Retroperitoneal Malignant Fibrous Histiocytoma with Renal Involvement

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Summary

Malignant fibrous histiocytoma is a mesenchymal tumour which can involve the genitourinary organs primarily or by secondary extension. Both conditions are rare. We report four cases of retroperitoneal malignant fibrous histiocytoma involving the kidney by local extension. Diagnosis was difficult because of diverse, non-specific clinical features and may only be reached at operation or post mortem. Prognosis is poor. Although en bloc tumour resection with nephrectomy was possible in two patients, they returned with recurrences.

Key Words: Malignant fibrous histiocytoma, Kidney, Retroperitoneum

Introduction

Malignant fibrous histiocytoma (MFH) was first described by O'Brien and Stout[1]. Retroperitoneal MFH can occur as a primary lesion of the kidney or involve the kidney by local extension. The latter appears to be less common[2].

Four such cases presented to our institution between 1974 and 1984. The rarity of this condition stimulated this review of the clinical, radiographic and pathological features of our patients.

Case Reports

Case 1

A 69-year-old woman, with history of duodenal ulcer, presented with abdominal pain and weight loss. On examination there was a tender mass in the right upper abdomen. A provisional diagnosis of gastric carcinoma was made.

Investigations showed that haemoglobin was 9.6 g/dl; serum electrolytes and creatinine were normal. Gastroscopy revealed no mucosal pathology but an extramural compression of the posterior gastric wall. CT scan showed a large right renal mass with extension across the midline, suggesting a right renal tumour. Urine analysis and microscopy were normal. Intravenous pyelogram (IVP) demonstrated a non

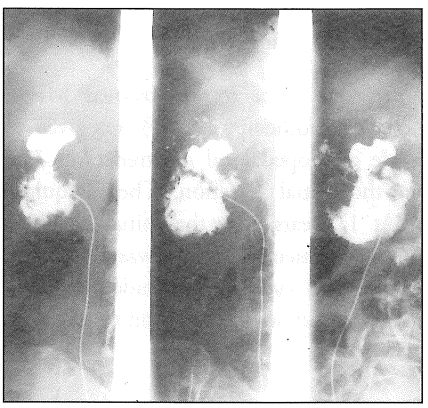

Fig. 1: Case 1. Dilated right calyceal system with leakage of contrast into the renal parenchymal

functioning right kidney. The left system was normal. Retrograde pyelogram showed moderately distended right upper and middle calyces. The lower calyces were irregular with leakage of contrast into the renal parencymal (Fig. 1). An arteriogram showed the lesion to be avascular.

At laparotomy a 15cm x 10cm retroperitoneal tumour adherent to the posterior abdominal wall, encasing the right kidney, inferior vena cave and aorta was found. Superiorly the tumour reached behind the stomach causing indentation of its posterior wall. Resection was not possible. A diagnostic biopsy was taken. Histology was that of a retroperitoneal MFH. She died two months after operation.

Case 2

A 41-year-old man, presented with fever, night sweats, dry cough, anorexia and weight loss. He was found to have an abdominal mass consistent with splenomegaly. Differential diagnoses included malaria, lymphoma and myeloproliferative disorders.

Blood films for malaria parasites were negative. Serum electrolytes and creatinine were normal. Ultrasound showed a normal spleen. However, there was a solid retroperitoneal mass displacing the left kidney anteriorly. IVP revealed a dilated left calyceal system. A diagnosis of retroperitoneal tumour with left renal involvement was made.

At laparotomy a tumour measuring 20cm x 25cm was found adherent to the left kidney. The tumour was excised en bloc with left nephrectomy.

Histopathology was one of a retroperitoneal MFH. Postoperative adjuvant radiotherapy (44Gy) was given to the tumour bed. He developed local recurrences at 7, 8 and 9 years after the initial resection. These required further excisions. At 11 years after the initial diagnosis he developed pulmonary metastases. He was started on adriamycin, vincristine and cyclophosphamide but failed to complete the course and was lost to follow up.

Case 3

A 65-year-old woman presented with history of left loin pain, fever, anorexia and weight loss. She was found to be hypertensive (BP=180/100 mmHg) with a left lumbar mass. The provisional diagnosis was a left renal tumour with hypertension.

Full blood count, serum electrolytes and creatinine were normal. Urine analysis and microscopy were unremarkable. CT scan showed a mass arising from the upper pole of the left kidney measuring 9 x 10 cms. It was suggestive of a renal cell carcinoma. IVP showed the mass displacing the left upper and middle calyces downwards.

