The Diagnosis of Malacoplakia of the Kidney by Percutaneous Renal Biopsy*

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SUMMARY

The case report describes a patient who presented with the haemolytic uraemic syndrome and was found to have bilateral palpable kidneys. Percutaneous renal biopsy showed malacoplakia. It is probable that the malacoplakia together with the disseminated intravascular lesions were the result of Escherichia coli infection. Malacoplakia should be considered in the differential diagnosis of bilaterally enlarged kidneys.


Malacoplakia (Greek: malak6s—soft, plax—a plaque) is a granulomatous lesion which occurs in the urinary tract, bladder, ureters, renal pelvis, and kidney;1 in the testis,2 and stomach and colon.3 It is generally held to be a benign inflammatory process which regresses spontaneously.2 Common diseases, such as coliform infections, tuberculosis, and sarcoidosis, have been frequently associated with malacoplakia.

The haemolytic uraemic syndrome is characterized by renal failure, micro-angiopathic haemolytic anaemia, and sometimes thrombocytopenia with typical histological changes in the kidney.4 Coagulation studies, histological changes, and peripheral blood findings suggest that the pathogenesis is disseminated, intravascular coagulation.4 This article describes a patient with malacoplakia of the kidneys and micro-angiopathic haemolytic anaemia, both believed to arise from a common aetiological factor, viz. a coliform bacilli infection of the kidney.

CASE REPORT

A Bantu female, aged 30 years, was admitted to hospital with a history of haemoptysis, haemetemesis and haematemesis, and a thrombocytopenia of 10 000/mm³.

The patient was mentally confused, unco-operative, and her speech was slurred.

Physical examination showed multiple purpuric spots on her upper chest wall, a right conjunctival haemorrhage and a pericardial effusion. Her blood pressure was 140/90 mmHg, and her heart sounds were normal, as was her respiratory system. The abdomen was slightly distended, there was no hepatomegaly, but bilateral renal masses were palpable. Central nervous system tests showed mental dullness, a right hemiparesis and hemi-anaesthesia, and loss of the right corneal reflex.

She was extremely ill and prednisone (20 mg t.i.d.) was administered. Initial investigations showed a normocytic, normochromic anaemia of 7.7 g/100 ml; leucocytes 6 000/mm³; platelet count 14 000/mm³; prothrombin index 100%; bleeding time longer than 8 minutes; and clotting time 7 minutes and 10 seconds. The fibrinogen level was not estimated during the acute phase. Seventeen days after admission, the level was normal, 371 mg/100 ml. Blood urea was 333 mg/100 ml. Urinary volume for 24 hours was 2 140 ml with pus cells, red blood cells, and a moderate growth of E. coli.

Within 7 days the patient improved markedly. There was no new evidence of haemorrhage. The platelet count rose to 171 000/mm³, blood urea was less than 200 mg/100 ml, creatinine clearance was 36 ml/minute. The blood picture was that of a micro-angiopathic anaemia. In spite of blood transfusion the haemoglobin dropped from 11.7 g/100 ml to 7.9 g/100 ml. The white cell count was 26 000 with a neutrophilia. Repeated urine examinations showed albumin, pus cells and E. coli on culture.

Three weeks after admission the patient still had leucocytosis, and micro-angiopathic haemolytic anaemia with a marked reticulocytosis. The platelet count was normal and the urea was 101 mg/100 ml. Clinically, she was mentally normal, without any speech defect, and with a mild residual spastic hemiparesis. Cerebrospinal fluid examination, brain scan and an electro-encephalogram were normal.

An intravenous pyelogram (IVP) showed bilateral uniformly enlarged kidneys (left kidney 17.5 cm, right kidney 17.0 cm in their long axes) with normal renal contours, and a renal angiogram showed superior displacement of the interlobar artery in the left kidney (Fig. 1).

Renal biopsy (Figs. 2, 3 and 4) showed normal glomeruli, but the medulla and part of the cortex were infiltrated with histiocytes and there was loss of the normal architecture. Numerous Michaelis-Gutmann calcospherites were present in these areas. These features are typical of malacoplakia. Iron deposits were present. In addition there was evidence of focal pyelonephritis. There was arteriosclerosis of the larger vessels but no fibrinoid necrosis or other pathology of the small vessels.

Ten weeks after admission the patient still had a mild leucocytosis, a normocytic, normochromic anaemia, a normal platelet count, a normal reticulocyte count and normal blood urea, but a persistent urinary tract infection. Clinically the patient was well, but the kidneys remained palpable and she still had a mild residual hemiparesis. The prednisone had been progressively reduced and then stopped. Her urinary tract infection was treated...
Fig. 1. Renal angiogram showing bilateral enlarged kidneys with normal contours. Vascular pattern appears normal except for the stretching of the interlobar vessel in the left kidney. The right kidney measured 17 cm and the left kidney measured 17.5 cm in the long axes.

with ampicillin, nalidixic acid and gentamycin, and her urine was sterile on discharge from the ward, 4 months after admission.

A second IVP showed reduction of kidney size.

