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Case Report Open Access

About the Rare Case of a Pelvic Primitive Neuro-ectodermal Tumor in a 37 Year Old Patient

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Abstract

Introduction: Primitive Neuro-Ectodermal Tumors (PNETs) belong to the group of malignant bone tumors. Their frequency is about 1% of sarcoma. The principal distinctive feature in a translocation concerning the EWS gene or t(11; 22)(q24; q12). They are even rarer when located in the extra-skeletal pelvic region.

Case report: We report a case of a 37 year old female patient with a PNET in the pelvic region that is found after a spontaneous abortion. The tumor is localized on the right latero-uterine ligament. The tumoral size is of 10 cm. After the coelioscopic diagnosis, the patient was treated with neo-adjuvant chemotherapy according to VIDE protocol and ensuring an important diminishing of the size of the tumor. She then had a total radical hysterectomy associated with a lymph node resection. The diagnosis was confirmed by histology, immunohistology examinations and molecular biology by hybridation in situ. The tumoral cells were classically small and round. They expressed CD99 and vimentine. Translocation t(11;12)(q24;q12) was positive. Two months later, the patient presented a tumoral relapse and received an adjuvant chemotherapy by VAI twice with a limited chemical and hematological tolerance that brought along palliative treatment.

Discussion: Extra skeletal PNETs found in the pelvic region are very rare. Our present case is also rare by the way in which it was found, by the age of the patient and its being in the pelvic region. Prognosis with these tumors is still very bad regardless of the optimization of a multidisciplinary answer.

Conclusion: The research of new biomarkers and target therapy could be a hope for the treatment of these rare and pejorative tumors.

Keywords: Primitive neuro-ectodermal tumors; Sarcoma; Spontaneous abortion

Introduction

Primitive neuro-ectodermal tumors (PNETs) are rare and aggressive tumors which are usually found in children and teenagers. Males are likely to get it but the rate of survival is higher in females. They are more frequently found in caucasian people than afro-americans. 20 to 25% of patients are metastatic when they are diagnosed and one third will have a tumoral relapse within 5 years of diagnosis regardless of adequate treatment [1,2]. PNETs show a similar histology, immunohistochemistry, and cytogenetics as Ewing's sarcomas [3]. Patients with these tumors have been treated according to protocols designed for rhabdomyosarcomas using vincristine, doxorubicin, and cyclophosphamide with ifosfamide and etoposide [3]. In the literature, the pelvic localization is very rare: 35 cases have been reported by Burgers [4]. In this report, we describe a rare case of a pelvis PNET.

Case Report

A 37 year old woman sent in by her gynecologist for an asymptomatic abdominal mass on the right side of the uterus randomly discovered after a non progressive pregnancy of 8 weeks. Her medical history consisted of a post-partum phlebitis and a miscarriage (G5P3 patient). An abdomino-pelvic scan showed the uterus and bladder pushed horizontally to the right by a heterogeneous mass presenting both liquid and solid appearance with arch like calcifications. The tumoral size is of 10 cm. The pelvic MRI (Magnetic Resonance Imaging) showed an egg like shape taking up the right side of the pelvis, with polycyclic outlines and a very heterogeneous matrix There was no modification with a Gadolinium injection. A coelioscopy was performed to take histological material. The pathological diagnosis of PNETs was made based on the presence of a tumoral proliferation composed of small round or spindle-shaped cells with a small amount of cytoplasma. We observed also a fibro-vascular tumoral stroma and any tumoral necrosis. The immunohistochemical (all antibodies and revelation system were those of Dako®) cell phenotype was positive for CD99 and vimentin and negative for CD56, chromogranin, synaptophysin, cytokeratin (AE1/AE3), Epithelial Membran Antigen, protein S100, desmin, CD3, CD20, estrogenic or progesterone receptors and alpha inhibin. The proliferation marker ki 67 was very high at 80%. Molecular biology using fluorescence in-situ hibridization were positive for translocation t(11;12)(q24;q12) (EWSR1 22q12 break (Kreatech®))Any metastasis were revealed by TEP (Positron Emission Tomography) or classical radiology (Chest X ray) Biologically, the liver and renal functions were normal and NSE was elevated:96,3ng/mL The patient was treated by a neo-adjuvant chemotherapy with VIDE protocol (Vincricstine, Ifosfamide, Doxorubicine, Etoposide); six cycles in all. After two and four cycles, the TEP scan showed a considerable reduction in size (4.8 cm) and in metabolic intensity of the tumoral mass. We saw stability after six cycles. The patient then had surgery (increased colpohysterectomy but incomplete resection due to the mass infiltrating the near totality of the psoas muscle). The tumor continued growing. The patient became symptomatic two months later with diffuse abdominal pains. Two adjuvant chemotherapeutic treatments by VAI (Vincristine, Dactinomycine, Ifosfamide) were then undertaken with worse clinical and hematological tolerances. After palliative care, the patient died one month later.

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Discussion

PNETs are a group of rare tumors. Their annual incidence is 2,9 per million population [5] first described PNET in 1918 [6]. Their understanding has significantly advanced over the last twenty-five years. They have a wide distribution of primary tumors sites, related to their supposed neural origin [7]. On microscopy, they appear as a monotonous collection of small round, darkly stained cells leading to the differential diagnosis with a rhabdomyosarcoma and a neuroblastoma Fortunately, PNETs are characterized by a certain number of specific translocations of the EWS gene which is found on the chromosome 22 classifying these tumor in the PNET family/Ewing sarcoma. Over added molecular anomalies often occur during the progression of the illness such as the p53 or the p16 mutations [8]. Because their large size, their numerous distant metastasis, their poor radio sensitivity and their incomplete surgical resection, Pelvic PNETs show often a pejorative prognosis. Burgers et al. [4] described 35 cases (15 boys and 20 girls aged between 3 and 31 years). 24 of these patients showed any metastasis in time of the primary diagnosis. All were treated by chemotherapy associated for 31 with radiotherapy. Moreover, 4 patients had surgery. 5 Patients developed metastasis (pulmonary and skeletal). The pulmonary metastasis were operated on followed by chemotherapy and sometimes radiotherapy. Patients treated only by radiotherapy showed intra thoracic relapses. Skeletal metastasis were developed in cases treated by radiotherapy first and chemotherapy.

The difference of the 5 years Survival without progression between patients without and with metastasis, was respectively of 19% and 40% [4].

Polychemotherapy and surgery were the basis of the treatment of PNETs. Polychemotherapy uses 5 drug approaches: Vincristine, Ifosfamide, Ciclophosphamide, Etoposide and Doxorubicine. Complete surgical excision reduces drastically the risk of local relaps but doesn't prevent metastatic spread [9].

Conclusion

Survival of patients with PNETs on a single location has considerable increased with the arrival of modern chemotherapies as the target therapy but also with new surgical techniques by coelioscopy. Prognosis for patients who already have a diffuse illness or who without response of the initial treatment remains worse.

A better comprehension of signals may be a future path in the treatment of these rare tumors.

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