RENAL NEOPLASMS — CLINICAL CASE

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CASE HISTORY: A 25-year old female patient was referred to St. Luke's Hospital with a history of hypertension detected 4 months previously when the patient visited her G.P. for a sore throat. She was then put on Bendrofluozide and a salt-free diet pending investigations. Systemic enquiry of the patient only revealed frequent frontal headaches, slight dyspnoea on exertion and a slight ache over her loins. She gave no history of haematuria or dysuria, no ankle oedema or puffiness around the eyes, no loss of weight, nausea or vomiting. The patient had been operated two years previously for a lump in the breast which histology showed to be due to mammary dysplasia. There were no remarkable features in her family history.

EXAMINATION: Her general physical condition was satisfactory, except for clinically detectable anaemia. Her tonsils were not enlarged of inflamed and there was no cervical lymphadenopathy. Her B.P. was 150/90. Pulse was regular at 96/minute and heart sounds were normal with no murmurs or extra sounds. Her JVP was not raised. Her respiration was regular with chest expansion equal on both sides. There were no signs of mediastinal shift. Breath sounds were vesicular and adventitious sounds were heard. Percussion was normally resonant.

The abdomen was not distended and moved symetrically with respiration. On palpation there was slight tenderness in the right iliac region and the right kidney was felt to be enlarged. There was no hepatosplenomegaly and no evidence of ascites.

The patient was kept at S.L.H. for a period of investigation of ten days during which she was noted to be occasionally pyrexic. Her diastolic pressure remained in the region of 90-100 mmHg. The patient was discharged on Bendrofluozide and was told to attend the outpatient department pending completion of her investigations.

INVESTIGATIONS: **Blood** — Haemoglobin 11.3 mg%; white cell count 9,300/mm^3 with a differential count of 69% neutrophils, 1% eosinophils, 21% lymphocytes and 9% monocytes. The blood picture showed slight hypochromia with anisocytosis and poikilocytosis. The neutrophil series exhibited a shift to the left. Platlets were normally represented. Serum protein electrophoresis showed no significant departure from the normal pattern with a total protein of 6.8 gm/dl.

Urine — pH 6.0, protein and glucose absent (a repeat showed traces of protein). Microscopy showed rare RBC's occasionally WBC's and a few transitional cells. Culture revealed a streptococcal species.

Renal function tests — Blood urea 17 mg%, serum creasinine 1.0 mg% with a creatinine clearance of 69.

Radiology — A chest X-ray showed no abnormalities. A plain abdomen film showed the right kidney to be obscured by an indistinct shadow of soft density. An intravenous pyelogram (IVP) showed the right pelvicalyceal system to be displaced by pressure from above. This was caused by a large space-occupying mass growing near the upper pole of the kidney. The left kidney, pelvis and ureter, and the bladder were normal.

DIFFERENTIAL DIAGNOSIS: The tentative diagnosis at this point was that of a **renal tumour.**

Truly benign tumours of the kidney are so rare that it is considered best to treat all kidney neoplasms detected clinically as malignant. In general, benign tumours are trivial and clinically insignificant. The angiomas may however give rise to profuse haematuria. The commoner malignant tumours of the kidney are characteristically seen in two periods of life: during infancy below the age of 7 years, and during adult life after the age of 40 years. Between the ages of seven and forty malignant tumours are unusual and present a totally different pathology.

(TABLE 1).

The clinical presentation of renal tumours depends on their pathology. The presence of blood in the urine is usually noticed by the patient and makes him seek early advice. Its transient or intermittent nature is however an important cause for delay. Pain is not a prominent feature and is usually of a continuous dull aching character producing discomfort or a feeling of weight in the loin. Renal colic is rare and is often due to the passage of blood clots through the ureter. An intermittent pyrexia with an evening temperature reaching 103°F may be a feature. A swelling in the loin may be a relatively late presentation in adult tumours but is the prime presenting feature in the nephroblastoma. A raised sedimentation rate (ESR), hypertension or polycythaemia which have no ready explanation should all raise suspicion of a renal tumour.

