Discussion

That the anuria was apparently painless yet due to bilateral obstruction of the ureters by calculi is noteworthy. Herring,⁴ in a study of 10 000 urinary calculi, found only 89 to contain cystine, of which about 70% were composed of pure cystine, as in our case. Thus urinary tract calculi may be due to cystinuria even in a young child. Cystinuria⁵ is a recessive or incompletely recessive genetic abnormality. Urinary concentrations of cystine, lysine, arginine, ornithine, and cystine-homocysteine are raised. Cystine, the least soluble of the urinary amino-acids, tends to precipitate when present in excessive concentrations. Cystine stones, however, occur only in some cystinuric patients, who may develop them presumably either in utero or during infancy, but the precise reasons for their formation are uncertain. Predisposing causes such as infection or anatomical abnormalities of the urinary tract are unusual.

Most medical treatments aim at reducing the concentration of urinary cystine. Penicillamine or a "high fluid intake" may cause dissolution of cystine calculi, but the immunological abnormalities induced by penicillamine, hypertension, heart failure, electrolyte disturbances due to fluid and alkali excess, and impaired renal function are important considerations in management. Our aim in this patient is to prevent further stone formation and avoid electrolyte imbalance. We are reluctant to use penicillamine because of its side effects.

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Hull Royal Infirmary, North Humberside, Yorks

M J FARR, MRCP, consultant nephrologist
D W W NEWLING, FRCS, consultant urologist
A H ELDRISSEY, MD, DCH, paediatric registrar
G A MIRGHANI, MD, DCH, paediatric senior house officer
R J PUGH, FRCP, consultant paediatrician

IgA deficiency and neutropenia

Immunodeficiencies may be associated with neutropenia.^{1 2} We report here a patient with IgA deficiency and neutropenia whose clinical state and neutropenia responded to corticosteroids. This does not seem to have been reported.

Case report

The patient first presented with thrombocytopenia, lymphadenopathy, and splenomegaly at the age of 10 years. He had had many attacks of "bronchitis." There was no family history of recurrent infections. A full blood count was normal except for thrombocytopenia. An antinuclear factor test was negative. Marrow aspirate showed an active marrow with normal megakaryocytes. After splenectomy the platelet count rose to $390 \times 10^9/l$ ($390\,000/mm^3$) and remained normal. Splenic histology was normal. During the next few years he had recurrent attacks of bronchopneumonia and otitis media.

He was readmitted at the age of 14 with fever and cough. Laboratory findings were as follows: leucocyte count $3.1 \times 10^9/l$ ($3100/mm^3$), neutrophils 8%, lymphocytes 74.5%, monocytes 15%, and eosinophils 1.5%; platelet count $161 \times 10^9/l$ ($161\,000/mm^3$); Paul-Bunnell and extended Widal tests negative; bone marrow aspirate showed active myelopoiesis; IgG 10.8 g/l, IgA 0.3 g/l, IgM 7.5 g/l; 51% of lymphocytes formed spontaneous rosettes with sheep red cells (normal 50%-60%); lymphocyte response to phytohaemagglutinin stimulation normal; surface immunofluorescence staining test showed IgG in 6.4% lymphocytes (control 13.3%), IgM in 5.2% (control 11%), IgA in 2.7% (control 6%), and a slightly reduced B-cell population. Assay of antibody to *Escherichia coli* and antibody response to primary antigenic challenge with pneumococcal polysaccharide were negative. Mantoux test (1/1000) was also negative although the patient had been vaccinated with BCG. Platelet agglutinin and complement fixation test for incomplete platelet antibodies were negative. Leucocyte agglutinin was positive. Tests for other antibodies, including antinuclear antibodies, smooth muscle antibodies, and mitochondrial antibodies, were all negative.

The patient was given cephaloridine 6 g/day for seven days with no effect.

