Adenocarcinoma of the stomach and Fanconi's anaemia

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Summary

A case of Fanconi's anaemia is described. The patient developed gastric ulcers at 14 years and died of adenocarcinoma of the stomach at the age of 21 years. The need fully to investigate gastric pathology by endoscopy and biopsy, as well as radiology, is stressed. So far as the authors are aware this is the first description of adenocarcinoma of the stomach occurring in Fanconi's anaemia, a condition known to predispose to malignancy.

Introduction

Fanconi's anaemia is an inherited disorder characterized by chromosome abnormalities, aplastic anaemia and multiple congenital abnormalities. Leukaemia and other malignancies have been noted to occur more frequently, but adenocarcinoma of the stomach, so far as can be ascertained, has not been reported before. This case is important in that the mode of presentation with worsening anaemia and gastrointestinal blood loss could have been wrongly ascribed to treatment with corticosteroids.

Case report

A 21-year-old male with known Fanconi's anaemia had only recently been discharged from hospital after a maintenance transfusion of 5 units of blood. He had been treated since early childhood with prednisolone 2.5 mg and oxymethalone 50 mg each twice/day and, 7 years previously, had suffered indigestion and haematemesis which had then been ascribed to steroid therapy. A barium meal showed an ulcer on the lesser curve close to the pylorus with a second ulcer on the greater curve also adjacent to the pylorus. He was treated medically and recovered and thereafter had remained well on a combination of enteric coated prednisolone and oxymethalone.

A few days before his last admission he had complained of lower back pain. Physical examination and spine X-rays showed no abnormality and there was no evidence of gastrointestinal blood loss. Two days later he was admitted shocked and distressed with a brisk haematemesis and melaena. Resuscitation was commenced with whole blood, platelet concentrates and fresh frozen plasma and, when stable, an attempt was made at gastroscopy but the procedure was abandoned because of further bleed-

ing. Later, he bled again and was subjected to laparotomy. At operation there was a large shallow ulcer on the lesser curve of the stomach with much surrounding oedema and induration involving the mesentery. The postoperative course was stormy and, despite multiple transfusions with blood, fresh frozen plasma and platelets, the patient died on the eighth postoperative day.

The gastrectomy specimen revealed the source of bleeding which was an ulcerated carcinoma one cm in diameter at the distal line of resection. Microscopy showed the tumour to be a poorly differentiated adenocarcinoma of the stomach principally characterized by small polyhedral cells arranged in columns and groups, infiltrating the stomach walls and extending to the perigastric connective tissues as well as the distal line of resection. The gastric mucosa away from the tumour appeared normal, there was no evidence of lymphatic or vascular invasion.

Discussion

Leukaemia, hepatoma and oro-genital and oesophageal squamous carcinomas have been found to occur more commonly with Fanconi's anaemia whether or not treated with androgenic anabolic steroids (Bourgeois and Hill, 1977; Swift, Zimmerman and McDonough, 1971; Sarna *et al.*, 1975). Most cases of Fanconi's anaemia are treated with prednisolone and androgenic steroids and these may exacerbate gastrointestinal ulceration and bleeding. Consequently, a patient presenting with gastrointestinal blood loss might be considered as having steroid-related blood loss. The present observation would appear to make a thorough endoscopic and barium study of the upper gastrointestinal tract mandatory in patients presenting with signs or symptoms of acute or chronic blood loss or where there is sudden deterioration in an otherwise stable patient.

References

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