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Medulloblastoma like Neuroectodermal Tumour Arising from Ovarian Teratoma in a 16 Year Old Patient: A Case Report

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Case Report

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Abstract

Malignant neuroectodermal tumor with differentiation medullo-blastoma-like in an immature ovarian teratoma is a very rare and aggressive tumor. Only few cases are reported and most occur in adolescence. In our experience we performed a conservative surgery for a FIGO stage 1A tumor but bone recurrence followed within 5 months from surgery. Association of CHT (cisplatin-etoposide followed by tandem high-dose Thiotepa and auto transplantation of hematopoietic stem cell) and external beam radiotherapy was effective for the management of the recurrence but no guidelines are still available.

Keywords: Neuroectodermal Tumor; Abdominal Swelling and Pains

Abbreviations: ES: Ewing's sarcoma; AHCR: Autologous Hematopoietic Cell Rescue; PNET: Peripheral Primitive Neuroectodermal Tumor.

Introduction

The most common pediatric gynecological disease is the adnexal tumors, more precisely ovarian tumors. The incidence of ovarian tumors increases with age and it is considerably higher in girls over 14 years of age. Abdominal swelling and sometimes abdominal pain are the most common symptoms, and the diagnosis of ovarian tumor is usually set by trans-abdominal ultrasound scanning. Alpha-fetoprotein and CA-125 are the most frequently used biomarkers for diagnosis setting and subsequent patient follow-up [1]. Functional cysts, ovarian torsion and benign masses are the most common tumors detected in adolescents and young women. Ovarian neoplasm makes up a small percentage of the tumors affecting children and adolescents, i.e., less than 2% of all tumors and about 0.9% of the malignant ones [2,3]. Borderline ovarian serous tumors are even less frequent (5-10% of all ovarian serous tumors, which, in their turn, make up 20-50% of all ovarian tumors), and they are therefore reported in literature as clinical case studies or small series of cases.

The case reported in this paper is that of a 16-year-old adolescent with a malignant neuroectodermal tumor with differentiation medullo-blastoma-like in an immature cystic teratoma. The occurrence of a malignant neuroectodermal tumor in a mature cystic teratoma is extremely rare, and it manifests in 3 forms: differentiated, primitive, and anaplastic. The medulloblastoma belongs to the subgroup of primitive neuroectodermal tumor of the ovary. Only nine cases of ovarian medulloblastoma have been reported in the literature, and all of them are presented without information about the specific pathological subtype.

Peripheral primitive neuroectodermal tumor (PNET) is a soft tissue sarcoma of neuroectodermal origin, and is the second most common sarcoma among children and young adults. It is considered one of the small round blue cell tumors. It shares many morphological features with Ewing's sarcoma (ES), which arises usually in the bone, while PNET arises in soft tissue. PNETs in general are aggressive cancers associated with a high mortality [4].

Case Report

This paper reports the case of a 16-year-old adolescent who is hospitalized for abdominal swelling and pains the onset of which occurred several weeks. She had no family or personal pathological medical history. The physical examination reveals a round mobile relatively well-delimited and slightly painful on palpation tumor located in the hypogastrium.

The CA-125 biomarker had pathological value 87,4 UI/ml (cut off 35,0), as well as CA 19-9: 49,7 UI/ml (cut off 35,0), instead alpha-fetoprotein was normal and chorion gonadotropin was negative. We studied also HE4 that was normal: 53 pmol/l (cut off premenopausal women 70). The abdomen ultrasound scan reveals a 13 cm big well-delimited complex mass, with papillary structures, located in front of the uterus and on top of the urinary bladder.

The right ovary exhibits several follicular cysts, and the uterus was regular. The pelvic MRI detects a 13/11/10 cm big well-delimited liquid cyst, located in the pelvis and belonging to the left ovary. The inner anterior and inferior wall of the cyst presents multiple papillary structures, with contrast uptake after contrast medium injection. No pelvic suspicious lymph nodes are revealed.

A laparoscopic surgical procedure is performed, the peritoneal cavity is explored and the well-delimited tumor is observed. It has no adherence to the neigh-boring organs and it belongs to the left ovary. The tumor is considered to be in the FIGO stage IA. In order to avoid the accidental rupture of the capsule the surgeon decided for laparotomy. A left salpingo-oophorectomy was performed. The frozen section revealed a sex cord stromal ovarian tumor. Peritoneal washing was performed, multiple peritoneal biopsies (bladder peritoneum, ovarian dimple, omental and right ovarian biopsies). After the surgery the patient was treated with antibiotics and support therapy, and was discharged three days after the surgical procedure.

The histopathological examination performed in order to determine the exact clinicopathological feature revealed a malignant neuroectodermal tumor with massive differentiation medullo-blastoma-like in a ovaric teratoma. The results of biopsies were negative and also peritoneal washing. The CT thorax abdomen and pelvis following the surgical treatment did not reveal any distant metastases.

