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Case Report Open Access

Adult Nephroblastoma (Wilms Tumor): A Case Report

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Background and Rationale

Nephroblastoma (Wilms tumor) is the most common primary malignant renal tumor in children. Rarely, it may present in the adult age group. This is a case report of a diagnosed case of a 29-year-old female presenting with right-sided flank pain and hematuria. Abdominal sonogram revealed a right renal mass measuring 1.55 × 1.88 × 1.62 cm on the lower pole of the right kidney. CT scan showed a 2.1 × 1.9 × 1.9 cm solid mass on the lower pole of the right kidney. Partial nephrectomy was performed and a final diagnosis of adult Wilms tumor was made. Rarity of the tumor favours documentation in literature.

Case Presentation

This is a case of a 29-year-old female, who presented with haematuria and dull aching pain in the right flank. The general examination of the patient was unremarkable, with no lymphadenopathy or bony tenderness. The chest, cardiovascular and neurological examinations are unremarkable. Abdominal examination revealed a mildly tender mass palpable in the right lumbar area. Routine laboratory investigations including a complete blood count, chest X-ray, and renal function tests were normal. Urine examination showed numerous of red blood cells per high power field. An ultrasound examination was done, which showed an echogenic mass measuring 1.55 × 1.88 × 1.62 cm on lower pole of the right kidney. An abdominal CT scan was also done, which showed a 2.1 × 1.9 × 1.9 cm, solid mass on the lower pole of the right kidney. She was then advised to undergo a right partial nephrectomy.

Grossly, the partial nephrectomy specimen consists of a light brown, fairly ovoid, rubbery tissue measuring 3.2 × 3.0 × 2.0 cm. This is covered by a light gray, smooth, thin capsule on one side. Sections show a fairly circumscribed cream white, firm mass measuring 2.2 × 2.1 × 1.6 cm, which abuts the capsule and is 0.1 cm from the nearest margin of resection. The mass has a solid, finely granular cut surface, with no foci of haemorrhages or necrosis seen. The adjacent renal parenchyma appears normal. The renal capsule is intact.

Microscopically, the tumor is highly cellular comprising mostly of blastemic and epithelial elements. The diffusely proliferating blastemal cells are seen associated with immature tubular formations and immature glomeruli. The epithelial elements comprised of tubules are seen admixed with blastemic cells. The tubular pattern resembled pseudo-rosettes at places. The blastemic component has solid areas showing cells with oval nuclei and scanty cytoplasm. There is no area showing anaplasia. A final diagnosis of Adult Wilms tumor with biphasic pattern and a favourable histology, stage 1 is made (Figures 1 and 2).

Discussion

The most common kidney tumor in adults is renal cell carcinoma (RCC). Wilms tumor in subjects who are older than 16 years is rare. Only 3% of Wilms tumours are reported in adults, which explain the difficulties in diagnosis and treatment of this tumor entity in this age group [1]. In two hundred (200) cases reported in adults, it is unclear whether some of them may have been sarcomatoid renal carcinomas or some other renal neoplasm. Presence of abortive or embryonic glomero-tubular structures within an immature spindle cell stroma are diagnostic for Wilms tumor and are not found in renal cell carcinoma . Some renal cell carcinomas may have glandular elements with predominance of sarcomatous or undifferentiated cells, which are often misdiagnosed as Wilms tumours [2].

Most adults present with local flank pain and haematuria, and majority of them have a history of weight loss and sudden drop of performance status. In contrast to the palpable boggy mass, which is

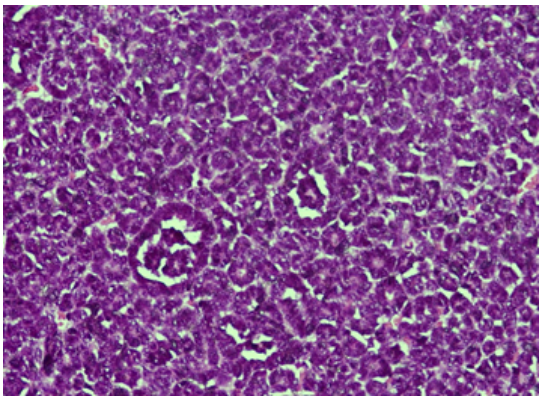

Figure 1: Blastema cells with embryonal glomerular component.

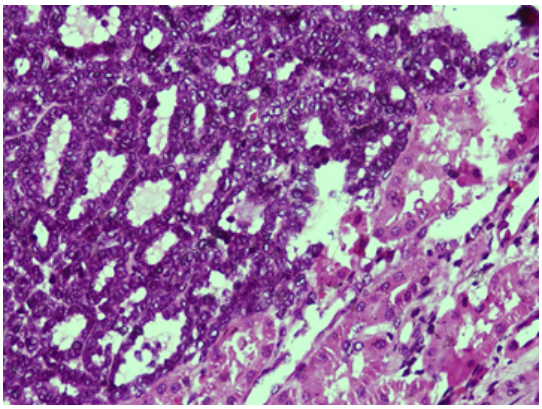

Figure 2: Epithelial elements composed of primitive tubular structures.

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more common in children [1]. In adults, Wilms tumor is larger and ill-defined with areas of necrosis and haemorrhage. In children, tumours are mainly asymptomatic, or children present with a painless swollen abdomen [1].

