Adult nephroblastoma

**N. Naqos, Z. Bouchbika, A. Taleb, N. Benchkroune, H. Jouhadi,
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ABSTRACT

Introduction: Nephroblastoma is rare in adults. It accounts only 0.5% of all renal tumors and it is often diagnosed in advanced stages. Histology, grading and staging are similar to pediatric nephroblastoma (PN) but its prognosis in the same clinical stage is more pejorative. However some reports use the current pediatric protocols to treat adult nephroblastoma (AN) taking into consideration tumor stage and grade. Others recommend using advanced disease regimens for all stages and grades and no standardized treatment is defined yet.

Case Report: A 29- year-old female who was diagnosed with stage IV nephroblastoma. She underwent radical nephrectomy and received multi-agent chemotherapy according to the International Society of Pediatric Oncology (SIOP) protocol No. 9301, a very important reduction of metastasis size occurred after three cycles.

Conclusion: Adult nephroblastoma is rare, its prognosis is poorer than that of children when the disease is compared stage for stage, but the outcome for adult patients having this tumor is steadily improving if it is early detected.

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CASE REPORT

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Keywords: Adult, Nephroblastoma, Pediatrics, Wilms tumor

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INTRODUCTION

Wilms tumor (WT) or nephroblastoma, is one of the most frequent malignant renal tumor in children [1], it is extremely rare in adults, its incidence is 8 per million in children [2], whereas its incidence is lower than 0.2 per million per year in adults, There is no difference between histological features of nephroblastoma in children and in adults, Treatment guidelines for pediatric nephroblastoma are well established but are less clear for adults [1–4], in view of the rare occurrence of this tumor in adults it is extremely difficult to carry out randomized clinical trials and therapeutic guidelines have not been defined yet.

Current therapies are based on trials and studies for children conducted by the Société Internationale d'Oncologie Pédiatrique (SIOP) and the Children's Oncology Group (COG) [3]. The prognosis for adult was reported to be worse than that for children [1, 2] because Adult nephroblastoma has more aggressive behavior and shows worse therapeutic response [2]. In children the overall survival is 90% for stage I and 70% for stage IV, this decrease with 76 % for stage I and with 11% for stage IV in adult [4].

We report a new case of adult nephroblastoma (AN) in which we discuss the diagnosis, treatment and outcome of this rare tumor for which the therapeutic strategies are more developed for pediatric groups and on which there are no large studies.

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CASE REPORT

A 29-year-old female was admitted to our department after 5 months history of left loin pain. Physical examination revealed hypertension and a palpable left loin mass. Biologically, the blood analysis were normal: hemoglobin: 12 g/dl, neutrophil: 3.4 g/l, leucocyte: 4.5 g/l, platelets: 340 g/l, creatinine: 58 mmol/l. Computed tomography (CT) scan of the abdomen showed a large left renal tumor with extension into renal vein and para-aortic lymphadenopathy with liver metastases, CT thorax confirmed multiple bilateral pulmonary lesions consistent with metastatic disease. A radical, transabdominal nephrectomy was performed. The operative specimen weighed 1540 g with a size of 13.5x13.5x8 cm surrounded by peritoneal grease with irregular outer surface. The kidney measures 19/17/15 cm and presents a white yellowish upper pole tumor of 13.5x13.5x8 cm encapsulated. It has a soft consistency with necrotic bleeding site estimated at 20%. The next of renal parenchyma measures 5x6.5x3 cm (Figure 1). Histology confirmed a diffuse blastemal and epithelial Wilms tumour, it shows tumor proliferation with dual component: epithelial and mesenchymatous; the epithelial component makes glandular and trabecular structure bored with cylindrical basophile cells with big size, with hyperchromic nucleus and atypical mitosis. The epithelial structure is furrowed with fusiform cells big nucleus size, all are developed in a stroma surrounded by a fibrous tick capsule respecting the adjacent parenchyma. No image of vascular embolism was seen, the renal hilum was unaffected as well as the ureteral cuts and the adrenal (Figure 2). The lymph node dissection was negative 0N/6N. There were no postoperative complications.

Chemotherapy according to the SIOP 93-01 protocol was started as soon as possible. After 2 cycles of chemotherapy combining actinomycine D at 15 mg per kg and per injection/3 weeks, adriamycin with the dose of 50 mg/m² each 3 weeks and Vincristine 1.5 mg/m² d1-d8, a very good clinical response was seen and a marked reduction in metastases size occurred (Figures 3 and 4). After six cycles of chemotherapy a complete remission of liver metastases was obtained with a very good reduction of lung metastases, unfortunately two months later the patient died after a sudden crisis headache.

DISCUSSION

Nephroblastoma is rare in adults and could have a more aggressive clinical behaviour than that in children, it is frequently diagnosed at more advanced stages, stage III disease represents 50% of AN.

