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Adult's Wilms tumor: A case report

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A B S T R A C T

Nephroblastoma is the most frequent renal tumor in childhood population. Rarely, it can occur in adults. In this case, the diagnosis is frequently challenging for pathologists. No standard guidelines are available for this neoplasm in adults. It needs multidisciplinary collaboration for optimal management. Herein we report a 26-year-old man presenting with a non-metastatic right nephroblastoma. He underwent a radical nephrectomy. He is currently in complete remission after a follow-up of 18 months.

1. Introduction

Wilms Tumour or nephroblastoma is the most frequent renal tumor in paediatric population.[1] It is seldom present in adults. It is frequently diagnosed post operatively after radical or partial nephrectomy for suspicious renal mass. No standard management guidelines are available for adults population.[1] Diagnosis and treatment can be challenging for both clinicians and pathologists.

Herein we present a case of a 26-year-old patient who presented a nephroblastoma, treated by radical nephrectomy.

2. Case presentation

A twenty-six-year-old man was referred to the urology outpatient department for gross haematuria and right flank pain evolving for 12 months. No significant medical comorbidities were present. There was no significant family or psychosocial history. The patient denied any weight loss or anorexia. Clinical examination revealed right lumber contact. No palpable abdominal mass was found. No Varicocele was revealed. No clinical or biological paraneoplasic syndrome was found.

A thoraco-abdomino pelvic contrast-enhanced computed tomography was realised. It showed an enhancing upper polar right renal mass measuring 73 x 69 × 62 mm. It was tissular and well encapsulated. It was intimately blended to inferior surface of liver. There was a suspicion of adherence to the ipsilateral adrenal gland. No renal vein or inferior vena cava thrombus was found. Perirenal fat was infiltrated. Contro-lateral kidney was unremarkable. No distant metastases were found (Fig. 1).

The patient underwent right radical nephrectomy with adrenalectomy by right subcostal incision, without inconvenience. The post operative course was uneventful. The patient was discharged on the second post operative day (Fig. 2).

Histopathological examination of operative specimens showed tumoral biphasic proliferation patterns: The lesion is composed of undifferentiated blastemal cells with hyperchromatic nuclei, with frequent mitosis. Other areas display primitive epithelial differentiation with pseudo-rosettes formations. No heterologous component was found. Immunohistochemistry showed the expression of wT1 and CD56. Ki67 was present in 40%. Adrenal gland showed no tumoral infiltration. These findings concluded to the diagnosis of nephroblastoma (Fig. 3).

No local recurrence or metastatic progression were found during 18 months of post discharge follow-up.

3. Discussion

The most common primary malignant renal tumor in children is Wilms tumour (85% of renal tumors, approximately 5–6% of children neoplasm) with peak incidence between 3 and 4 years old.[1] It is also called nephroblastoma as it originates from the totipotent cells of the metanephrogenic blastemal remnants.[1] However, about 3% of Wilms Tumors are reported in adults, mostly diagnosed following nephrectomy for suspect renal mass.[1] It represents 0.5% of all renal neoplasms.[1]

Clinical presentation of adults with Wilms' tumor differs from that of children. The main symptom of adults is flank pain and the majority of them have a history of weight loss and a sudden drop in performans status, while children often report an abdominal indolent palpable mass.[2,3]

Metastatic stage disease within diagnosis is more common in adults

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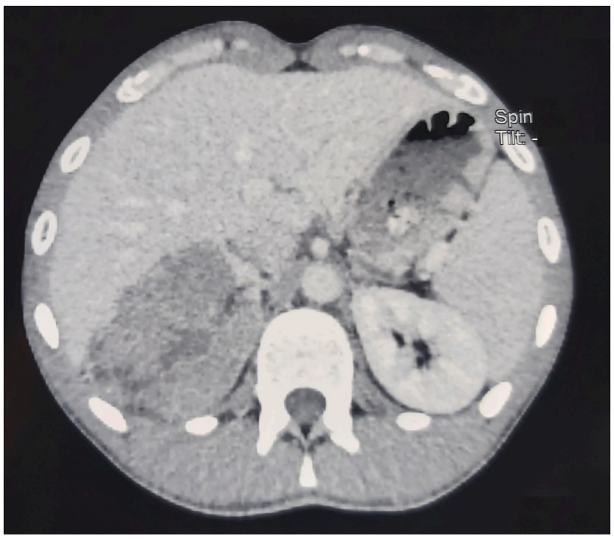

Fig. 1. CT showing a tissular well encapsulated right renal mass, with heterogenous enhancement.

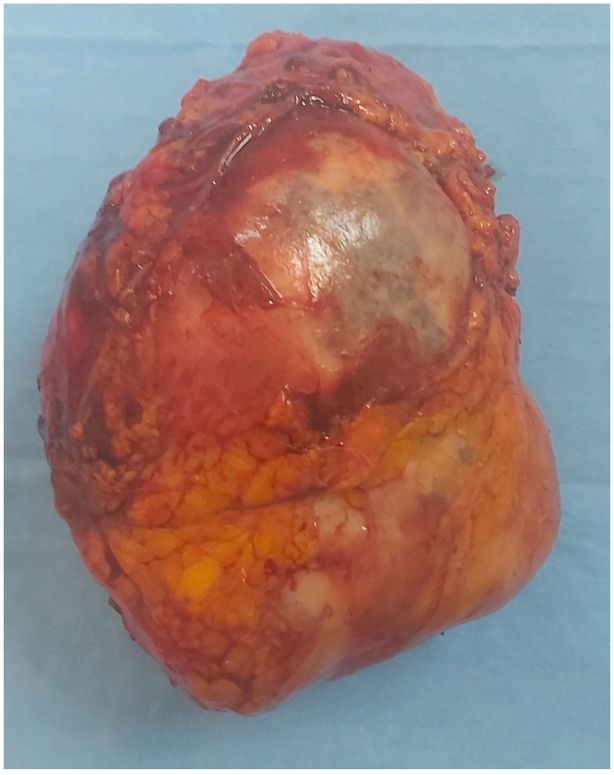

Fig. 2. Right enlarged nephrectomy specimen.