Prednisone 40 mg/day was then started. The fever lysed within 24 hours, and the neutrophil count rose from $0.18 \times 10^9/l$ to $1.71 \times 10^9/l$ (180 to $1710/mm^3$) over 20 days. Over the next five years attempts to withdraw the steroid were unsuccessful. Every time he would become neutropenic and feverish and would respond only to an increase in prednisone dosage. He also developed eczema. He was last admitted at the age of 21 with pneumococcal septicaemia. His leucocyte count then was $6.2 \times 10^9/l$ ($6200/mm^3$), neutrophils 81%, lymphocytes 12%, monocytes 7%; IgG 6 g/l, IgA 0.39 g/l, and IgM 1.98 g/l. Necropsy showed generalised lymphadenopathy with bilateral pleural effusion and no evidence of lymphoma.

Immunoglobulin levels in members of the immediate family were normal.

Discussion

This patient had low IgA, normal IgG, and high IgM serum concentrations. B-cells were slightly reduced, and the number and function of T-cells were probably normal. Recurrent bronchopneumonia, otitis media, thrombocytopenia, neutropenia, eczema, and a raised level of IgM are not unusual in selective IgA deficiency.³ IgA deficiency is associated with autoimmune phenomena and with the presence of various autoantibodies.³ Furthermore, immunodeficiency may be associated with neutropenia,^{1 2} but the cause of this is not clear. The beneficial effect of prednisone on our patient's neutropenia and infection, and their exacerbation when prednisone was withdrawn, pointed to an immunologically mediated destruction of his polymorphs, in which the leucocyte agglutinin might have been implicated. His immune deficiency was unlikely to have been the result of splenectomy in childhood, since after splenectomy children usually have low serum IgM but normal IgA and IgG concentrations.⁴ We therefore think that his IgA deficiency was probably congenital and the neutropenia was due to the presence of a leucocyte antibody which was controlled with prednisone.

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Department of Clinical Haematology, University College Hospital Medical School, London WC 1

R P NG, MB, MRCP, senior lecturer in haematology
T A J PRANKERD, MD, FRCP, professor of haematology

Acute dilatation of stomach as late complication of drug overdose

To our knowledge acute dilatation of the stomach complicating severe drug poisoning has not been reported.

Case report

A 16-year-old girl was admitted to hospital four hours after taking an unknown quantity of amitriptyline, orphenadrine, and flurazepam. Her pupils were widely dilated and she was deeply unconscious showing no response to painful stimuli. The heart rate was 120 per minute and the systolic blood pressure only 80 mm Hg. A cuffed endotracheal tube was passed and a minimal amount of tablet fragments was recovered by gastric lavage. Subsequently convulsive episodes occurred and an electrocardiogram showed an idioventricular rhythm. Supportive measures were instituted and cardiac monitoring showed resumption of sinus rhythm four hours after admission.

Her general condition gradually improved and after 48 hours she was fully conscious and superficially well, the pulse rate being 90 per minute and the blood pressure 120/75 mm Hg. On the third day, however, she

became acutely ill complaining of generalised abdominal discomfort. The pulse rose to 120 per minute and the blood pressure fell to 90/50 mm Hg. The abdomen became considerably distended, with no audible bowel sounds, but vomiting did not occur and her temperature was normal. Supine and erect abdominal x-ray films showed considerable distension of the stomach with gas and fluid, but neither the small nor large bowel showed distension or fluid levels. Gastric aspiration produced 2500 ml of light brown fluid and continuous drainage yielded a further 6000 ml of fluid over the next 24 hours. During this period she received a replacement intravenous infusion of normal saline with potassium supplements and her plasma electrolytes were maintained within the range of normal. Sustained improvement occurred—the pulse, blood pressure, bowel sounds, and urine output gradually returning to normal. After a further few days of convalescence she had recovered fully and was discharged.

Discussion

Acute dilatation of the stomach is a life-threatening condition which has been encountered and studied more intensively in surgical rather than medical practice.¹ Awareness of its gravity and the more routine use of the nasogastric tube have reduced the frequency and mortality of this complication after surgery or trauma. Although it has been described in diabetic coma and as a complication of the treatment of anorexia nervosa,² it remains a rare but important medical emergency since any delay in treatment carries a high mortality.³ Both amitriptyline and orphenadrine may affect the gastrointestinal tract adversely, producing paralytic ileus and faecal impaction,⁴ which have been attributed to their anticholinergic properties. A search of published work, however, has failed to uncover any account of acute gastric dilatation complicating such drug poisoning. Our experience with this patient indicates the need to bear this possibility in mind and also emphasises that, like the cardiac arrhythmias, it may be a late complication.

