Chapter 15

NEPHROBLASTOMA (WILM'S TUMOR)

It occurs in 1 per 10,000 live births and is probably present from birth. Recognition is in first 5 years of life with peak incidence at 2nd and 3rd year.

Tumor may arise in utero and be detected at birth or shortly afterwards but rare after 5 years.

Link between nephroblastoma and aniridia and other developmental malformation such as duplication has been reported.

Incidence of nephroblastoma is much higher in siblings of affected twins.

Tumor is larger than kidney at time of diagnosis and may be very large. Diagnosis of small tumor is by accident or due to investigation of hypertension which is due to renin production by tumor or kidney. Tumor is largely extrarenal when superficial but when deep seated compresses and displaces renal tissue.

Nephroblastoma is bilateral in 10% cases. This could be secondary spread or multifocal origin.

Nephroblastoma consists of embryonic renal tissue with epithelial or mesenchymal elements. Epithelial element varies in differentiation upto glomerular and tubular production and mesenchymal elements may be primitive or differentiated to muscle, osteoid tissue or cartilage. Local spread occasionally takes place. Blood spread is common. Secondary deposits may be found in lungs, liver and vertebrae.

Nephroblastoma must be distinguished from neuroblastoma and hamartomas.

CLINICAL FEATURES

Predominant signs are abdominal mass, abdominal distension and abdominal pain. Haematuria is uncommon. Systemic hypertension is present. Pyrexia and occasionally secondary mass detected on X ray chest are presenting features. Polycythaemia may exist due to excessive erythropoietin. High serum and urine levels of mucopolysaccharide isolated as hyaluronic acid are often obtained from child with disseminated nephroblastoma.

DIAGNOSIS

Child with abdominal pain, abdominal distension, haematuria and abdominal lump should be investigated with intravenous urogram.

Differentiation from other forms of malignant tumors and cysts is required. X ray chest and bones are indicated for evidence of secondary spread.

Plain X ray abdomen shows calcification around edge of nephroblastoma in 15% cases. Calcification is more commonly due to neuroblastoma and in that tumor urogram usually shows displacement of kidney outward and downwards. Occasionally contralateral kidney is also displaced.

Teratoma may be indistinguishable unless containing specific tissue such as teeth which can be shown radiologically.

Other conditions are hydronephrosis, renal cysts, renal venous thrombosis and pyelonephritis.

Tumor cells are seen in urine in few patients only.

Renal biopsy carries risk of spread of malignancy and hemorrhage.

Ultrasound is helpful in distinguishing between intrarenal cysts and tumors.

TREATMENT

A. Surgery:

Transperitoneal approach and early tying of renal vessels is advised.

B. Chemotherapy:

Actinomycin 15 microgram per kg for 5 days with first dose on day of operation. Leucocyte and platelet count should be monitored. Repeated courses can be given. Cyclophosphamide and vincristin are also used.

C. Radiotherapy:

Initial treatment prior to surgery will shrink tumor. Radiation nephritis may be produced.

Infant aged less than 1 year with operable lesion and with no evidence of secondary spread requires nephrectomy, added chemotherapy with optional radiotherapy.

Child aged more than 1 year should have nephrectomy, actinomycin D and radiotherapy to metastasis.

If infant or child has inoperable lesion chemotherapy and radiotherapy followed by nephrectomy later if possible is advised.

When patient has bilateral nephroblastoma nephrectomy on worse side with radiotherapy to smaller tumor is possible but seldom cures. Chemotherapy with actinomycin D or vincristin or both is helpful.

PROGNOSIS

Younger child has better prognosis. Localized tumor has much better prognosis. Bilateral tumors have poor prognosis.

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