PEDIATRIC CHROMOPHOBE RENAL CELL CARCINOMA: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

We report a case of an extremely rare pediatric renal tumour, chromophobe renal cell carcinoma in a 12 years old boy, who presented with dysuria and painless mass of right kidney. The clinical, radiological and pathological features of this neoplasm will be discussed, along with a review of the literature.

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INTRODUCTION

Chromophobe renal cell carcinoma is an anecdotal childhood tumour with distinct biological and histopathological characteristics [1]. To date, fewer than 20 cases of pediatric chromophobe renal cell carcinoma have been reported [2]and it is the first reported case of our institute.

CASE REPORT

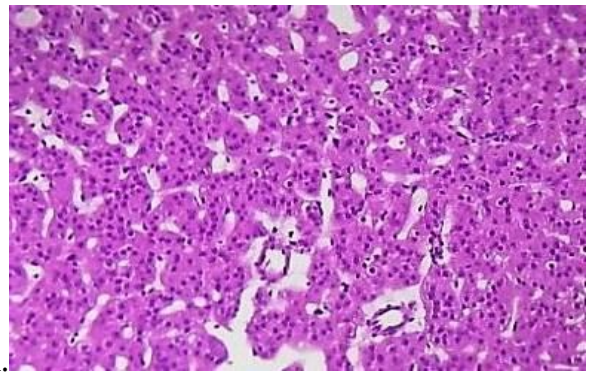
A 12 years old boy, student of class 7th, presented to our institute in December 2015 with complaints of dysuria and painless right sided abdominal swelling of short duration. Local examination of the abdomen revealed a mass in right lumbar region measuring 8 x 6 cm, firm in consistency, not attached to the overlying skin but seem to be arising from the right kidney.

Abdominal ultrasound revealed right renal mass with differential diagnosis of nephroblastoma and renal cell carcinoma. Contrast enhanced CT scan of abdomen was done, which reported the lesion as right renal tumour limited to the right kidney, followed by renal biopsy for exact typing of the tumour.

Histopathology of the renal biopsy revealed a tumour composed of cords and trabeculae lined by round to oval cells having monomorphic nuclei with few cells having perinuclear halo (Figure-1a). An extended panel of immunohistochemistry was applied which showed positivity for CK7, CD117, PAX8 (Figure 1b,c,d) and negativity for vimentin and CD10, thus a definitive diagnosis of chromophobe renal cell carcinoma was made.

Right sided nephrectomy was done and resected specimen was sent to some other institute for histopathology, which concurred with our diagnosis. So, patient was advised follow up by oncologist and he is doing well now.

a.



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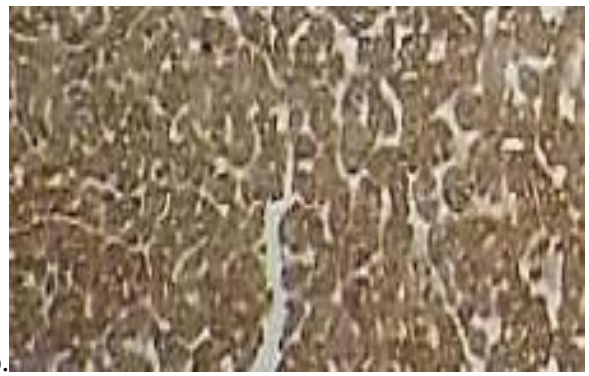

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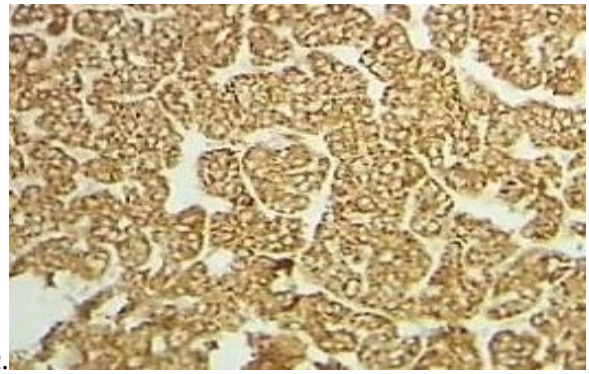
c. 

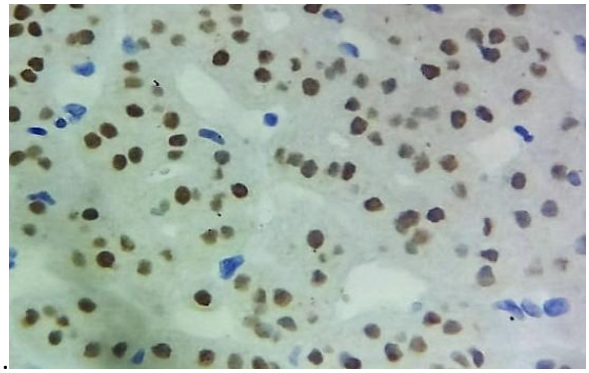
d. 

Figure-1. Photomicrograph of Chromophobe renal cell carcinoma

a. Haematoxylin-Eosin (original magnification 20x)

b. CK7 (original magnification 20x)

c. CD117 (original magnification 20x)

d. PAX8 (original magnification 40x)

DISCUSSION

Chromophobe renal cell carcinoma is a rare childhood tumour which was first reported by Fujii et al in 1998[1]. Renal cell carcinoma (RCC) represents 3-4.3% of all pediatric renal tumours, with a median age of presentation between 7-17 years without any gender predominance [3]. The reported incidence of pediatric RCC is 0.01/100000 population [4].

The most common presenting complaint of pediatric RCC is abdominal pain (43%), followed by hematuria (37%), abdominal mass (16%), fever (13%) and weight loss (5%). At the time of diagnosis only 12% children are asymptomatic [3].

Pediatric RCC has four subtypes. Xp11(TFE) translocation RCC being the most common (20-40%) subtype which may occur following chemotherapy, followed by papillary RCC (30%) which may occur in the presence of preexisting tumours like Wilms tumour, metanephric adenoma or metanephric adenofibroma. Renal medullary carcinoma and oncocytic RCC are not much common subtypes and usually occurs in patients with the sickle cell gene and previously diagnosed with neuroblastoma respectively. Chromophobe RCC is very rare and thought to arise from the same cell type as renal oncocytomas.

It may appear in the setting of two genetic syndromes: Birt-Hogg-Dube syndrome associated with BHD gene or hereditary paraganglioma syndrome associated with SHD gene [2].

There is no definitive therapy for children with RCC. Surgery is curative if the tumor is localized and completely resected. In children with metastatic or residual RCC, chemotherapy as well as radiation therapy are not proved to be significantly effective [5].

RCC has a favourable long-term prognosis in children as compared to adults. Factors like symptomatic presentation, histology, stage, grade and performance status affect the prognosis. Tumour stage is the most important predictor of disease prognosis and helps to determine treatment protocols [6].

The low frequency of RCC in children suggests limited number of studies in this age group. However, it has distinct clinicopathological characteristics and exhibits significant differences from its adult analogue [7].

CONCLUSION

The role of pathologist is fundamental in the diagnosis of chromophobe RCC. Biopsy is useful in excluding other diagnoses and immuno - histochemistry has a vital role in it.

AUTHORS CONTRIBUTION

Syed Salman Ali: Entire research work, sample collection, analysis, literature review and write-up.

Rabia Ahmed: Concept and overall supervision.

Shoaib Naiyar Hashmi, Iqbal Muhammad, Saeed Afzal, Syed Naeem Raza Hamdani: Literature review**.**

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