Clinically, the tumor is often discovered as an incidental abdominal mass either by routine palpation or on a CT-scan or ultrasound done for other reasons. More commonly however, flank pain and/or hematuria are the first indications of the condition. Systemic symptoms such as weight loss, malaise, weakness, and fever are

Figure 1: The operative specimen showing the kidney 19x17x15 cm with a white yellowish upper pole tumor of 13.5x13.5x8 cm encapsulated.

Figure 2: Tumor proliferation with blastema, glomerular and tubular formations, and mesenchyme (Gx250).

indicative of advanced disease [5, 6]. The most common metastatic sites are the lungs, liver and the bones [3]. The classic histology of nephroblastoma is that of a triphasic embryonal neoplasm containing varying amounts of blastema, stroma and epithelial cells forming abortive tubular or glomerular structures. For diagnosis to be made not all the classic features are necessary, as some Wilms tumors may contain blastemal or epithelial features only [7]. According to all available studies and due to the very small number of reported cases, randomized trials cannot be undertaken and results of pediatric trials should be considered. And Until recently, no standardized treatment for AN was found in literature [3, 8]. There

Figure 3: Computed tomography pulmonary scan showing the pulmonary metastases before (A) and after (B) 3 cycles of chemotherapy.

were only a few reports of small series of patients with AN who were treated with different protocols involving chemotherapy, surgery, and radiotherapy [8]. The results of these early studies are somewhat contradictory. Fortunately, the more current guidelines proposed by the National Wilms Tumor Study Group (NWTSG) and The International Society of Pediatric Oncology (SIOP) have helped to direct physicians [8].

National Wilms Tumor Study Group (NWTSG) realises nephrectomy at the time of diagnosis for all resectable primary tumors. Surgery is then followed by chemotherapy and radiation therapy to sites of metastatic or residual disease after surgery [9]. In contrast, The International Society of Pediatric Oncology (SIOP) recommends administering several weeks of chemotherapy before nephrectomy [10].

Figure 4: Computed tomography abdominal scan showing the liver metastases before (A) and after (B) 3 cycles of chemotherapy.

In 1982, the NWTSG reported 31 patients with AN who were treated between 1968 and 1979 [8, 6]. 51.7% had stage III and stage IV disease. The three-year survival rate was 24%, compared with 74% for patients with pediatric nephroblastoma (PN). These results indicated a need for new therapeutic approaches for patients with AN. In 1990, Arrigo et al. [8, 11] from the NWTSG studied 27 adult patients who were treated between 1979 and 1987. Their results demonstrated a survival rate of 67% at three year. Moreover, the national Wilms Tumor Study (NWTSG) recommends a multidisciplinary treatment: for stages III and IV, a large nephrectomy associated with a triple Chemotherapy (actinomycine D, vincristine and doxorubicin) during 15 months then a radiation of tumor bed and a double chemotherapy without radiation for both stage I and II [12].

For our patient we followed the NWTSG's recommendations, after surgery a combination of vincristine, dactinomycin and doxorubicin was used, after three cycles a remarkable reduction of about 80% of lever and lung metastases was obtained.

In the study of the International Society of Pediatric Oncology [3], 30 AN were treated: 66% of them were localized whereas 33% were metastatic at the time of diagnosis. Unlike protocols for children, the majority of adult in SIOP study underwent primary surgery; whereas preoperative chemotherapy was administered to only 4 out of 30 patients. The event-free survival was 57% at 4 years and the overall survival was 83% [3].

Terenziani et al. [13] reported their institutional experience regarding AN. Between 1983 and 2001, 17 patients with AN who were older than 16 years were treated according to pediatric nephroblastoma (PN) guidelines. In this series, the overall survival was 62.4% at 5th year.

Due to the frequency of lung metastases in stage III and IV, Hentrich et al. justify a systemic pulmonary irradiation of lung metastases they consider that an irradiation of bed tumor may complete surgery following by chemotherapy (Dactinomycin + Vincristine + Doxorubicin). Lately, in relapsed AN patients, high-dose chemotherapy followed by autologous stem cell rescue has been used as the salvage therapy but only a small number of patients have obtained long complete remission [1, 14]. The use of the new combination chemotherapy regimens based on the SIOP-2001 second-line protocol has not improved the outcome in patients with recurrent AN. The best approach in the treatment of AN still needs to be defined. Abu-Ghosh et al. [15] reported an overall response rate of 82% in 11 patients treated with ifosfamide-carboplatin etoposide chemotherapy for poor-risk relapsed nephroblastoma. Italiano [16] reported one case of a short but striking response to paclitaxel in an adult who had recurrent massive disease. Our patient presented with advanced disease. His clinical follow-up suggests that treating AN like childhood Wilms tumor remains an effective option. New modalities of treatment are being tested. One example is the use of recombinant interferon- α to treat a case of recurrent adult nephroblastoma [17]. Whether these new modalities of treatment will prove to be more effective, they await additional data, hopefully from standardized clinical studies on adults.