The malignancy was classified as a FIGO Stage IA PNET of the ovary (T1a NX M0). Subsequently, she was managed by a multidisciplinary team comprising of a gynecologic oncologist, a medical oncologist and a radiotherapist. In consideration of the FIGO stage no adjuvant therapy was administered. After 5 months because of backache, vertebral, sacral and cranial metastasis were diagnosed in other institution by a CT scan and led the physicians to decide for chemotherapy (III cycles of cisplatin-etoposide and III cycles of high dose thiotepa followed by auto-transplantation of hematopoietic stem cells) and EBRT ended in December 2019. From that date the patient followed a close follow up with no further progression or recurrence.

Discussion

The risk of malignant transformation develops in about 0.17 to 2% of mature cystic teratomas, and occurs most often in postmenopausal women [5].

Peripheral primitive neuroectodermal tumors usually have more neuroendocrine features. It is the second most common sarcoma among children and young adult, 4 it usually contains well-formed rosettes or pseudorosettes [6]. Most PNET occur in the soft tissues, however, rare cases have been reported in the ovary [7]. Usually these tumors behave clinically in an aggressive fashion, only a small percentage of patients with these tumors survive. The survival rate varies from 10.8 month to 3 years [7]. Peripheral primitive neuroectodermal tumor of the ovary with 2 successful spontaneous pregnancies has been described. The oldest age reported with ovarian PNET is a case of a 78-year-old woman, and it was associated with endometrioid adenocarcinoma [8].

The rarity of PNET of the ovary precludes randomized clinical study to guide the management of this disease hence the optimal treatment strategies have not been established. In general, ovarian PNET malignancies are highly aggressive and the prognosis is poor especially in the presence of extra-ovarian spread. One of the largest studies by Morovic and Damjanov [9] identified that disease stage appears to be the most important prognostic factor in PNET of the ovary. The majority of patients with Stage I disease (nine out of eleven cases) were alive and free of disease at a follow-up period of between three to five years. Many of the patients with Stage IA disease were treated with staging laparotomy, total hysterectomy bilateral salpingo-ophorectomy, omentectomy and pelvic/para-aortic lymphadenectomy only, whereas the Stage IC patients received chemotherapy in addition to surgery. On the other hand, women with Stage III or IV disease were often treated with surgery in combination with chemotherapy and or radiotherapy. Despite treatments, the prognoses of these women with advanced disease remained poor, the diseases were highly aggressive and rapidly gave rise to metastases and death.

Fertility-preserving surgery followed by chemotherapy for early stage PNET of the ovary with successful pregnancies

has been reported recently by Demirtas, et al. [10]. He reported a young patient with Stage IC PNET of the ovary who was treated with unilateral salpingo-oophorectomy, wedge resection of the right ovary and complete pelvic and para-aortic lymphadenectomy. She received adjuvant chemotherapy consisting of bleomycin, etoposide and cisplatin (BEP protocol). Her disease recurred shortly after and salvage chemotherapy with VIP protocol was administered. Seven months later, the patient was pregnant and delivered a healthy baby girl at term. Sixteen months later, the patient again delivered a healthy infant. However, the two studies by Morovic and Damjanov, and Demirtas et al. described patients with primary PNET of the ovary and not patients with malignant transformation to PNET within a mature cystic teratoma, which is the case here.

The optimal chemotherapy regimen for the treatment of cPNET transformed from germ cell tumors of ovary remains unknown. Management of these cases is based on case reports and largely extrapolated from our knowledge of the transformed male germ cell testicular tumors. For PNETs that are transformed from germ cell tumors, some authors advocate the use of cisplatin-based chemotherapy, such as BEP (Bleomycin, Etoposide and Cisplatin), directed at the germ cell tumors (Demirtas, et al. [10] many other authors however felt that despite arising within germ cell tumors, PNETs are usually resistant to cisplatin-based treatment [11] and therefore advocate chemotherapy regimens comprising of doxorubicin, ifosfamide and cyclophosphamide, directed at the transformed PNET component [12,13]. In one of the largest series by Ehrlich Y and colleagues [13] CAV/IE (Cyclophosphamide/Doxorubicin/Vincristine alternating with Ifosfamide/Etoposide), was the treatment of choice for PNETs transformed from testicular teratoma. Yet other authors had used integrated chemotherapy regimens that target both PNET and germ cell tumor [14]. Good outcomes were also described with tandem courses of High-dose thiotepa with autologous hematopoietic cell rescue (AHCR) in young patients with recurrent or refractory solid tumors particularly medulloblastoma [15].

Conclusion

The best treatment strategy for this kind of rare ovarian cancer is yet unclear, but the evaluation from a multidisciplinary team of physicians is mandatory to take into consideration all the possible strategies and try to offer a tailored treatment for each patient.

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