

PRIMITIVE NEUROECTODERMAL TUMOR (PNET) OF THE PROSTATE: CASE REPORTEL. Mouhtadi S.,*¹ Filali N.,¹ Abahssain H.,² Harrak S.,¹ Mrabti H.,³ Boutayeb S.³ and Errihani H.⁴¹Resident in Medical Oncology, National Oncology Institute of Rabat Morocco.²Doctor Specialist Medical Oncology Department, National Oncology Institute of Rabat Morocco.³Professor of Medical Oncology Department, National Oncology Institute of Rabat Morocco.⁴Head of Medical Oncology Department, National Oncology Institute of Rabat Morocco.***Corresponding Author: EL. Mouhtadi S.**

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INTRODUCTION

Primitive neuroectodermal tumor (PNET) is a malignant tumour of the neural crest which is now included in the PNET/EWING family. Showing to share the same characteristic cytogenetic overlaps and a t-translocation.^[11,22] (q24 ; q12).^[1]

It mainly affects soft tissues and bones, rarely visceral sites such as the brain, parotid gland, lung, kidney, urinary bladder, uterus, pancreas and more rarely the prostate.^[2,3]

The treatment is a combination of chemotherapy, radiotherapy and surgery, but with a poor results with high mortality.

In this study we present an extremely rare case of prostate PNET with a brief review of the literature.

CASE REPORT

Mr E.E. 32 years old, from Morocco, chronic smoker.

The patient presented in urology consultation in October 2019 with six months history of nocturnal pollakiuria and dysuria progressively evolving towards urine retention, pelvic discomfort, weight loss and a deterioration of general condition.

Rectal examination, the prostate was very large and the mass was hard and palpable with poor mobility, the rest of the clinical examination shows left inguinal adenopathy measuring 5cm, compressing the left lower limb with oedema without associated deep vein thrombosis.

The thoraco-abdomino-pelvic CT scan showed a voluminous process of the prostatic lodge measuring 15 cm and extending over 20 cm in height, containing large areas of necrosis, invading the lower bladder floor, the left ureter, the left external iliac vein and the pelvis by contiguity. Without metastasis sites, especially lung and bone.

The initial biological assessment had objectified renal insufficiency, a bilateral nephrostomy was carried out with normalisation of the renal function, no other biological abnormalities were noted. The PSA level was normal at 0.8 ng/ml.

The initial biological assessment showed a renal failure, after a bilateral nephrostomy the renal function was corrected, no other biological disorders were noted. The PSA level was normal at 0.8 ng/ml.

Prostate biopsies revealed a poorly differentiated malignant tumour process infiltrating the prostate parenchyma.

The immunohistochemical study confirmed that it was a round cell tumour proliferation expressing CD99, and not expressing CD56, synaptophysin, anti-chromogranin antibody, desmine and myogenin, all in favour of a sarcoma of the PNET/EWING family.

The decision of the multidisciplinary consultation meeting of the national oncology institute Rabat in November 2019, was a chemotherapy based on Vincristine, Doxorubicin, Cyclophosphamide, Etoposide and Ifosfamide according to the CAV/IE regimen, followed by discussion of a local treatment depending on the response.

During his hospitalisation for his first chemotherapy treatment, the patient presented a high fever at 39°C. The infectious assessment was in favour of a urinary infection, the antibiotic therapy showed a good evolution.

The patient received 6 cycles of chemotherapy with good clinical and biological tolerance, and a significant improvement in quality of life.

Until June in 2020 when the succeeding six rounds of chemotherapy were finished. CT scan showed the mass shrank to 8,2 cm, and then full-round external RTx was added afterwards.

A three-dimensional conformal radiation therapy was given at a dose of 59,4 Gy in (25 + 8) sessions.

Until the 25th session the tolerance is globally good without signs of toxicity.

DISCUSSION

Primary neuroectodermal tumours PNET of the prostate are aggressive tumours with a very poor prognosis.^[4]

The PNET diagnosis of prostate cancer is full of challenges.

They are extremely rare, representing less than 0.1% of prostate cancers in adults,^[5] particularly in young men between 35 and 60 years old. Only about ten cases are reported in the literature.^[6]

The age of PNET patients ranged from 20 to 49, which was much younger than the median age for prostate cancer. The median overall survival of these patients was 11 months.

Clinically, almost the same urinary symptomatology is described (pollakiuria, dysuria, haematuria, urine retention, pelvic pain and constipation).^[4-7]

Dysuria was the most common symptom, and no specificity of symptomatology was found compared to that caused by other malignant prostate tumours.

The most common sites of prostate cancer metastasis were bone, followed by lung and liver.

Approximately 40% of patients with PNET of the prostate have distant metastases.^[7] Lung metastases were first followed by bone and then liver metastases.^[10]

PSA is an essential serum marker for the diagnosis of prostate cancer. Nevertheless, none of the cases of prostate PNET recorded in the literature had a PSA above the normal range (reference <4.00 ng / ml).^[11]

In our case the initial symptom was dysuria, the patient's PSA level was normal and the patient did not present any metastases.

Radiology is important for diagnosis, evaluation of locoregional and distant extension, pre-therapy staging and post-therapy follow-up.

It describes a large heterogeneous mass with hemorrhage and necrosis. Sometimes it may be accompanied by adjacent osteolytic erosion.^[9] Similar to the radiological images of our patient. However, in general, the imaging features of pPNET showed no evident specificity.

The histology shows a proliferation of small undifferentiated round cells, which are arranged in a laminated, lobulated or nest-like pattern. The presence of the characteristic structure of Homer Wright's rosettes or pseudo rosettes is inconstant, and the diagnosis is confirmed by immunohistochemistry.^[12]

Neuronal markers include mainly chromogranin, Synaptophysin, CD56 and CD99. CD99 is the most important and sensitive marker.^[13-14]

No international standards for prostate PNET are currently available. However, surgery, radiotherapy and chemotherapy are strongly recommended.

Surgical resection is the optimal option for patients at an early stage.

Neoadjuvant chemotherapy avoids mutilating surgery, reduces tumour cell dissemination during surgery and significantly improves the survival rate of patients.^[15]

Chemotherapy regimen is the VAC protocol (doxorubicin + vincristine + cyclophosphamide) IE (ifosfamide + etoposide).

CONCLUSION

In summary, we report a case of an extremely rare malignant tumour, PNET of the prostate, which is usually shows an aggressive biological behavior and a severe clinical course, generally correlated with a poor prognosis.

But coordinated multidisciplinary collaboration in diagnosis and treatment should improve the survival of these patients.

In our case, combination chemotherapy consisting of ifosfamide, doxorubicin, vincristine, and etoposide, and adjuvant radiation therapy without resection of the tumor provide a successful therapeutic result until now.

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