CONCLUSION

Wilms tumor or nephroblastoma is an exceptional tumor in adults. This diagnosis, often histological, is generally established at a more advanced clinical stage than in children and the prognosis, for the same clinical stage, is comparatively poorer, it must be kept in mind that only the early detection of the tumor offers patients the best chance for survival.

Author Contributions

N. Naqos – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation

of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Z. Bouchbika – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

A. Taleb – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

N. Benchkroune – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

H. Jouhadi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

N. Tawfiq – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

S. Sahraoui – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

A. Benider – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

1. Zdrojowy R, Sawicz-Birkowska K, Apoznanski W, et al. Adult Wilms tumour. *Int Urol Nephrol* 2011 Sep;43(3):691–6.
2. Vehbi Ercolak, Ismail Oguz Kara. A case report of adult Wilms' tumour, *ContempOncol (Pozn)* 2014;18.
3. Reinhard H, Aliani S, Ruebe C, Stockle M, Leuschner I, Graf N. Wilms' tumor in adults: results of the Society of Pediatric Oncology (SIOP) 93-01/Society for Pediatric Oncology and Hematology (GPOH) Study. *J Clin Oncol* 2004 Nov 15;22(22):4500–6.
4. Tahri A, Benchechroun N, Karkouri M, et al. [Nephroblastoma in adults. Three case reports]. [Article in French]. *Ann Urol (Paris)* 2001 Sep;35(5):257–61.
5. Dawson NA, Klein MA, Taylor HG. Salvage therapy in metastatic adult Wilms' tumor. *Cancer* 1988 Sep 1;62(5):1017–21.

6. Fosså S. Rare and unusual tumors of the genitourinary tract. *Curr Opin Oncol* 1992 Jun;4(3):463–8.
7. Huser J, Grignon DJ, Ro JY, Ayala AG, Shannon RL, Papadopoulos NJ. Adult Wilms' tumor: a clinicopathologic study of 11 cases. *Mod Pathol* 1990 May;3(3):321–6.
8. Arifi S, Mellas N, Kaikani W, et al. Adult Wilms' Tumor: Report of 2 Cases and Review of the Literature. *UroToday International Journal* 2010;3:1944–5784.
9. Grundy P, Green D, Coppes M. (2002) Renaltumors. In: Pizzo PA, Poplack DG (eds) *Principles and practice of pediatric oncology*. Lippincott Williams & Wilkins, Philadelphia.
10. Tournade MF, Com-Nougué C, de Kraker J, et al. Optimal duration of preoperative therapy in unilateral and nonmetastatic Wilms' tumor in children older than 6 months: results of the Ninth International Society of Pediatric Oncology Wilms' Tumor Trial and Study. *J Clin Oncol* 2001 Jan 15;19(2):488–500.
11. Arrigo S, Beckwith JB, Sharples K, D'Angio G, Haase G. Better survival after combined modality care for adults with Wilms' tumor. A report from the National Wilms' Tumor Study. *Cancer* 1990 Sep 1;66(5):827–30.
12. Kalapurakal JA, Nan B, Norkool P, et al. Treatment outcomes in adults with favorable histologic type Wilms tumor—an update from the National Wilms Tumor Study Group. *Int J Radiat Oncol Biol Phys* 2004 Dec 1;60(5):1379–84.
13. Terenziani M, Spreafico F, Collini P, et al. Adult Wilms' tumor: A monoinstitutional experience and a review of the literature. *Cancer* 2004 Jul 15;101(2):289–93.
14. Petty R, Culligan D, Bissett D. Successful salvage of relapsed adults Wilms' tumour with high dose chemotherapy and autologous peripheral blood stem cell rescue. *Clin Oncol (R Coll Radiol)* 2002 Aug;14(4):329.
15. Abu-Ghosh AM, Krailo MD, Goldman SC, et al. Ifosfamide, carboplatin and etoposide in children with poor-risk relapsed Wilms' tumor: a Children's Cancer Group report. *Ann Oncol* 2002 Mar;13(3):460–9.
16. Italiano A, Sirvent N, Michiels JF, Peyrade F, Otto J, Thyss A. Tumour response to paclitaxel in an adult with relapsed nephroblastoma. *Lancet Oncol* 2005 Apr;6(4):252–3.
17. Tawil A, Cox JN, Roth AD, Briner J, Droz JP, Remadi S. Wilms' tumor in the adult—report of a case and review of the literature. *Pathol Res Pract* 1999;195(2):105–11.

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