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Clinical case

Ewing's sarcoma of bladder: about a Moroccan case treated in the medical oncology department of FEZ

Sarcome de la vessie d'Ewing : à propos d'un cas traité dans le service d'oncologie médicale de Fès

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Abstract

The Ewing's sarcoma of bladder is a rare entity. To our knowledge, only 16 cases of Ewing's sarcoma have been documented in the literature. The management of this type of tumor in this localization is not well identified, and it is similar to the management of PNET / EWING tumors of the limbs. We reported the case of patient aged of 31 years old who has presented a dysuria and a macroscopic hematuria. the cystoscopy showed a large tumoral process on the anterior surface and on the dome of bladder. the biopsy founded the round cell tumor proliferation. the immunohistochemical and the translocation research t(11,22) confirmed the PNET tumor. the radiological assessment showed the Ewing sarcoma locally advanced of bladder. The patient has been treated by palliative chemotherapy (VAC- IE protocol). Our paper reports our case by performing a review of the literature of published cases.

Keywords: Ewing sarcoma, bladder tumor, chemotherapy.

Résumé

Le sarcome de la vessie d'Ewing est une entité rare. A notre connaissance, seuls 16 cas de sarcome d'Ewing ont été documentés dans la littérature. La gestion de ce type de tumeur à cette localisation n'est pas bien identifiée et calquée à la gestion des tumeurs des membres PNET / EWING. Nous rapportons le cas d'un patient de 31 ans ayant présenté une dysurie et une hématurie macroscopique. La cystoscopie a montré une grosse tumeur sur la surface antérieure de la vessie et sur le dôme. La biopsie a identifié la prolifération des tumeurs à cellules rondes. La recherche immunohistochimique et la translocation t (11,22) ont confirmé la tumeur PNET. L'évaluation radiologique a montré un sarcome d'Ewing localement avancé en dépend de la vessie. Le patient a été traité par chimiothérapie palliative (protocole VAC-IE). Notre papier rapporte notre cas en effectuant une revue de la littérature des cas publiés.

Introduction

Ewing sarcoma (ES) develop in the bones or nearby soft tissues. Three main types of ES have been described: ES of the bone, extra-osseous (extra skeletal) ES and primitive neuroectodermal tumor (PNET). He is the second most common primary

bone tumor seen in children and adolescents. However, extraosseous sarcomas of the Ewing family in adults are very rare. ES involving the urinary bladder is rare, with only 16 cases reported in the literature to our knowledge.

Clinical case

We report the case of a patient aged 31 years old, alcoholic smoking addict for 13 years, who has presented for 5 months dysuria. The symptomatology was aggravated by the appearance of macroscopic hematuria. The patient consulted in the urology department, the cystoscopy showed a large tumoral process on the anterior surface and on the dome of bladder measuring 8 cm.

The biopsy founded the round cell tumor proliferation. The immunohistochemical complement showed the tumor cells expresses of CD99, lack of expression of myogenic, Melan A, cytokeratin and CD 45 which is in favor of a neuroectodermal tumor in vesical localization. The translocation research t(11,22) confirmed the PNET tumor (figure 1)

A thoraco abdominopelvic CT scan showed a bulky tumoral process centered on the bladder measuring 21 * 17 * 10 cm, it comes into contact with the anterior abdominal wall without separating lip, it infiltrates the sigmoid and the prostate and it pushes back the iliac vessels external outside (figure 2)

The tumor is unresectable. The patient has been transferred to oncology department. At admission the patient presented macroscopic hematuria. The physical examination showed a patient with index of karnofsky between 60 et 80, a huge abdominopelvic mass measuring 20 cm. The decision was to start the chemotherapy with the protocol VAC-IE (Vincristine+ Doxorubicin+ Cyclophosphamide + Mesna/ Etoposide, Ifosfamide + Mesna)

The patient has received 3 cures with disappearance of macroscopic hematuria and abdominal pain and stabilization of tumoral mass. After 6 cures, the patient presented a clinical and radiological progression. The decision was to start a palliative care, the patient died a 1 month later.

