of the ileal wall (Figure 1). The suspicious omental areas which were resected showed non-specific inflammation, and the peritoneal fluid revealed pus cells and Escherichia coli on culture; however, no malignant cells were seen.

In view of the metastatic disease and the poor general condition of the patient, no further adjuvant treatment was offered. At an eight-month follow-up, the patient is alive and asymptomatic and has learnt to manage his ileostoma well.

While the small intestine accounts for nearly 75% of the total length of the GI tract and more than 90% of the mucosal surface area, less than 25% of all alimentary tract neoplasms and less than 2% of all malignant tumours originate from the small intestine. This could be due to the relatively rapid transit time, liquid content of stools, low bacterial population, and a high concentration of IgA within the lumen. Metastatic tumours of the small bowel outnumber the primary tumours and usually occur as part of generalised peritoneal carcinomatosis. Rarely are these metastatic tumours solitary. The different mechanisms postulated for isolated small bowel metastasis include retrograde lymphatic spread following blockade of paraaortic or mediastinal lymph nodes, lymphatic embolisation, haematogenous spread or peritoneal seeding, including direct implantation. The ileum is the commonest site of metastatic lesions in the small intestine, probably because of the large number of Peyer’s patches, which make it the best trapping zone for hematogenic metastasis.

Ileal metastasis of squamous carcinoma may arise from the cervix, lung and other sites, but metastasis from oesophageal carcinoma is rare. Although equally uncommon, the possibility of a primary squamous cell carcinoma of the ileum was excluded since these usually arise in the presence of bowel duplication, and more importantly, the patient already had a primary tumour in the oesophagus.

Despite the inevitable concern of finding generalised abdominal carcinomatosis, laparotomy is indicated to relieve intestinal obstruction, either by a palliative intestinal resection or by a bypass surgery. Although the overall prognosis is poor, significant palliation or occasionally, long-term survival may be achieved in the absence of disseminated disease.

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Yet another cause for drug-induced pulmonary fibrosis

Sir,

Hydroxyurea is a drug mainly used for the treatment of chronic myeloid leukaemia and polycythaemia. Its most common toxic effects are myelosuppression, nausea and skin reactions.

An 80-year-old Caucasian woman was referred by her primary care physician to a haematologist with findings of haemoglobin 20.4 Gm% and PCV 0.62 in May 2002. A diagnosis of primary polycythemia was made by the haematologist after a detailed work-up. The patient was started on hydroxyurea 1 g/day for 2 months (approx 20 mg/kg/day). This was subsequently reduced to 1 mg and 0.5 mg on alternate days, when the patient showed an adequate response as seen by a reduction in her haemoglobin and PCV.

Four months after this, she presented with gradual onset of shortness of breath and wheeze. She had received 2 courses of antibiotics from her primary care physician with no resolution of symptoms. Chest X-ray during admission showed marked honey-combing of the lungs at both bases and lung function tests were in keeping with a restrictive pattern of lung disease, with a FEV1 63% predicted and FVC 67% predicted and a FEV1/FVC of 104%. A CT scan of the chest (Figure 1) confirmed the presence of pulmonary fibrosis in both the bases.

This suggested a predominant involvement of sympathetic functions rather than parasympathetic. In another study, abnormal and spontaneous changes in circadian rhythm of temperature, pulse and blood pressure were observed, with febrile peaks in the absence of an infective focus, suggestive of an autonomic dysfunction. Meenakshi-Sundaram et al reported dysautonomia in 38% (19/50) of patients with WD of varying duration and severity. These studies provide convincing evidence about autonomic dysfunction in WD. However, they are often asymptomatic and diagnosed with the help of clinical and electrophysiological tests.

There are conflicting reports regarding the mechanism of autonomic dysfunction in WD. Some reports favor a central origin. Chu et al reported a significant prolongation of mean latencies and mean central conduction time favoring a central involvement. Meenakshi-Sundaram et al too favored central origin as they found normal peripheral nerve conductions in all. However, von Giesen et al found significantly higher thresholds for warm sensation in sural and peroneal nerves with the help of quantitative sensory testing indicative of damage to unmyelinated warm-specific C fibers, favoring a peripheral origin of dysautonomia.

In conclusion, one it should be noted that autonomic dysfunction occurs in WD and is probably underdiagnosed. Hence, WD should be excluded in a young individual presenting with autonomic dysfunction.

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