

Renal Cell Carcinoma in Adolescents

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INTRODUCTION:

Malignant renal neoplasm: Because benign tumors of kidney are rare, so it is a good rule that all neoplasm of kidney which are recognized clinically should be treated as malignant. They are uncommon between the ages of 7&40⁽¹⁾. Renal cell carcinoma is the most common tumor of the kidney in adult. It arises from renal tubular cells.. It is rare in children & is more frequent in men than women. It is an adenocarcinoma & is the most common neoplasm(75%) of kidney. The tumor may invade locally & metastasize by way of blood stream & the lymphatics. Regional lymph nodes are involved in approximately 30% of patients, unfortunately sign & symptoms are usually meager until the disease is advanced. Gross haematuria is the most frequent presenting complaint. Diagnosis of mass in the kidney is best established by intravenous urogram(IVU). CT scan may confirm the diagnosis. Radical nephrectomy is treatment of choice & offers the only known chance of cure. The over all 5 years survival rate of patient is 35%. Angiomyolipoma (Hamartoma) is a benign tumor of the kidney that may rarely be confused with renal cell carcinoma. It occurs frequently in patient with tuberous sclerosis⁽²⁾. Wilms' tumor is the most common of malignant tumors of kidney in childhood. It is highly malignant mixed tumors consisting of connective tissue origin & epithelial structures ,it is usually discovered as palpable mass by mother or by examining physician. Treatment of primary tumor is radical nephrectomy. Adjunctive therapy with irradiation & chemotherapy is important in improving the survival. Transitional cell carcinoma is the most common tumor of renal pelvis & ureter. Squamous cell carcinoma may occur because of metaplastic changes. Clinical behavior of this tumor is dependant on degree of cellular differentiation & extent of invasion.⁽³⁾

Case Report :

A 12 -year-old boy was admitted to the hospital because of right hypochondrial pain & heaviness of 2 months duration , dull in it's nature ,it radiated to tip of right shoulder. He had history of weight loss (10 Kg) and loss of appetite during this period, with history of intermittent fever and night sweats with 2 attacks of painless haematuria.

On physical examination the patient was pale, right hypochondrial mass (smooth surface, rounded, 13x14cm in size, moved with respiration, tender & could not get above it). Investigation: revealed Hb was 9gm/dl, liver function tests were normal apart from serum alkaline phosphatase was 112 IU/L(30-85 IU/L normal value). ESR was 55 mm/hr, CXR was normal & normal urine examination. Abdominal ultrasonography (US) showed a rounded heterogeneous hyperechogenic right (Rt) hypochondrial mass of 12x11 cm ,displacing the Rt kidney tissue medially. The mass was of smooth outline with no cystic change with normal left kidney. Intravenous urography showed mass involving mid & upper pole of Rt kidney causing displacement & distortion of pelvicalyceal system, otherwise the kidney was functioning with no delay in excretion of contrast with normal left kidney. Fig. (1)

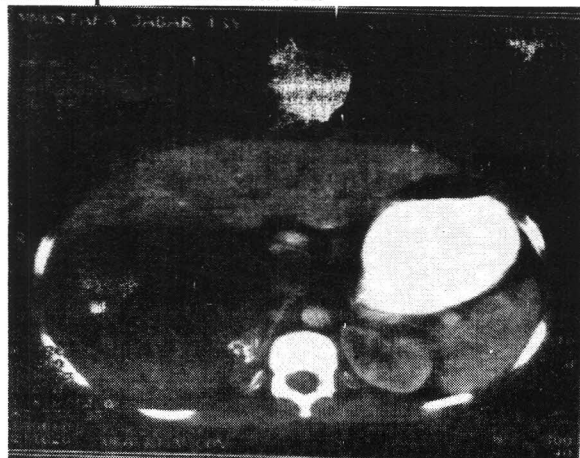
Figure (1): IVU showed evidence of large mass involving mid and upper pole of the right kidney.



CT scan of abdomen (Oral & IV contrast) revealed a well- defined large mildly hypo dense almost rounded mass occupying the whole of Rt kidney with few calcified spots seen at the inferomedial portion ,the mass pushed the intestinal loops inferiorly , it showed moderate thick walled mural enhancement after contrast ,with markedly hypo dense center, no associated renal vein or inferior vena cava involvement with regional lymphadenopathy (Radiological feature of Wilms'), left kidney was normal. Figure 2:

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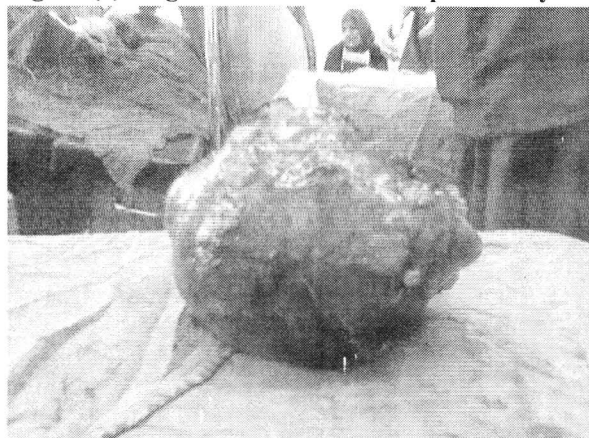
Figure (2): CT scan of the abdomen with contrast showing right renal mass with infero-medial spots of calcification.