Table 1: Reported cases of urinary bladder Ewing's sarcoma

Case reported	sex	age	presentation	treatment	Survival
1Banerjee et al., 1997	M	21	Microscopic hematuria, dysuria	Cystectomy then Vincristine-adriamycin-cyclophosphamide	18 months
2.Gousse et al., 1997	F	15	Macroscopic hematuria	Trans Urethral Resection + partial cyclophosphamide etoposide	12 months
3.Desai, 1998	F	38	Gross hematuria	Cystectomy+ total hysterectomy +bilateral salpingo-oophorectomy;	-

4.Lopez Beltran et al., 2006	F	21	Gross hematuria, dysuria, frequency	Radical Cystectomy+ total hysterectomy+ bilateral salpingo-oophorectomy+ Adjuvant CTx (unknown-1 year) + maintenance with imatinib	36 months
5.Osone et al., 2007	M	10	Gross hematuria, dysuria	Trans Urethral Resection then Cyclophosphamide-+pirarubicin vincristine(1,2,3,6cycles)+ifosfamide-etoposide (4,5,7 cycles)	24 months
6.Al Meshaan et al., 2009	F	67	Repeated hematuria	Trans Urethral Resection + partial cystectomy+ 3 courses chemotherapy for prior bladder tm (not for PNET)	8 months
7.Rao et al., 2010	F	14	Lump in lower abdomen	Sleeve resection of bladder and 3 cycles undefined CTx after recurrent	6 months
8. Busato et al., 2011	F	52	Pelvic pain, dysuria, frequency	Trans Urethral Resection then Doxorubicin-vincristine-cyclophosphamide dactinomycin /etoposide/ifosfamide	27 months +
9.Okada et al., 2011	M	65	Gross hematuria, dysuria	Trans Urethral Resection and Vincristine, ifosfamide, doxorubisin, etoposide(for lymph node recurrence);	7 months
10.Sueyoshi et al., 2014	M	10	Polyuria, lower abdominal swelling	3 courses neoadjuvant chemotherapy then Partial cystectomy and 4 courses adjuvant vincristine, doxorubicin, cyclophosphamide+ ifosfamide, etoposide	11 months
11. Şenol Tonyalı et al, 2016	F	38	Gross hematuria	Radical cystectomy, and Vincristine +doxorubicin+cyclophosphamide+ mesna/etoposide, ifosfamide + mesna	14 months
12.Mentzel et al., 1998	M	62	Fever, acute urinary retention, backache	Nephrostomy	Death
13.Colecchia et al., 2002	F	61	Bilateral hydronephrosis	-	-
14.Kruger et al., 2003	M	81	Lower extremities Lymphedema, fatigue, urge incontinence	Trans Urethral Resection + BilateraL	2weeks

15.Ellinger et al., 2006	M	72	Gross hematuria, oliguria	bilateral pelvic lymphadenectomy, ileum resection	2 months
16.Zheng et al., 2011	M	74	Frequency, dysuria, hematuria	Palliative surgery + 3 courses epirubicin, vincristine, cyclophosphamide	4 months
17 Current case	M	31	Dysuria, gross hematuria	The palliative chemotherapy with Vincristine+ doxorubicin+cyclophosphamide+mesna/etoposide, ifosfamide +mesna	12 months

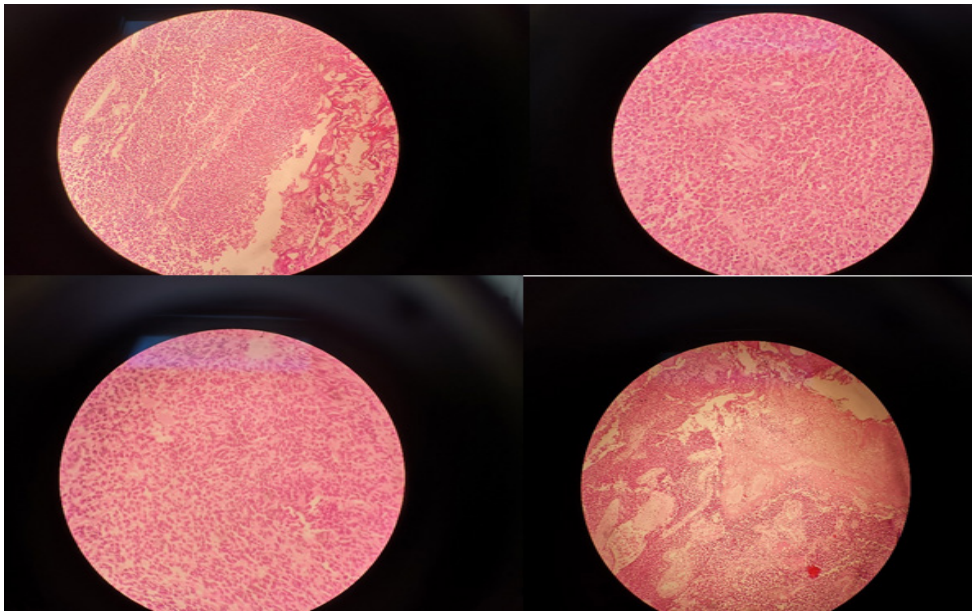

Figure 1: The round cell tumor proliferation with expression of CD99 and lack of expression of myogenic, Melan A, cytokeratin and CD 45 which is in favor of a neuroectodermal tumor in vesical localization

