

Peripheral primitive neuroectodermal tumor of seminal vesicles: Is there a role for relatively aggressive treatment modalities?

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Summary *A 50 year old white man received an incidental ultrasound diagnosis of hypoechoic mass interesting the right seminal vesicle. A CT scan showed the presence of a 7.8 cm roundish cyst, originating from the right seminal vesicle. He had been followed by the removal of the right seminal vesicle and both the cystic lesion. The histological findings of the specimen documented the presence of small round cells compatible with Ewing's sarcoma/PPNET. The patient received also adjuvant chemotherapy and radiation treatment. After 10 years, the follow-up is still negative.*

KEY WORDS: Seminal vesicles, Sarcoma, Peripheral primitive neuroectodermal tumor.

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INTRODUCTION

The seminal vesicles are an unusual site of primary malignancy in the urinary tract and primary malignant Ewing's sarcoma or its variant *peripheral primitive neuroectodermal tumor* (PPNET) are even more rare (1-2). The prognosis of extraskeletal PPNET is generally poor and an aggressive, multimodal approach is usually required (3). At our knowledge only one case of PPNET involving the seminal vesicles has been reported, with scarce information about the follow-up. Herein is described a case of PPNET arising from the right seminal vesicle with a ten year follow-up.

CASE REPORT

A 50 year old white man received an incidental ultrasound diagnosis of hypoechoic mass interesting the right seminal vesicle. At the time, he was 174 cm in height, 82 kg in weight and the body surface area was 1,97 m²; the performance status was grade 0.

The family and his own personal medical history was not significant. He did not refer any professional exposure to carcinogens and used to smoke 7-8 cigarettes per day. Complete blood cell count, routine chemistry profile and

urine analysis were all normal. The general physical examination did not reveal anything particular. At DRE, a palpable mass was recognized at the level of the right portion of the anterior rectal wall dislocating the prostate gland.

The abdominal ultrasound examination of the pelvis showed a roundish cyst within the right seminal vesicle of 8 cm in the widest diameter, with septa and corpuscular material, inside (Figure 1). The further imaging diagnostic work up consisted in a pelvic *computed tomography* (CT) scan and *intravenous pyelography* (IVP). The CT scan confirmed the presence of a 7.8 cm roundish cyst, originating from the right seminal vesicle; the urinary bladder resulted compressed and anteriorly dislocated (Figure 2). No wall contrast enhancement was detected and a plane of dissection was appreciable from the bladder, rectum and the lateral pelvic wall (internal obturator muscle). CT scan revealed another oval-shaped cyst of 3 cm in the widest diameter originating from the right seminal vesicle with the same densitometric features. The IVP confirmed also that the right supero-external margin of the bladder wall was compressed (Figure 3). The patient underwent a TC guided *fine needle agobiopsy* (FNAB) of the larger lesion that gave no significant findings. Therefore the patient was surgically explored through a midline incision of the lower abdomen. A cystic lesion was found against the right seminal vesicle that raised-up the bladder. Frozen section biopsies of the cystic lesion documented fibrous tissue with chronic inflammation. The removal of the right seminal vesicle and both the cystic lesions was performed. The post operative period was uneventful and the patient was discharged in few days. The histological findings of the specimen documented the presence of small round cells compatible with Ewing's sarcoma/PPNET (Figures 4-5).

After surgery the patient received adjuvant chemotherapy with a total of 13 cycles (Adriamycin, Vincristine, Cyclophosphamide, Actinomycin D, Ifosfamide, Etoposide), as well as radiation treatment (48 Gy) on the pelvis. After 10 years, the follow-up is still negative: PET total body was always negative and the patient is in a good general condition with normal urinary continence and good erectile function.

DISCUSSION

PPNET, histologically characterized by the presence of small round cells extracranially in soft tissues and bones. Ewing's sarcoma and *primitive peripheral neuroectodermal tumor* (PPNET) have been originally described as two distinct pathologic entities (4).

Because of their similar histologic and cytogenetic characteristics, these tumors are now considered to derive from a common origin cell and to be a part of a spectrum of neoplastic diseases known as the *Ewing sarcoma family of tumors* (ESFT), which also includes *extraosseous Ewing sarcoma* (EES), adult neuroblastoma, malignant small-cell tumor of the thoracopulmonary region (Askin tumor), paravertebral small-cell tumor, and atypical Ewing's sarcoma. They all derive from embryonal neural crest cells. The essential feature to diagnose PPNET or Ewing's sarcoma is the histoimmunochemistry with CD99. *Ewing family of tumors* (EFTs) represents a neoplastic entity characterized by specific chromosomal rearrangements.

The most commonly detected translocation involves the fusion of EWSR1 to one of the genes encoding ETS family of transcription factors, usually FLI1 or ERG.

The detection of specific translocations by *fluorescence in situ hybridization* (FISH), *reverse transcription-polymerase chain reaction* (RT-PCR), or both has become the diagnostic hallmark for the EFTs (5). PPNETs are uncommon and the primary PPNET of the seminal vesicles is quite exceptional. Differential diagnosis must include rhabdomyosarcomas, leiomyosarcomas, phylloides tumors, malignant fibrous histiocytomas, chondrosarcomas.

Differential diagnosis must also include benign conditions like benign fibroepithelial tumor (6), prostatic utricle cyst, the prostatic abscess, the hydrops, the cyst and the empyema of seminal vesicles or the ectopic ureterocele. Usually surgical treatment of sarcomas of the pelvis is extremely aggressive.

The choice to treat the present patient in a conservative

way was related to CT scan images that showed only roundish cystic lesions without any contrast enhancement in the cystic wall and the presence of a well defined plane of dissection from the bladder, rectum and lateral pelvic wall.

Usually in case of sarcoma of the seminal vesicles, they are treated by means of an aggressive surgical approach as radical prostatectomy or radical cystectomy with an inevitably negative impact on the quality of life of the patients. In the case here described, the very conservative surgical approach was followed by an adjuvant therapy: such a treatment provided good clinical results, lasted in the long term period.

Figures are posted in Supplementary Materials” on www.aiua.it

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