Renal myelofibrosis: an unusual cause of renal impairment

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Case History

A 58-year-old man, diagnosed with myelofibrosis 9 years previously, presented with a 2 month history of increasing breathlessness, malaise and ankle oedema. He had undergone splenectomy 4 years earlier, and had received intermittent treatment with hydroxyurea. On admission he was taking fluoxetine and penicillin V. Examination revealed him to be hypertensive (170/90) with widespread lymphadenopathy (left and right inguinal, left axilla and left supraclavicular nodes). Investigations showed Hb 8.1 g/dl, MCV 100 with a leucocythoerythroblastic film, plasma creatinine 198 μmol/l, urea 10 mmol/l and normal electrolytes. Lymph node and bone marrow biopsies showed myelofibrosis Renal ultrasound revealed normal sized kidneys of increased echogenicity consistent with an infiltrative process.

The patient subsequently underwent a renal biopsy, histology of which showed extramedullary haematopoiesis and myelofibrosis infiltrating renal and perirenal tissue (Figure 1). The patient subsequently received chemotherapy with ‘FLAG’ (fludarabine, cytarabine, arabinoside and G-CSF), with recovery of renal function (creatinine 80 μmol/l) and blood pressure returning to normal.

Discussion

Extramedullary haematopoiesis (EMH) is usually a complication of chronic haematological conditions such as myelofibrosis, and normally occurs in the reticuloendothelial system (liver, spleen and lymph nodes), and less commonly in the pleura, pericardium, adrenal glands and kidneys [1–3]. EMH is often asymptomatic, may cause organomegaly and rarely end-organ damage.

Renal parenchymal EMH presenting as renal failure responsive to chemotherapy and radiotherapy has been previously described [3]. EMH may also involve perirenal tissue and the collecting system [4–6]. In this case extramedullary haematopoiesis was found in the renal medulla and in the hilar connective tissue where it was associated with myelofibrotic tissue similar to that found in the marrow and the lymph node. It is probably this component of the infiltrate which caused obstruction and which responded to chemotherapy. Extramedullary haematopoiesis at this site probably preceded infiltration by the myelofibrotic process. Renal extramedullary haematopoiesis and myelofibrosis is a rare, but treatable cause of renal impairment.

Suggested Reading

Fig. 1. (A) The needle biopsy core at low power shows renal medullary tissue on the left (see B) and the connective tissue of the renal pelvis on the right (see C). (H&E ×100). (B) Clusters of cells with small dark nuclei between the renal medullary tubules are islands of extramedullary haematopoiesis. (H&E ×400). (C) In the connective tissue of the renal pelvis extramedullary haematopoiesis is seen as clusters of small dark nuclei. The large atypical megakaryocytes and the sclerotic stroma represent myelofibrosis. (H&E ×400).