DISCUSSION

Malacoplakia, as a disease process, was first recognized by Michaelis and Gutmann in 1902 and was subsequently described in greater detail by Von Hansemann. The lesions in the urinary tract comprise soft, pale-yellow, flat or umbilicated elevations about 6 mm in diameter, with peripheral hyperaemia. They may be confluent. Microscopically, they are composed predominantly of large xanthomatous cells containing the characteristic Michaelis-Gutmann bodies. These cells are thought to be of connective tissue origin. The plaques do not extend beyond the mucosal layers, but have been reported to involve the renal parenchyma by extension from the renal pelvis.

The Michaelis-Gutmann bodies are inclusion bodies consisting of concretions of calcium, well demonstrated by the von Kossa stain. Small amounts of iron have also been found. These bodies have been produced experimentally by incubating broth, blood, and urine with E. coli bacilli. These bodies are not specific for malacoplakia; similar bodies have been described in basal cystitis.

Bunting has emphasized the association of iron and calcium in interstitial deposits of various types, throughout the body.

The aetiology of this granulomatous reaction is obscure. It is believed to be a chronic inflammatory response to a
nonspecific irritation. It has been associated with tuberculosis, sarcoidosis, and coliform-bacilli urinary tract infections. According to Bleisch and Konikov, any theory for the genesis of malacoplakia must explain: (i) the large phagocytic cells and their inclusions; (ii) the frequent association of coliform bacilli with the disease, and yet the rarity of the disease compared to the higher incidence of coliform urinary-tract infections; (iii) the association of the disease with tuberculosis; and (iv) the 4 to 1 preponderance of the disease in females. The incidence of coliform urinary-tract infections and tuberculosis is high in our Bantu female patients. Our patient had no evidence of tuberculosis on chest radiography and investigation of the urinary tract, including cystoscopy, was negative.

The pathogenesis of the neurological lesion could have been hypertensive arterial disease; haemorrhage from thrombocytopenia, or disseminated intravascular coagulation.

The patient's blood picture in the acute stage showed the typical features of micro-angiopathic haemolytic anaemia, viz. red cell fragmentation, reticulocytosis, and thrombocytopenia. The finding of multiple lesions suggests that the bleeding tendency and the abnormal blood picture could be due to disseminated intravascular coagulation.

It has been suggested that the haemolytic uraemic syndrome is an example of the Schwartzman reaction. We consider that the probable precipitating factor in our case was an endotoxin from the patient's own urinary-tract infection.

We think that this patient developed malacoplakia as the result of a chronic, coliform-bacilli infection of the urinary tract, and that an acute exacerbation precipitated disseminated intravascular coagulation.

### Fig. 3. Renal biopsy specimen showing numerous, black-staining Michaelis-Gutmann bodies (calcospherites). (Von Kossa stain.)

### Fig. 4. Renal biopsy specimen showing tubules containing inflammatory cells. (H. and E. stain.)

### Fig. 5. The blood picture and blood urea changes during the course of the disease.
Malacoplakia should be considered in the differential diagnosis of bilateral enlarged kidneys, which includes congenital polycystic kidneys and bilateral hydronephrosis. To our knowledge this is the first recorded case of malacoplakia diagnosed on percutaneous renal biopsy.

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REFERENCES

Mitral Valve Replacement with the
Starr-Edwards Prosthesis*

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SUMMARY
In 124 patients with mitral valve disease, valve replacement with a Starr-Edwards prosthesis was undertaken; 12 patients died after operation and there were 10 late deaths. Fourteen patients have residual disability, and in 9 patients periprosthetic valve insufficiency was corrected at a second procedure. The remaining 88 patients are leading normal lives and have been transformed from cardiac cripples into useful citizens.

Valve replacement is usually necessary in the management of mitral valve disease other than noncalcific, pure mitral stenosis. Reconstructive operations succeed in only a minority of young patients with pure mitral incompetence. The Starr-Edwards mitral valve prosthesis, in use in many centres, is easy and safe to insert and the final haemodynamic result is predictable. Its disadvantages are its expense, and the fact that it is a ball-and-cage prosthesis with a lateral flow orifice. As a foreign body in the circulation it carries the hazards of thrombosis with subsequent embolism; infective endocarditis with partial dehiscence of the sewing ring from the annulus, and haemolysis. Late poppet fatigue is unlikely with the presently available valve, the ball of which is metallic.

Our surgical experience of mitral valve disease includes valve replacement with the Hammersmith, Beall and Starr-Edwards prostheses and mounted inverted aortic homografts; reconstructive procedures include annuloplasty, cusp extension and chordal repair. The purpose of this article is to outline the manner in which patients have been selected for mitral valve replacement, and the operative results and long-term surveillance in a consecutive series of 124 patients in whom mitral valve replacement with a Starr-Edwards prosthesis was undertaken. Experience extends over 3 years. During the period in which 124 Starr-Edwards mitral valves were inserted, 30 mounted, inverted, aortic homografts (with 2 operative deaths) and 14 Hammersmith valves (with 3 operative deaths) were inserted in the mitral area, and 30 conservative, reconstructive procedures were undertaken for mitral incompetence. The Hammersmith valves inserted in the series were the only valves available. Homograft valve replacement was undertaken when a suitable homograft valve was available.

Age and sex are outlined in Fig. 1. Table I is a classification of the surgical pathology of the mitral valve, with an explanation of the criteria for subdivision into 6 groups. In Table II are listed the pre-operative disabilities.