Boari flap was undertaken, and histological examination confirmed the presence of a single pTaG1 papillary transitional cell carcinoma in the lower third of the ureter. Subsequent cystoscopies at three and six months were normal. One year after the surgical procedure the patient developed painless haematuria. IVU demonstrated normal upper tracts and bladder, but cystoscopy revealed multiple superficial pTaG1 papillary bladder tumours. These were managed with a combination of endoscopic treatment and intravesical chemotherapy with ethoglucid. The patient remained tumour-free for 2 years when he developed recurrent multiple pTaG1 bladder tumours. These were treated with transurethral resection and intravesical mitomycin, followed by intravesical BCG. The bladder became clear of tumour and remained so for over 3 years. However, despite normal cystoscopy the patient reported haematuria. On IVU the kidneys and bladder were normal but there was a filling defect in the reconstructed lower third of the right ureter (Figure 1). A right nephroureterectomy was performed with excision of a generous margin of the bladder, and histological examination revealed a single pTaG1 tumour in the Boari flap and another pTaG1 tumour in the right renal pelvis.

COMMENT

With a single low-grade tumour in the lower ureter, our patient seemed an ideal candidate for localized ureteric resection and reconstruction by a Boari flap. However, it subsequently became evident that this single tumour was the first manifestation of a multifocal disease process in the urothelium. Over seven years the patient developed multiple superficial tumours in the bladder and kidney as well as in the Boari flap. Considering that a Boari flap is part of the bladder, tumour recurrence in it is not surprising but it does not seem to have been reported previously.

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Autoimmune enteropathy with goblet-cell antibodies

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Autoimmune enteropathy is a rare disease characterized by protracted diarrhoea and severe malabsorption. To make the diagnosis all other causes of chronic diarrhoea have to be excluded. Most patients present in infancy and have serum antibody to epithelium. There is a strong association with other autoimmune conditions and many patients have a family history of autoimmune disease[1].

CASE HISTORY

A 19-year-old woman with multiple autoimmune disorders (Table 1) had an 11-year history of protracted diarrhoea and severe malabsorption. Initial investigations revealed severe fat malabsorption but pancreatic function tests were normal. A sweat test as well as DNA tests for the most common mutations of the cystic fibrosis gene were negative. Barium meal and follow-through showed normal small-bowel appearances but the transit time was extremely short with contrast medium appearing in the colon after only 15 minutes (normal >1 h). The following serum intestinal hormones were in the normal range: somatostatin, gastrin, glucagon, neurotensin, vasoactive intestinal polypeptide, pancreatic polypeptide. An initial jejunal biopsy specimen was morphologically normal with normal disaccharidase levels. Repeat jejunal and gastric biopsies showed mild lymphocytic infiltration but the jejunal villous architecture was preserved. Gliadin antibodies were absent. Colonoscopy was attempted but the patient deteriorated during the procedure and only a rectal biopsy specimen could be obtained. This showed pronounced depletion of goblet cells and scattered apoptotic cells, a picture resembling graft-versus-host disease and indicating an autoimmune process. Serum antibodies to epithelium were absent but an IgG antibody directed against mucin which stained the inclusions in goblet cells was demonstrated by indirect immunofluorescence.

Pancreatic enzyme supplements and elemental diet had not prevented diarrhoea and weight loss, so immunosuppressive treatment was tried with steroids, oral and intravenous cyclosporin, tacrolimus and intravenous gammaglobins. This too was ineffective. The patient lost weight rapidly despite high oral caloric intake and eventually required partial parenteral nutrition. This initially led to a good weight gain but she died from what

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Table 1 **Autoimmune disorders in the patient**

Age (year)	Diagnosis	Investigations
4.5	Monoarthritis left knee	SCAT and ANA negative
7.0	Autoimmune thyroiditis	Microsomal and thyroglobulin antibodies negative
13.5	Progressive dementia	MRI and CT scan: generalized cerebral atrophy CSF studies: 8 oligoclonal bands suggesting autoimmune process Anti-neuronal antibodies negative
15.5	Insulin-dependent diabetes mellitus	Islet-cell antibodies positive
15.7	Immune complex deposition in cornea	
18.0	Partial lipoatrophy	Insulin antibodies positive

SCAT=Sheep cell agglutinin test; ANA=antinuclear antibodies; MRI=magnetic resonance imaging; CT=computed tomography; CSF=cerebrospinal fluid

clinically appeared to be overwhelming sepsis. No pathogenic organism was isolated.

COMMENT

The autoimmune enteropathy in this patient was unusual in showing absence of epithelial antibodies but presence of goblet-cell antibody. We have only found one other report, from Australia, describing a similar patient—a boy aged 4 with near normal epithelial cells on biopsy but total destruction of goblet cells in the presence of a goblet-cell antibody[1,2]. In view of the normal pancreatic function tests we cannot explain the fat malabsorption but we postulate that it was due partly to lack of mucin, arising from the absence of functioning goblet cells.

For treatment, cyclosporin has been reported beneficial in several cases of typical autoimmune enteropathy[3,4] but this drug was ineffective in the Australian patient as in ours. These two cases suggest a variant of autoimmune enteropathy resistant to existing immunosuppressive therapy.

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