Along with the differential diagnosis of a renal tumour one must consider a **solitary renal cyst.** These are usually globular and unilateral. The patient usually complains of a painless mass. This is smooth, globular and tense, Large cysts may cause a dull ache over the loin. The diagnosis depends on the pyelogram which demonstrates a globular mass with elongation and distortion of adjoining calyces. An encapsulated neoplasm may produce an identical picture making diagnosis difficult without the use of an arteriogram.

Adrenal tumours, particularly if malignant, may produce a pyelogram showing displacement of the upper pole of the kidney and possibly a distortion of the pevicalyceal system. Three tumours may originate from the adrenal medulla. The **ganglioneuroma** may be found in 15% of cases. It is a symptomless relatively benign tumour which may occur at any age and may grow to a large size. Affecting children below the age of five years is the highly malignant **neuroblastoma.** This usually presents as an abdominal swelling with signs and symptoms attributable to metastasis and general effects of malignancy. The common adrenal medulla tumour affecting adults it the **phaeochromocytoma** which may attain a diameter of about 5 cm. This produces intermittent or continuous catecholamine secretion which gives rise to the symptoms of paroxysmal or persistent hypertension, headache, palpitations, vomiting, dyspnoea, weakness and pallor.

Tumours of the adrenal cortex are usually functional. These include the small rare adrenocortical adenoma which causes features of **Primary Aldosteronism.** Sodium retention and a fall in potassium give rise to the features of hypertension and episodic muscular weakness associated with polyuria and polydypsia. Another functioning tumour of the adrenal cortex is that which may give rise to **Cushing's syndrome** or the **Adrenogentital syndrome.** These very rarely may be malignant large carcinomas weighing as much as 4,000 gm. Extension beyond the adrenal capsule with invasion of the kidney may occur with the more aggressive tumours. The benign adrenomas are well encapsulated tumours which may reach a mass of 200 gm.

OPERATION: The patient was readmitted ten days later for a right-sided transperitoneal nephrectomy. During operation a large tumour measuring 18 x 12 x 12 cm. overlying the lower pole of the right kidney was found. The tumour was attached to the inferior vena cava causing an aneurism containing a thrombus. A radical nephrectomy was performed and the tumour mass affecting the IVC was removed by resection of the anterior wall of the vein. Major bleeding could not be prevented and was controlled by massive blood transfusion and fluid infusion. The anterior wall of the IVC was repaired and mesh sutures were placed in the lower abdominal IVC to restrict embolism.

PATHOLOGY: On incision the renal tumour was shown to be bordered laterally by surviving renal tissue. The tumour was yellowish in ap-

TABLE 2: CLASSIFICATION & STAGING OF NON-HODGKIN LYMPHOMAS

FOLLICUAR LYMPHOMAS	DIFFUSE LYMPHOMAS	STAGING
Small lmphoid cell	Small lymphoid cell varients 1. Small lymphocytic 2. SL with plasmacytoid differentiation 3. Ttypical SL 4. Convoluted lymphocytic (thymic)	STAGE O:- no detectable disease STAGE I:- localization to single node or adjacent group of nodes.
Mixed lymphoid cell	Mixed lymphoid cell	STAGE II:- involvement of more than one region of nodes but on one side of diaphragm. A) without general symptoms B) with general symptoms.
Large lymphoid cell	Large lymphoid cell Varients 1. Histiocytic 2. Burkitt's 3. Mycosis fungoides 4. Undefined	STAGE III:- involvement of both sides of diaphragm. A) without general symptoms B) with general symptoms STAGE IV:- generalized disease demonstrable in bone, lungs, GIT, skin or kidneys.

pearance with extensive areas of haemorrhage. Microscopy showed a sarcomatous tumour with lymphoblasts as the component cells. There was extension into the inferior vena cava as evident from the microscopy of the thrombotic mass.

