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Primitive Neuroectodermal Tumor of the Prostate, A Case Report and Literature Review

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ABSTRACT

Sarcomas of the prostate are rare and represent less than 0.1% of cancers of this gland, represented mainly by leiomyosarcomas. It is a rare type of tumor, with a 5-year survival rate of approximately 53%. occur mainly in children and young adults. These tumours show predilection for bones and soft tissues in the paraspinal region and lower extremities . Its therapeutic means represent a real problem for the urologist. We report the case of a young patient admitted for urine disorder, the paraclinical examination revealed a prostatic PNET.

Introduction

Sarcomas of the prostate are rare and represent less than 0.1% of cancers of this gland, represented mainly by leiomyosarcomas [1].

A primary neuroectodermal tumor (or PNET, standing for “primitive neuroectodermal tumor”) is a malignant tumor, a cancer of the neural crest. It is a rare type of tumor, with a 5-year survival rate of approximately 53%. [2]

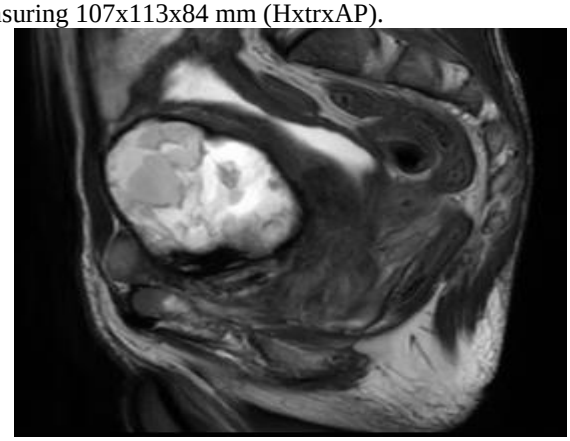
In this observation, we report the case of a young patient admitted for urine retention and in whom the paraclinical examination revealed a prostatic PNET.

Observation

Mr E.H., 32 years old, having consulted in the emergency department for complete urinary retention.

The questioning found no particular history besides a notion of pollakiuria with progressive dysuria , and we found a pelvic mass in the examination . So the patient had an urinary catheterization which gave 1,500 mililiter urine , hematic at the begining.

The PSA level was not too hight 0.9 ng / ml, and an MRI was performed showing the presence of a large prostatic process in heterogeneous T2 signal containing an area with T2 hypersignal (liquid) enhanced in an intense and heterogeneous way after injection, by diffusion hypersignal measuring 107x113x84 mm (HxtrxAP).



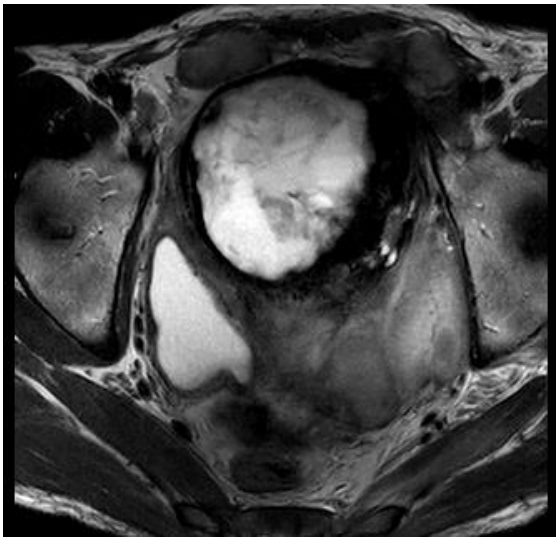


Figure 1. MRI showing a large prostatic process in heterogeneous T2 signal containing an area with T2 hypersignal.

The patient had a prostate biopsy. The histology examination showed a prostatic parenchyma massively invaded by a tumor proliferation made up of ovoid cells.

The nuclei were slightly vesicular and quite monomorphic, and the stroma was reduced to a rich capillary vascular network. And it was concluded to an undifferentiated malignant tumor of primary neuroectodermal type (PNET).

The immunohistochemical study showed tumor cells strongly expressing CD99 , CD56 , AE1 and AE3, desmine and myogénine, confirming the diagnosis of a prostatic PNET. A thoraco-abdominal-pelvic tomodensitometric examination, performed as part of the extension assessment, did not show any secondary location.

The patient had radiotherapy sessions (60 Grays on the tumor and 50 Grays on the pelvis).

After a 12-month, he still had urinary symptoms mainly like urinary burns without metastasis at the control CT scan.

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Discussion

Sarcomas of the prostate are rare and represent less than 0.1% of cancers of this gland, represented mainly by leiomyosarcomas [1].

A primary neuroectodermal tumor (or PNET, standing for "primitive neuroectodermal tumor") is a malignant tumor, a cancer of the neural crest. It is a rare type of tumor, with a 5-year survival rate of approximately 53%. [2]

Primitive neuroectodermal tumours (PNETs) are composed of small malignant undifferentiated cells and occur mainly in children and young adults. These tumours show predilection for bones and soft tissues in the paraspinal region and lower extremities [3].

Primitive peripheral neuroectodermal tumour has been described in a variety of primary visceral sites ; Extraskeletal PNET mainly occurs in the paravertebral region, the chest wall and the lower extremities, and less commonly in the pelvis, retroperitoneum or upper extremities [4].

MRI features of PNET of the kidney have been described in the literature as a heterogeneous lobulated mass with irregular septae within. The mass shows low signal intensity on T1W images and appears heterogeneous on T2W images. Haemorrhage and necrosis are also seen in the periphery of the tumour. The septae within the mass enhance on post-contrast scans [5].

Microscopically, PNETs are characterized by primitive round cells reactive to anti-CD99 antibody (Mic-2). CD99 positivity distinguishes PNET from other rare entities such as lymphoma, rhabdomyosarcoma and other neuroendocrine cancers [6].

Its therapeutic means represent a real problem for the urologist [7,8], because these tumors frequently recur despite radical surgery. Lung and bone metastases have been reported [8,9].

Conclusion

Pathologists, urologists and oncologist should be familiar with this rare entity to treat it better. The development of research work on the specialized prostatic stroma will make it possible to better understand it.

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