Radiographically, WT may present as in homogeneous masses with large areas of low density and less contrast enhancement than the normal parenchyma. Some are seen as complex, largely cystic mass, with some solid components in abdominal CT scans. The mass may contain calcifications. Sonograms of some patients with WT show venous thrombosis. Although invasion of renal vein and inferior vena cava are not used in staging, metastases and poorer prognosis were found in five of six patients with renal vein and/or inferior vena cava tumor thrombus. Angiographic findings in adults Wilms tumors were described as "poor vascularity with some neoformation of vessels, having a fine wavy or zigzag pattern [2]."

Adult Wilms tumor is diagnosed based on the criteria given by Kilton, Mathews, and Cohen [4]. These include 1) the tumor under consideration should be a primary renal neoplasm; 2) presence of primitive blastemic spindle or round cell component; 3) formation of abortive or embryonal tubules or glomerular structures; 4) no area of tumor diagnostic of renal cell carcinoma; 5) pictorial confirmation of histology and 6) patient's age >15 years. Kilton et al. reported 35 cases of adult Wilms' tumor complying with all the above criteria [3].

The differential diagnosis of an adult Wilms tumor with mainly epithelial differentiation includes metanephric adenoma. Metanephric adenoma is a rare benign kidney tumor found in childhood through adulthood, with 50% cases present with hematuria and abdominal pain. Grossly, metanephric adenoma appears unilateral, solitary, well circumscribed but unencapsulated. Microsections of a metanephric adenoma shows a pushing border with no capsule or infiltration into surrounding kidney parenchyma with proliferation of tumor cells with small, uniform round to oval nuclei with delicate chromatin, inconspicuous nucleoli and scant cytoplasm. On the other hand, a well-differentiated nephroblastoma would show a tumor capsule with distinct triphasic pattern with blastemal, stromal and epithelial components with tumor cells showing elongated or columnar nuclei with frequent mitotic activity. Immunostains may differentiate these two entities using a panel of IHC including CD56, CD57, WT-1, AMACR, CK7, and Cytokeratin AE1/3. Metanephric adenoma is usually CD56 negative, AMACR positive in 10%, WT-1 positive, Cytokeratin: AE1/3 positive in 50% but usually CK7. Well-differentiated nephroblastoma is usually positive for CD56 and CD57 with cells showing blastemal and epithelial differentiation stain positive for WT-1 [4].

A predominant blastemic Wilms tumor has a strong resemblance to small, blue round cell tumours, which commonly include lymphoma, peripheral neuroectodermal tumor and rhabdomyosarcoma; and rarely metastatic small cell tumours from lung, immature teratoma, and primary renal cell sarcoma. Extensive search for any other components is needed as a poorly differentiated renal carcinoma can have large sarcomatous areas resembling blastema [5]. Other differential diagnosis for nephroblastoma would include neuroblastoma, which is commonly found in the adrenal gland. Homer-Wright pseudo-rosettes often seen, and which stain for chromogranin, synaptophysin and NSE, negative for WT-1 in neuroblastoma [4].

Wilms tumor in adults has worse prognosis than in the paediatric population, a phenomenon for which there is no adequate explanation [3,6]. WT in adults, often present as large masses and are locally advanced at the time of diagnosis [7]. Even when comparing tumours with same stage, the prognosis still appears to be worse in adults than in children [8].

As adult Wilms tumor is rare, randomized trials cannot be performed. It has been suggested by most authors that to evaluate concepts for adequate treatment, results of randomized trials with childhood Wilms tumor should be extrapolated. National Wilms Tumor Study (NWTS) and other studies have recommended multimodal therapy for the disease with surgery, chemotherapy (actinomycin D, vincristine and doxorubicin) for 15 months and tumor bed irradiation in the case of stage 3 diseases. Less aggressive therapy with two drugs is advised in stage 1 and 2 diseases [5,6]. Satisfactory results have also been published with cisplatin and etoposide in patients with stage IV disease and patients in progression after conventional chemotherapy [6].

A study done by Arrigo et al for the National Wilms Tumor Study in 1990 reported on 27 adult patients with an event-free survival of 67% was achieved. This is in concordance with the paediatric population. They recommend treating risk-adapted adults depending on the tumor stage and histology [1]. The improvement of prognosis is due to central monitoring of patients and to standardized treatment according to the paediatric protocol [1]. The difficulties in correct diagnosis may lead to inappropriate or delayed treatment hence may be a contributory factor in poorer prognosis of adult Wilms tumor. The presence of RCC simultaneous with Wilms tumor does not necessarily worsen the prognosis. Localized RCC may be cured by surgical removal alone and may not need further treatment [1].

In addition to routine histo-pathologic examination, molecular and genetic studies such as aspiration cytology and cytogenetic analyses of Wilms Tumors may be necessary to gain further insight into the biology of individual tumors and may improve outcomes of aggressive therapy [7,9,10].

Conclusion

The rarity of Wilms tumor in adults warrants documentation. Wilms tumor should be considered in an adult patient presenting with pain in the flank and a renal mass. Prognosis is noted to be poorer than in adults than those of children when the disease is compared stage for stage. Yet with multimodal therapy, the outcome for adult patients diagnosed with Wilms tumor is steadily improving. In addition, molecular and genetic studies may be pursued to investigate into the biology of individual tumours hence improve outcomes of treatment.

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