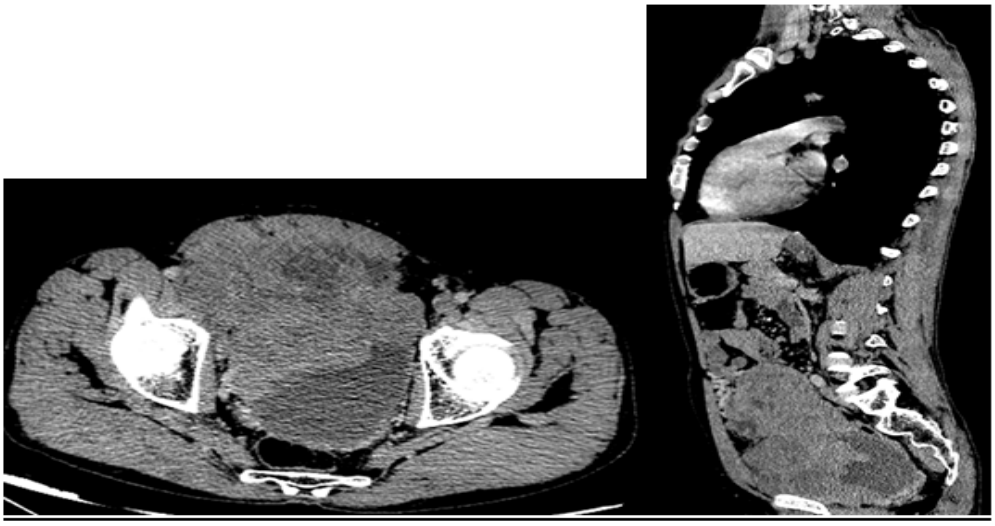

Figure 2: locally advanced ewing sarcoma located on the bladder and extending to neighboring organs.

Discussion

Ewing's sarcoma/Primitive neuroectodermal tumor is rare entity in bladder localization. It has not been possible to establish definitive guidelines regarding its management and treatment due to the very small number of reported cases; to our knowledge only 16 cases have been documented in the literature to day. Review of those previous reports including the present case is shown in (Table 1) (1-7). There was no gender tendency, with eight female and eight male patients, with a mean age of 43.81 years old (range: 10-81). Hematuria was the most frequent presenting symptom. Some patients presented with bilateral hydronephrosis, renal failure and edema in lower extremities, which were associated with advanced disease. The diagnosis of primary ewing sarcoma in the urinary bladder is difficult. Its differential diagnosis includes other small round cell malignancies, particularly small cell neuroendocrine carcinoma, embryonal rhabdomyosarcoma, small cell variant of malignant melanoma, non-Hodgkin lymphoma, synovial sarcoma, and desmoplastic small round cell tumor. These diagnoses can be ruled out in most cases if a relevant immunohistochemical panel is applied. Molecular analysis supports the final decision of ewing sarcoma /PNET by showing the EWS gene rearrangement via FISH, and the reciprocal translocation t (11; 22).

The radiologic imagery showed five metastatic cases at the time of diagnosis and 2 tumors had perivesical infiltration.

The radical surgical treatment was performed in 8 cases (total cystectomy in 4, partial cystectomy in 4) One patient with a frozen pelvis was inoperable. The remaining patients were managed with transurethral resection or conservatively. The use of adjuvant or neoadjuvant chemotherapy were in 8 cases. The palliative chemotherapy was used in 2 cases.

The recommended chemotherapy regimen consists of vincristine, doxorubicin and cyclophosphamide, alternating with ifosfamide and etoposide. (8,9)

The prognosis for recurrent, locally advanced or metastatic disease is reserved. patients who received adjuvant or neoadjuvant chemotherapy had the best survival compared to patients who received surgical treatment only without chemotherapy. (10).

Conclusion

Ewing's sarcoma/Primitive neuroectodermal tumor (ES/PNET) can also rarely occur in visceral organs such as urinary bladder. Treatment includes radical surgical treatment with chemotherapy in the localized stage. Palliative chemotherapy remains the only means of treatment in the metastatic and locally advanced stages. The chemotherapy protocols are constituted by the drugs used in the limb ewing sarcomas. The prognosis of this disease remains very reserved.

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Conflict of interest: none

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