

became yellow a week before this episode. Chronic lymphatic leukaemia had been diagnosed 8 years previously and remained well controlled; because of this he took an antibiotic whenever he developed a cold but did not do so on this occasion. In the past he had had many febrile upper respiratory infections, but had never experienced an aura during these. Previously he had not had multiple attacks, headache or post-aura light sensitivity. In childhood he had had no travel sickness or recurrent abdominal pain. His sister had migraine with aura; no other family member was known to have migraine currently or in the past.

COMMENT

Alvarez¹ described similar personal experiences. His aura began in youth, occurred about once a month for no apparent reason, and was not followed by a headache until age 67: the headache was then mild but he did not state its site or duration. Aged 72, he had two attacks on one day and two more three days later associated with 'a trace of fever' due to a saphenous vein thrombosis. On one occasion after an aura in the morning, he noticed that afternoon at a musical recital that he felt 'distressed by a brilliant after-image of the piano which I got if I looked at it for more than a few seconds.' Perhaps the white piano keys were responsible. Alvarez also recorded one patient whose vision remained hypersensitive for five hours after an aura.

Migraine aura without headache is well recognized. The International Headache Society Classification² specifies the aura duration as usually less than sixty minutes, though sometimes longer. However, scant attention has been given to the mechanism(s) of increased number or changes in pattern of attacks as in this case. The additional headache here could have arisen from meningeal vasodilatation by extension of mucosal vasodilatation in the nasopharynx and paranasal sinuses secondary to the infection.³ Observations during migraine episodes indicate that the headache arises from nociceptive nerve endings of the meninges,⁴ the brain itself being insensitive to pain. More difficult to explain is the increased frequency of the visual aura and its increased duration. If the aura arises in the occipital cortex⁵ perhaps the pyrexia was contributory.

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Combined surgical and endoscopic clearance of small-bowel polyps in Peutz–Jeghers syndrome

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In Peutz–Jeghers syndrome (PJS) a combined surgical and endoscopic procedure allows effective clearance of the multiple hamartomatous polyps from the entire length of the small bowel.

CASE HISTORIES

Case 1

A woman of 18 was referred with intestinal obstruction. 4 years previously PJS had been diagnosed after an episode of intussusception. She had the characteristic perioral pigmentation. At that time she had undergone near-total clearance of her small-bowel polyposis by combined laparotomy and small-bowel enteroscopy, with subsequent yearly surveillance and polypectomy by small-bowel enteroscopy. At the latest admission a small-bowel meal suggested further extensive polyposis. Repeat combined laparotomy and small-bowel enteroscopy allowed removal of over 40 polyps by snare loop diathermy. The entire length of the small bowel was visualized, as well as the ascending colon to the hepatic flexure. No enterotomies were required to remove the polyps.

Case 2

The second patient, now aged 17, was diagnosed at age 11 years. She had required an emergency ileo-caecal resection after intestinal intussusception. Since the diagnosis of PJS, surveillance endoscopies were conducted and several small gastric polyps were removed over the years. The present episode began with colicky abdominal pain and she proved to have iron-deficiency anaemia. A small-bowel meal revealed several small-intestinal polyps. At laparotomy with

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on-table enteroscopy, multiple small polyps were cauterized and several moderate-sized polyps were removed by snare polypectomy. Four larger polyps were removed via enterotomies. She made an uneventful recovery with return of bowel function in 4–5 days. On histological examination there was no evidence of dysplasia or malignancy.

COMMENT

PJS is an autosomal dominant condition with incomplete penetrance characterized by hamartomatous polyps of the gastrointestinal tract and melanin pigmentation of the mucocutaneous membranes. Over the past two decades there has been a change in management. Formerly these patients were managed by a combination of gastroduodenoscopy and colonoscopy with polypectomy. Surgery was reserved for dealing with complications such as small-bowel obstruction, intussusception and gastrointestinal haemorrhage. Polyps were identified at laparotomy by a combination of transillumination and palpation; however, Spigelman *et al.*¹ demonstrated that 38% of polyps were not palpable. Thus patients were exposed to repeated laparotomies and small-bowel resections with the risk of developing short-bowel syndrome.

The combined approach of intraoperative endoscopy was described by Mathus-Vliegen and Tytgat² in 1985. This allows direct visualization of the whole length of the small bowel during the laparotomy itself.³ The procedure is performed under general anaesthesia. The endotracheal tube is of small diameter to facilitate passage of the scope⁴ and a nasogastric tube allows suctioning of air and secretions from the stomach (which can blow up with the insufflated gas). A paediatric colonoscope is then passed via the mouth and with the assistance of the surgeon is guided through the stomach and duodenum into the small intestine. Often manual pressure must be applied to the scope to achieve passage through the bends of the duodenum. At this point the operating surgeon has to ensure that the bowel is not torn or the mesentery of the small intestine excessively stretched with resultant bleeding, haematoma and infarction. In most cases, the polyps are then easily identified and can be snared via the enteroscope. This reduces the number of enterotomies required and also decreases the potential complication rate. The surgeon is able to assist the endoscopist in the snaring of the polyps as well as in

guiding the endoscope through the entire bowel by pushing the small bowel over the endoscope. As the endoscope proceeds it becomes less easy for the endoscopist to manoeuvre, but the surgeon can often direct it to the right position. Bulky lesions and thick-stalked polyps require open polypectomy,⁴ and the enterotomy site can then be used as a point of entry for the scope. We often employ a sterile laparoscopic camera sheath to aid with sterility. The endoscope is passed proximally as far as the duodenojejunal flexure and polypectomies are performed on withdrawal of the scope. The scope is then passed distally to the ileocaecal valve and polypectomy is performed, again on scope withdrawal. The snared polyps can then be milked back to the enterotomy site and retrieved for histological examination. We use single layer closure for the small-bowel enterotomies.

This combined procedure is simple, safe and effective and renders the patient symptom-free for long periods. Disadvantages are the prolonged postoperative ileus that can result from handling of the bowel and stretching of the mesentery, and reactionary haemorrhage from polypectomy sites. Small-bowel leakage is another hazard, though less likely than with the multiple enterotomies of a conventionally performed Peutz–Jeghers laparotomy. In view of the risk of further polyp formation and of malignant change, these patients require ‘top and tail’ endoscopy along with imaging of the small bowel every 2 years. Polyps that are causing symptoms, or are greater than 1.5 cm in size, or are suspicious of malignancy should be managed by intraoperative enteroscopic polypectomy if possible, or by open resection if not.⁵

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