Metastatic Ewing’s sarcoma/PNET of kidney in 40 year old patient

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ABSTRACT

INTRODUCTION: Primary renal Ewing sarcoma/PNET is an uncommon and very aggressive tumor. We report the case of a young woman who underwent nephrectomy for a renal mass from unknown etiology. Histologic analysis found small tumoral cells in rosette formation, and immunohistochemical staining was positive for CD99, and focally positive for vimentin and Protein S-100. A post operative abdominal computed tomography (CT) scan revealed a residual renal processus with hepatic wounds and abdominal metastatic nodes.

After 6 cycles of chemotherapy including Vincristine, Doxorubicin, Cyclophosphamide, the response was considered as good.

DISCUSSION: Ewing’s sarcoma/PNET of kidney is a member of the family of small round cell tumors and it should be differentiated from Wilms tumor, neuroblastoma, rhabdomyosarcoma and lymphoblastic lymphoma. The principle management of its treatment have been extrapolated from the treatment of osseous Ewing sarcoma of bone.

CONCLUSION: Despite aggressive treatment, primary renal Ewing sarcoma/PNET has a poor prognosis. It requires a multidisciplinary approach including oncologists, urologists and radiation oncologists.

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1. Introduction

The extra-osseous Ewing’s sarcoma represents nearly 6% of the Ewing’s sarcoma family of tumors (ESFT). It was introduced initially in 1969 by Tefft who reported a series of five patients with round cell tumors that come from the paravertebral soft tissues. It often involves the trunk, the extremities, the soft tissues of head and neck, and the retroperitoneum. However, renal localization is very rare and has a poor prognosis. Because of the rarity of this disease, there is no consensus concerning the treatment of renal Ewing’s sarcoma/PNET.

We report the case of a young female patient with a renal metastatic Ewing’s sarcoma/PNET which was successfully managed by chemotherapy.

2. Case report

A 40 years old woman was referred to the National Institute of Oncology with severe back pain one month after a right nephrectomy, performed because of a renal mass, in a peripheral hospital. Only abdominal tenderness was found on clinical exam. There was no organomegaly and no hematuria. Histological report described macroscopically a renal mass about a renal mass measuring 190 mm × 100 mm × 90 mm, white gray, friable, with areas of necrosis and hemorrhage. Microscopic examination showed a malignant tumor composed of monomorphic cells, with slightly irregular nuclei and dense chromatin. These cells fitted together in some places forming rosettes (Fig. 1). At immunohistochemistry, the cells were positive for the PNET markers: CD99 and focally positive for Vimentin antacors and protein S-100, but negative for Epithelial membrane antigen (EMA), Leucocyte common antigen (LCA) and Desmine (Fig. 2).

A post-operative abdominal computed tomography (CT) scan revealed a residual mass in the right renal space which involved liver. The mass abutted onto the duodenum, the right psoas muscle and the right colic flexure. Peritoneal nodes and ascites were also found. The computed tomography of the thorax, the bone scintigraphy, the blood electrolytes and the renal function were all normal. The disease was staged at IV.

Our patient was treated by six cycles of chemotherapy every three weeks including Vincristin 1.4 mg/m2, Adriamycine 60 mg/m2, and Cyclophosphamide 1250 mg/m2. The tolerance of chemotherapy was good. The response was estimated at 90% according to the Recist guideline version 1.1. The patient was in good control for four months after the last cycle of chemotherapy.

3. Discussion

Primary renal Primitive Neuroectodermal Tumor (PNET) is a very rare condition. The first case was reported by Seemayer et al. Since then, there have been about only 60 cases reported in the

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There is no consensus of treatment for metastatic renal PNET because of its rarity. The treatment is extrapolated from the treatment of osseous Ewing sarcoma. It is based on combination of chemotherapy included active drugs such as Doxorubicin (A), Vin-cristin (V), Cyclophosphamide (C), Ifosfamide (I) and Etoposide (E).

The IESS-III study published in 2003 by Grier conducted in the pediatric oncology group children’s cancer group (POG-CCG) study (INT-0091), concluded that patients with non metastatic Ewing sarcoma were randomized to receive chemotherapy with either VACD alone or alternating with ifosfamide and etoposide (VACD-IE) for a total of 17 cycles. The five event free survival rate was 69% in the VACD-IE group as compared with 54% in the VACD alone group. Overall survival was also significantly better among patients in the VACD-IE group (72% vs 61% in the VACD group). However, the addition alternating cycles of Ifosfamide (I) and etoposide (E) to the VDC (vincristin, D–actinomycin, cyclophosphamide) regimen did not improve the outcome of the patients with metastases.

Otherwise, the EURO–EWING study is designed to evaluate the efficacy and safety of multiagent induction chemotherapy with six courses of vincristine, ifosfamide, doxorubicin, and etoposide (VIDE), local treatment (surgery and/or radiotherapy), and high dose therapy followed by stem cell transplantation in 281 patients with Ewing’s sarcoma with primary disseminated disease. The event free survival and overall survival rates at 3 years for the entire study cohort were 27% and 34% respectively. The event free survival rates were 57% and 25% respectively for patients with complete and partial response after HDT/SCT. The relevant risk factors are the patient’s age, tumor volume, and extent of metastatic spread.

About 30–40% of patients with Ewing’s sarcoma experience recurrence. The only prognostic factor identified in relapse seems to be time to relapse: patients relapsing later than 2 years from initial diagnosis have a better outcome. The treatment option for patient with relapsed or refractory disease participation in clinical trial. The guidelines proposed the following regimen: Ifosfamide with etoposide and carboplatin, Ifosfamide and etoposide, docetaxel in combination with gemcitabine, Temozolomide and irinotecan.

Despite aggressive treatment, the prognosis of rPNET is poor. The median survival for advanced disease is only 2 years. In the retrospective study reported by Yuvaraja (16 patients with renal PNET/Ewing sarcoma): 5 patients (31%) had metastases (2 lungs, 1 lung and lymph nodes, 1 lymph nodes, and 1 liver). The median survival was estimated at 15 months.

In the case we report, the age of occurrence of rPNET at 40 years old which is unusual. The diagnosis of PNET/Ewing’s sarcoma of the kidney was established on morphologic and immunohistochemical criteria. We could not perform molecular analysis, because of a lack of technical structures.

We treat our patient with 6 cycles of chemotherapy every three weeks including: Vincristin 1.4 mg/m², Adriamycin 60 mg/m² and Cyclophosphamide 1250 mg/m². The response was considered as good, and the control was kept for four months. The patient died 4 months later in the unknown circumstances.

4. Conclusion

Because of its rarity, the management of PNET/Ewing sarcoma of the kidney remains difficult, and the results are uncertain. The prognosis is poor, despite aggressive treatment. More experiences are needed to clarify the management strategy.

Conflict of interest statement

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Ethical approval

Written informed consent was obtained from the patient of the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor in chief of this journal of request.

Author contributions

Mouna Kairouani participated in the care of the patient and wrote the article. Saber Boutayeb participated in the writing of article and validated the content. Maha MOKRIM and Nawfal Mellas participated in the care of the patient. Hind El M’rabi and Hassan Errihani: validated content and form of the article all authors read and approved the final manuscript.

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