(10% in the pediatric population versus 30% in the adult population), and up to 50% of cases are in an advanced stage (III–V).[2] Distant metastasis of nephroblastoma usually occurs in lungs, liver and less frequently in bones, skin, bladder, large intestine, central nervous system, and the opposite kidney.[3]

There is no significant histological and radiological difference between adult and childhood Wilms' tumors. In fact, they share the classical triphasic histopathological features: Blastemal, epithelial and stromal features. The histological appearance is characterized by marked structural diversity, and the occurrence of all three types in the same case is uncommon.[3,4]

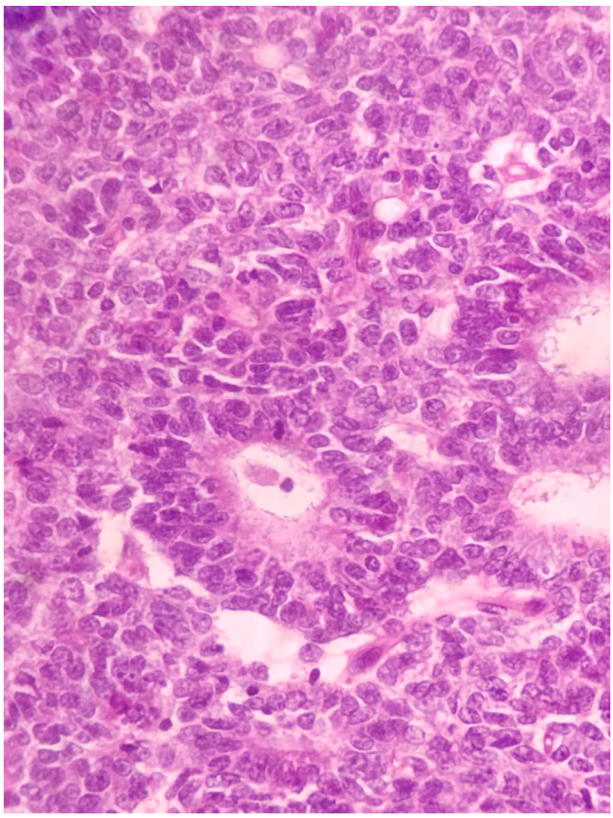

Fig. 3. Photomicrograph showing histopathologic features of biphasic proliferation of undifferentiated blastemal cells with hyperchromatic nuclei with areas displaying primitive epithelial differentiation with pseudo-rosettes formations.

Kilton and al. has established, in 1980, six pathological diagnosis criteria for nephroblastoma in adults, which are the following[3,4]: The tumour should be a primary renal neoplasm. Presence of primitive blastemic spindle or round cell component. Formation of abortive or embryonal tubules or glomerular structures. No area of tumour diagnostic of renal cell carcinoma. There should be pictorial confirmation of histology. Patient's age above 15 years.

Nephroblastoma is classified to one of three risk groups depending of the histopathologic features, which is necessary for therapeutic schemes.[3] Blastemal-predominant Wilms' Tumors are more aggressive than others types, with a poorer outcome (High risk group). Epithelial and stromal kinds represent intermediate risk tumors. Predominance of mesoblastic features put the tumor in low risk group.[3]

The standard paediatric treatment implies a multimodal approach with radical nephrectomy with lymphatic nodes dissection as the cornerstone of management, associated with exclusive chemotherapy or concomitant radiotherapy in most patients.[2] By comparison, due to the scarcity of cases counting only about 300 published papers, no clinical studies reporting a standard management guidelines are available for adult Wilms' tumor (1,2). In most isolated cases, the management is extrapolated from paediatric guidelines.[2]

Some authors suggested multiple factors that might explain the survival decrements in adults, including unfamiliarity of adult oncologists and pathologists with this diagnosis, and thus a potential delay in diagnosis and starting postsurgical therapy.[5]

Other factors are inadequate staging due to low rates of lymph node sampling, and underutilization of proven adjuvant chemotherapy and radiation.[5]

The treatment intensity, particularly the dose of vincristine, is often decreased in adults, as they have more competing health comorbidities and may develop more chemotherapy-related toxicity.

Improved outcomes in adults who were diagnosed timely and treated appropriately according to the pediatric protocols have however been reported.[5] Postoperative radiotherapy is frequently used in pediatric population presenting nephroblastoma. However due to the delay of histological diagnosis of nephroblastoma, radiotherapy is often abandoned, since it has to be debuted within 10 days after surgery. More time is usually required by the pathologist to confirm the histological diagnosis.[5]

It is obvious that management of this entity in adult population is challenging. For that reason, most papers recommend multidisciplinary collaboration between adult and paediatrician physicians.

4. Conclusion

The paucity of nephroblastoma in adult population makes its diagnosis and management challenging. A standardized model of care and management of nephroblastoma in this population, needs to be established. A closer collaboration between paediatric and adult oncology systems may increase mutual knowledge of the disease on one hand and treatment tolerability on the other hand.

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