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Dumfries and Galloway Royal Infirmary, Dumfries DG1 4AP

J HOW, MB, MRCP (UK), registrar in general medicine (now lecturer in haematology, University of Aberdeen)

R W STRACHAN, MB, FRCPE, consultant physician

Retrosternal goitre presenting as bleeding oesophageal varices

Oesophageal varices are normally due to portal hypertension. Occasionally, however, they are seen in association with mediastinal obstruction, usually with malignant superior vena caval obstruction: the so-called "downhill" varices. Bleeding is rare. The following case presented with massive gastrointestinal haemorrhage in the absence of clinical superior vena caval obstruction. Large, bleeding mid-oesophageal varices were demonstrated endoscopically and radiologically, which disappeared after the excision of a large, benign, retrosternal goitre.

Case report

A 64-year-old woman was admitted as an emergency. Three days previously she had suddenly developed severe retrosternal and epigastric pain, followed by a small haematemesis. Subsequently she complained of episodic retrosternal pain and melaena. There was no relevant previous medical history. On examination she was pale, ill, and hypotensive. The left lobe of the thyroid was enlarged and irregular. There was mild epigastric tenderness and melaena on rectal examination. There were no other abnormal physical signs. Haemoglobin was 9 g/dl on admission and rapidly fell further. Endoscopy showed considerable oesophageal displacement to the right, and large bleeding varices were seen at 15-20 cm. Below this, there was no abnormality. A chest radiograph showed displacement of the trachea by a large superior mediastinal mass, and a barium series confirmed the marked oesophageal displacement and showed varices in the midoesophagus in the region of the obstruction. A thyroid scan showed a large retrosternal goitre.

Chest radiograph showing large superior mediastinal tumour.

Portal pressure measured by splenic puncture was normal. At laparotomy, the liver and spleen appeared healthy and the portal vein pressure was normal. A liver biopsy was taken, the abdomen closed, and a sternotomy performed. The left lobe of the thyroid was greatly enlarged and descended behind the trachea to the right side, displacing the oesophagus. The lobe was mobilised and excised. Subsequent progress was uneventful. No further bleeding occurred and a gastroscopy four months later showed no trace of varices. The large, nodular tumour removed was shown histologically to be a benign colloid goitre. The liver biopsy was normal.

Discussion

The submucosal and perioesophageal venous plexus of the mid-oesophagus drains into the azygos system and thence into the superior vena cava (SVC). This anatomical arrangement probably accounts for oesophageal varices with SVC obstruction. Most cases have been due to neoplastic lymph node enlargement. Felton and Lessure reviewed 30 cases in 1964.¹ The cause was bronchogenic carcinoma in 15, Hodgkin's disease in two, with single cases of metastatic carcinoma, surgical caval ligation, and an undetermined mediastinal mass. Seven were thought to have mediastinal fibrosis, and three had retrosternal goitres. The preponderance of malignant tumours was confirmed by Gerstenberg in 1971.² Keminger and Kolb in 1972³ described two cases of cervical oesophageal varices with retrosternal goitres and advised phlebography prior to thyroidectomy.

The present case is unusual in that SVC obstruction was not clinically apparent. The situation of the tumour would, however, readily account for pressure on the superior vena cava, or more locally on the azygos system or oesophageal plexus. Phlebography would have been interesting, especially since the varices appeared to be predominantly lateral to and below the obstruction, so that it is not clear that they pursue a "downhill" course. The other feature of interest is the mode of presentation. Most varices of this sort are asymptomatic, though bleeding has occasionally been reported.^{4,5} The case demonstrates the value of early endoscopy in the management of upper gastrointestinal bleeding. Evidently, the possibility of mediastinal obstruction needs to be at least considered in the differential diagnosis, though in many cases a chest radiograph will suffice to exclude it.

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North Manchester General Hospital, Crumpsall, Manchester M8 6RB

P V BARBER, MB, MRCP, senior registrar in chest diseases

J D EDWARDS, MB, MRCP, registrar in medicine

B A ENOCH, MB, MRCP, consultant physician

M SHAFIQ, MB, FRCS, consultant surgeon

A G VALLON, MB, CHB, senior house officer