The patient's blood pressure was controlled with prazosin 0.5mg tds, propranolol 40mg bd and frusemide 40 mg daily. At operation a large tumour was found adherent to the left kidney. En bloc excision of tumour and nephrectomy was performed.

Histopathology showed a retroperitoneal MFH. She developed local recurrence 2 years later and defaulted before treatment was effected.

Case 4

A 36-year-old man presented with progressive weakness, fatigue, fever and weight loss. Examination revealed the patient to be severely anaemic with hepatosplenomegaly.

Investigations were directed towards haematological disorders; haemoglobin was 7.3 g/dl, white cell count 14000/ul (90% neutrophils) and ESR 142mm/hr. Serum alkaline phosphatase was 800 IU/l. Direct Coomb's test was negative. Skeletal survey was normal. Electrophoresis showed no evidence of monoclonal gamaglobulinaemia. Fe^{59} utilization was above normal. Cr^{51} red blood cell survival was below normal. Bone marrow biopsy showed abnormal plasma cells. A diagnosis of plasma cell dyscrasia was made. He was treated with blood transfusion and melphalan.

One year after the initial diagnosis, he was readmitted on five further occasions with anaemia requiring blood transfusions. On his sixth admission he developed cardiac failure and gross abdominal distention. He died during admission.

A post mortem showed that the entire left kidney and the upper pole of right kidney were replaced by tumour masses. Histopathology of the left mass showed

necrotic tissue; the right mass proved to be retroperitoneal MFH. There were metastatic nodules in the spleen, liver and the lumbar vertebrae.

Discussion

Retroperitoneal MFH is often difficult to diagnose because patients present with non-specific clinical features as illustrated by our patients. Even though the kidneys were involved by local spread of the disease, none of our patients presented with urinary symptoms. The only consistent feature was the presence of abdominal masses.

Urine analysis, microscopy and creatinine were normal in all cases. Case 4 demonstrated the diagnostic difficulty in the days when limited radiological facilities were available and the actual diagnosis was only made at post mortem. Presently, our ability to diagnose this condition has been facilitated by the availability of ultrasound and CT scan. Despite this it is sometimes not possible to distinguish a renal tumour from retroperitoneal MFH, as illustrated in Case 3.

Review of all four cases showed essentially similar histological features, the tumours were all of the storeiform-pleomorphic type (Fig. 2). There was generally clear evidence of fibroblastic proliferation with collagen deposition by tumour cells, as well as the large numbers of histiocytes. Aggregates of foamy macrophages and foci of infiltrating lymphocytes were a constant feature. Tumour giant cells as well as Touton-type and foreign body giant cells were present. Necrosis was, as a rule, minimal. In two cases, there were areas of histological differentiation reminiscent of hemangiopericytoma; metaplastic bone formation was present in Case 2. Immunohistochemical markers showed that the majority of the tumour cells were positive for vimentin, alpha 1-antitrypsin, alpha 1-antichymotrypsin.

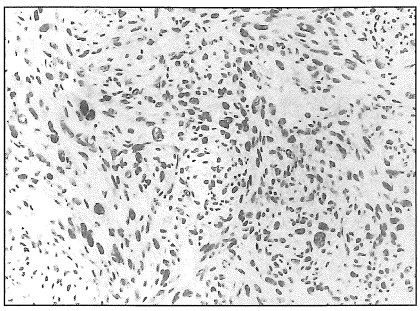

Fig. 2: There is cellular atypia and pleomorphism, scattered tumour giant cells, focal lymphoid aggregates with storeiform arrangement of the sarcomatous cells. (H&E stain X 200)

The prognosis of retroperitoneal MFH is worse than other sites because of its late and non-specific presentation. The 5-year survival rate is about 14 per cent[3]. Little success have been obtained with adjuvant therapy. The most effective treatment is complete surgical excision[3]. Unfortunately this may not be possible because of tumour size, proximity to or involvement of vital structures. In this series, in the two cases where en bloc excision of tumour with nephrectomy was possible, both patients had reasonable disease free survival period with recurrences at 2 and 7 years after resection. The latter demonstrated that prolonged survival was possible with repeated resection of recurrences. Unfortunately both of them were lost to follow up so their exact periods of survival were not known. We advocate en bloc resection of retroperitoneal MFH with nephrectomy in cases where resection is possible.

Although retroperitoneal MFH with renal involvement is rare the diagnosis should be suspected in patients who present with suspected renal tumours but with no obvious symptoms and signs referable to the genitourinary system.

References

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