At operation:

Radical right nephrectomy was done through long right paramedian incision & large right renal mass was found. After the peritoneum incised, pararenal fascia entered dissection of right kidney was done. Renal vein was gently mobilized using right angled clamp, the artery was located behind the vein and ligated in continuity (This may be more difficult from an anterior approach because the artery lies behind the vein) then renal vein gently palpated. There was no evidence of metastasize within its lumen, and divided between ligatures. The renal artery was divided and kidney mobilized along with its facial and fatty covering. The ureter was then divided as low as possible. Whole specimen was sent for histopathology along with few lymph nodes from nearby area figure 3.

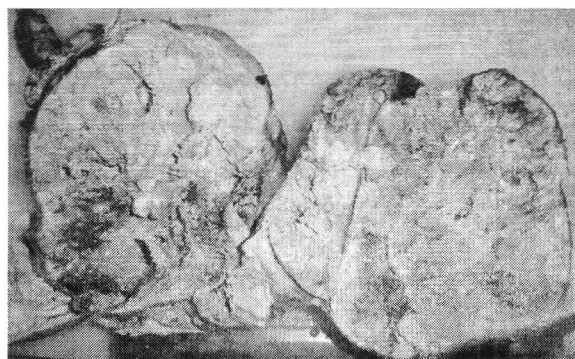
Figure (3): Right renal mass after nephrectomy



Histopathological examination revealed grossly that the kidney is extensively replaced by a large gray to tan colored mass (13x18cm) with areas of hemorrhage and necrosis. The residual renal parenchyma (8x2cm) delimits the tumor posteriorly and laterally. The specimen (kidney and its tumor) weights 1.200 g. The abnormal mass engulfs the

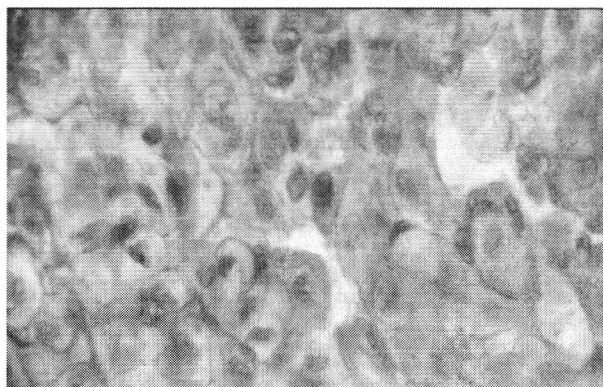
hilar region extensively and in such a way that it is difficult to delineate the pelvi-calyceal system, ureter and major hilar vessels. Fig. (4)

Figure (4): Macroscopical appearance of the right renal mass.



Microscopically : Renal cell carcinoma, showing classical nests and sheets of polyhedral cells with clear cytoplasm. In places glandular arrangements by eosinophilic malignant epithelial cells are noted fig.5.

Figure (5): Microscopical appearance of renal cell carcinoma



The cancer invades and destroys the pelvi-calyceal system and is extending to the hilum and hilar vessel. 3 N1 MX stage III.

Discussion:

Malignant renal neoplasm commonly occur below 7 and above 40 years of age. Renal cell carcinoma occurs in adult and rarely occurs in children. While Wilm's tumor usually discovered in the 1st 4 years of age⁽⁴⁾. The youngest patient presented with renal cell carcinoma was reported by (Leone)⁽⁵⁾ and the patient was about 16 years. While our case presented at 12 years of age. Although renal cell carcinoma usually presented as frequent attacks of haematuria⁽³⁾ on comparison to Wilms' tumor which presents usually as palpable mass by mother or physician. Our case was diagnosed by history of haematuria, right renal mass with fever, confirmed by IVU & CT scan. The findings were within the

features of Wilms' tumor clinically & radiologically (Well defined large mildly hypodense rounded mass with few calcified spots at inferomedial portion by CT scan). Surprisingly the histopathological finding revealed that the current case was renal cell carcinoma & the tumor engulfing pelvicalyceal system with area of hemorrhage & necrosis. This case is considered as unique because it is the first pediatric case of renal cell carcinoma at age of 12 years. Renal cell carcinoma should be kept in mind as one of malignant tumor occurring in the kidney at younger age group. Angiomyolipoma (Hamartoma) is very rare benign tumor of the kidney which can be regarded as one of the differential diagnosis of renal cell carcinoma in adolescents⁽²⁾.

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