End stage renal disease due to bilateral renal malakoplakia

S Hegde, M G Coulthard

Malakoplakia typically affects the bladders of immunocompromised adults who have defective intracellular killing of *Escherichia coli*. Renal malakoplakia is rare in children and generally has a good outcome. In the case presented, however, it caused end stage renal failure in a 5 year old girl. The management dilemmas surrounding renal transplantation are highlighted.

**CASE REPORT**

A 5 year old girl was febrile, shocked, and encephalopathic after three days of bloody diarrhoea and two days of anuria. This presentation, plus her biochemistry (plasma creatinine 7.3 mmol/l, ionised calcium 0.6 mmol/l, and phosphate 3.2 mmol/l) and her haematology results (haemoglobin 88 g/l, white cells 16×10⁹/l, neutrophils 9×10⁹/l, platelets 89×10⁹/l) suggested haemolytic-uraemic syndrome, though she did not have a typical blood film. Investigations six months earlier for poor weight gain had shown normal urinalysis, full blood count, plasma creatinine, and blood pressure. Her older sister had had a ganglioneuroma resected at 5 years of age.

She was treated with fluid resuscitation, and intravenous cefotaxime before transfer to our unit for peritoneal dialysis. Computed tomography of her head, cerebrospinal fluid examination, and metabolic screen were normal. Blood culture grew *Escherichia coli*, but urine was not obtained before antibiotic treatment. Stool grew no pathogens. C reactive protein was raised at 459 mg/l. Renal ultrasound appearance changed over eight days from being slightly enlarged (78 mm long) with decreased corticomedullary differentiation, to being 88 mm long with a well defined right lower pole hypechoic mass which distorted the internal architecture. These findings were confirmed on computerised tomography, and the mass was biopsied.

Most of the glomeruli were sclerosed, the tubules contained neutrophils and necrotic debris, and the interstitium was massively expanded by a cellular infiltrate (fig 1A) including abundant large histiocytes with eosinophilic cytoplasm that had intracellular antibacterial activity, but no other pathognomonic laminated basophilic calcospherules called Michaelis-Gutmann bodies.

We are aware of only eight paediatric case reports involving the renal parenchyme in the English literature (see table 1). Escherichia coli was cultured from the urine of all eight, and the blood of four. The first case died, and two with unilateral involvement were successfully treated with nephrectomy. Of the five remaining bilateral cases, three were treated with antibiotics alone, of whom one died and two made a full recovery. Two additionally received prednisolone, plus azathioprine in one case; both recovered fully. We report a unique case of bilateral renal malakoplakia producing end stage renal disease in a 5 year old girl.

### Table 1

<table>
<thead>
<tr>
<th>References</th>
<th>Age (y)</th>
<th>Sex</th>
<th>E. coli cultured</th>
<th>Kidneys involved</th>
<th>Medical/surgical therapy*</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ravel (1998)</td>
<td>3</td>
<td>F</td>
<td>–</td>
<td>+</td>
<td>2</td>
<td>Dead</td>
</tr>
<tr>
<td>Trillo and Lorent (1997)</td>
<td>9.5</td>
<td>F</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Nephrectomy and Full recovery</td>
</tr>
<tr>
<td>Deridder et al (1996)</td>
<td>3</td>
<td>F</td>
<td>–</td>
<td>–</td>
<td>1</td>
<td>Nephrectomy and Full recovery</td>
</tr>
<tr>
<td>Wiglinkhuizen et al (1995)</td>
<td>0.5</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>2</td>
<td>Dead</td>
</tr>
<tr>
<td>Saleem et al (1994)</td>
<td>0.15</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>2</td>
<td>Full recovery</td>
</tr>
<tr>
<td>Honjo et al (1993)</td>
<td>0.09</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>2</td>
<td>Methylprednisolone and Full recovery</td>
</tr>
<tr>
<td>Present study</td>
<td>3.5</td>
<td>F</td>
<td>–</td>
<td>+</td>
<td>2</td>
<td>End stage renal failure</td>
</tr>
</tbody>
</table>

*Medical therapy excludes antibiotic treatment, which they all received.
DISCUSSION
Childhood renal parenchymal malakoplakia is rare; our case is only the ninth in the English literature, and the first ever to develop end stage renal failure. Unlike the majority of adult cases,\(^1\) she had no malignancy, immunodeficiency syndrome, or autoimmune disease, and had not been receiving steroids or immunosuppressant drugs. Indeed, analysis of her leucocyte function showed no abnormalities of bacterial killing.\(^9\)

However, we did not immunosuppress her with steroids or azathioprine as others have reported,\(^7,8\) because this has been implicated in the genesis of malakoplakia.\(^1\) Indeed, patients who developed malakoplakia while on immunosuppressive therapy for other reasons have been shown to have reversal of their clinical and leucocyte abnormalities after withdrawal of these agents.\(^10\) This poses a difficult management dilemma in our patient; renal transplantation is universally recognised to be the long term treatment of choice in children, but inevitably requires lifelong immunosuppression. If this can induce rare cases of de novo malakoplakia in the transplanted kidney\(^11\) and other organs\(^10\) of patients with no previous history of this condition, we fear that our child may be at particularly high risk of a recurrence of malakoplakia in the graft. Nonetheless, because of the much better quality of life that paediatric renal transplantation provides we plan to undertake this, using minimal immunosuppression, and long term cotrimoxazole.

Authors’ affiliations
S Hegde, M G Coulthard, Department of Paediatric Nephrology, Royal Victoria Infirmary, Newcastle, UK

Correspondence to: Dr M G Coulthard, Department of Paediatric Nephrology, Royal Victoria Infirmary, Newcastle NE1 4LP, UK; malcolm.coulthard@nuth.northy.nhs.uk

Accepted 12 March 2003

REFERENCES
End stage renal disease due to bilateral renal malakoplakia

S Hegde and M G Coulthard

Arch Dis Child 2004 89: 78-79

Updated information and services can be found at:
http://adc.bmj.com/content/89/1/78

These include:

References

This article cites 11 articles, 1 of which you can access for free at:
http://adc.bmj.com/content/89/1/78#BIBL

Email alerting service

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections

Articles on similar topics can be found in the following collections

- Artificial and donated transplantation (146)
- Homicide (20)
- Immunology (including allergy) (2018)
- Renal medicine (273)
- Urology (446)
- Child health (3922)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/