the presence of duodenal polyposis, biliary diversion by means of hepatodochojejunostomy should be employed.

References

Andre R, Duhamel G & Bruaire M
(1966) *Bulletin de la Société Médicale des Hopitaux de Paris* **117,** 505
Bartholomew L G, Dahlin D C & Waugh J M
(1957) *Gastroenterology* **32,** 434–451
Bartholomew L G, Moore C E, Dahlin D C & Waugh J M
(1962) *Surgery, Gynecology and Obstetrics* **115,** 1–11
Bolwell J S & James P D
(1979) *Histopathology* **3,** 39–50
Bussey H J R
(1970) *Gut* **11,** 970–978
Dormandy T L
(1957) *New England Journal of Medicine* **256,** 1093–1102
Erbe R W
(1976) *New England Journal of Medicine* **294,** 1101–1104
Foster D R & Foster D B E
(1980) *Postgraduate Medical Journal* **56,** 373–376
Gannon P G, Dahlin D C, Bartholomew L G & Beahrs O H
(1962) *Surgery, Gynecology and Obstetrics* **114,** 666–672
Linos D A, Dozois R R, Dahlin D C & Bartholomew L G
(1981) *Archives of Surgery* **116,** 1182–1184
Perzin K M & Bridge M F
(1982) *Cancer* **49,** 971–983
Reid J D
(1974) *Journal of the American Medical Association* **229,** 833–834
Utsunomiya J, Gocho H, Miyanga T *et al.*
(1975) *Johns Hopkins Medical Journal* **136,** 71–82

Syndrome resembling graft-versus-host disease in a patient with disseminated carcinoma[1]

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A patient with disseminated carcinoma is described who developed cutaneous and systemic features closely resembling those seen in chronic graft-versus-host (GVH) disease. The patient had not received a bone marrow or blood product transplant. The possibility that a GVH-like reaction was induced by alteration of her 'self-antigens', consequent upon her malignancy, is considered.

Case report

Mrs E B developed dry eyes and a dry mouth at the age of 66 years. Reduced lacrimal secretion was confirmed by Schirmer's test. Three weeks later an eruption appeared on her shins composed of erythematous telangiectatic macules. These lesions subsequently became papular and violaceous, resembling lichen planus. There was no improvement in the eruption following the application of topical corticosteroids. Within one month of the onset of her rash she developed ankle oedema, weight loss and diarrhoea. Absorption studies demonstrated that she was malabsorbing and, as a duodenal biopsy showed partial villous atrophy, her malabsorption was attributed to coeliac disease. However, she failed to respond clinically to a gluten-free diet and after 3 months on this diet there was no improvement in her sugar absorption studies.

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