DIAGNOSIS: Diffuse Lymphocytic Lymphoma of the right kidney extending to the inferior vena cava.

POST-OPERATION MANAGEMENT: The patient did very well being covered with fluid therapy and broad spectrum antibiotics. Her subsequent anaemia was treated with blood transfusion and iron tablet therapy. She was discharged 19 days after operation and was referred for future management with chemotherapy.

LYMPHOMAS: Most patients with Lymphomas present as otherwise healthy individuals with painless enlargement of a single or group of lymphnodes usually in the cervical chain. Occasionally, evidence of extranodal involvement is already present and indeed 25% of patients have initial complaints referable to hepatosplenomegaly. With more advanced disease, systemic manifestation including fever, night sweats, marked weight loss, weakness and anaemia may be evident. As would be expected, the manifestation of advanced widespread disease is truly protean. Involvement of the gastrointestinal tract may produce diarrhoea, sometimes with the full blown picture of the malabsorption syndrome. Multiple osteolytic lesions with bone pains and pathological fractures may occur. Renal enlargement may result from direct lymphomatous infiltration or from hydronephrosis resulting from obstruction to the lower urinary tract by tumour. Lymphomatous infiltration of the kidney as evidenced at necropsy was found in 85% of non-Hodgkin's lymphomas, the majority (73%) being Burkitt's Lymphoma. Infiltration in Hodgkin Disease was found in 10% of cases. Invasion of the kidney sufficient to cause renal failure is practically confined to the lymphosarcoma and the reticulum cell sarcoma, and is the cause of death in only 0.5% of cases of renal lymphomas.

Nervous system involvement can create a bewildering array of central and peripheral findings. The peripheral blood examination and bone marrow aspiration is in most lymphomas usually normal. Some types of lymphoma, particularly the diffuse lymphocytic lymphomas, are associated with a corresponding leukaemia.

Primary lymphomas of various other organs have also been reported, however documented primary lymphomas of the kidney is rare. Therefore the unicentric theory commonly accepted for other lymphomas has been questioned in the case of renal involvement. Until 1956 it was assumed that lymphoma of the kidney was not primary. In that year Knoepp reported a case of solitary lymphoma in the right kidney which radical resection apparently cured. Other authors have since documented cases which support the unicentric theory of lymphomastous lesion in the kidney.

Prognosis depends on the histological pattern and staging of the disease (TABLE 2). The overall 5 year survival rate is about 25% being worse with stage IV disease with a diffuse histological pattern. Radiotherapy and chemotherapy have however in recent years altered considerably the natural course of many of the lymphomas.

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TABLE 1: CLASSIFICATION OF RENAL MALIGNANT NEOPLASMS

NEOPLASM	AGE	INCIDENCE	PATHOLOGY
Adenocarcinoma (Grawitz tumour)	55-60	80%	Cubical or polyhedral cells arranged as solid alveoli, papillary cysts of tubules. The strome is scanty and rich in large blood vessels. Various degrees of anaplasia may occur.
Nephroblastoma (Wilms's tumour)	1-4	8%	Primitive or abortive glomeruli with poorly formed Bowman's spaces and abortive tubules all enclosed in a spindle cell stroma including cells of muscle, fat, bone and cartilage.
Malignant haemartoma Sarcomas fibrosarcoma liposarcoma leomyosarcoma osteosarcoma neurofibrosarcoma rhabdomyosarcoma		2%	Connective tissue malignant tumours of fibrous tissue, fat, smooth muscle, etc. Various degrees of anaplasia may be seen.
Lymphosarcoma and leukaemic deposits		50% of generalized disease	Involvement of the renal parenchyma with the malignant lymphoid cells, usually associated with stage IV disease.
Metastatic carcinoma from Lungs Breast Stomach	variable	8% of malignant disease	Resemble the primary carcinoma.