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Cutaneous Metastasis of Juvenile-Type Granulosa Cell Tumor

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1. Introduction

Granulosa cell tumors (GCT) of ovary is a malignant tumor originating from sex cord stromal cells. It accounts for 2%-5% of all ovarian cancers. The granulosa is classified separately in adult and juvenile pattern by clinical presentation and histologic characteristics. The juvenile type granulosa cell tumor (JGCTs) are extremely rare. The incidence of JGCTs on the GCTs is 6.2% [1,2]. It is diagnosed in young women and pre-pubertal girls at stage I disease. The literature is scanty. The guidelines of the optimal management are still controversial [3,4]. The prognosis of JGCT is excellent because of tumour recurrence or metastasis being rare. However, JGCT is regarded as a low-grade malignant neoplasm. There is a significant propensity for recurrence or metastasis. Recurrent or metastatic AGCT can manifests many years after initial surgery.

The pelvis is a common recurrence site. The abdomen, retroperitoneum, bone, liver, and lungs are also possible sites [4,5]. The cutaneous metastasis of juvenile-type granulosa cell tumor has never been reported. Here, we report a case of a woman with stage IC ovarian juvenile-type GCT with cutaneous metastasis 5 years after the initial diagnosis and surgery.

2. Case Report

Five years ago, a-12-year-old post-menarchal female with no specific prior medical history presented with 2 months of increasing abdominal distension. She reported heavy periods with intermittent bleeding. She denied previous sexual activity. Initial workup at a local hospital found peritoneal effusion and a pelvic mass with negative peritoneal cytology showing only mesothelial cells. Upon admission, her abdomen was enlarged to 1~2 cm below the xiphoid process. Physical examination revealed positive shifting dullness of her abdomen. The laboratory tests showed that the tumor markers including alpha fetoprotein (AFP), lactate dehydrogenase (LDH), Cancer antigen125 (CA125), carcinoembryonic antigen (CEA), and CA199 were within the normal range. Transabdominal ultrasound revealed a solid adnexal mass with cystic components, a normal uterus, and massive ascites. A contrast enhanced CT showed that a solid-cystic pelvic mass with the diameter of 45*43

millimeter and uneven thicken wall. The patient underwent laparoscopy examination. Approximately 8,000 ml green ascites were collected, and subsequent cytology exam was negative. A 20 mm × 15 mm × 6 mm right and ruptured ovarian tumor was observed. Unilateral salpingo-oophorectomy was performed, and the intraoperative rapid pathological examination reported ovarian GCT. The final pathological examination confirmed the diagnosis of juvenile -type GCT of the right ovary. Due to the capsule ruptured before surgery with tumor on ovarian surface, the patient was diagnosed with juvenile-type ovarian GCT (FIGO stage IC2). At least 6 cycles of chemotherapy were expected. The close follow up is required then. This year, the patient finds a mass in the incision. The contrast enhanced CT showed that a solid mass with the diameter of 4*3 millimeter in the incision. The patient admits to our hospital. The laboratory tests showed that the tumor markers including alpha fetoprotein (AFP), lactate dehydrogenase (LDH), Cancer antigen125 (CA125), carcinoembryonic antigen (CEA), and CA199 were within the normal range. The mass was incised in the surgery. The final pathological diagnosis of the recurrence of juvenile-type GCT. The 6 cycles of chemotherapy were expected after the operation. The patient has no recurrence in the period of follow up.

3. Discussion

The juvenile granulosa cell tumors (JGCTs) are rare ovarian tumors. They differ from adult granulosa cell tumors (AGCTs) with overall good prognoses. The AGCTs are well known for late recurrence [6]. The most JGCTs occur in individuals <30 years old. JGCTs have been observed mostly in children and young adults. The exact etiopathogenesis of JGCTs is unknown. In the other hands, the adult counterpart wherein mutations in the *FOXL2* gene have been identified as the key player in pathogenesis [7]. JGCT is an estrogen producing tumor. It hence is often associated with clinical manifestations of hyper-estrogenism. It is accounting for early clinical presentations in such patients. In premenarchal girls, isosexual precocious pseudo-puberty, bleeding per vaginum, irregular menstruation, virilization or hirsutism are common. Most common clinical symptoms of JGCT include abdominal pain and abdominal distention. Radiological features are often non-specific. The pure cystic masses without any papillary projections or less commonly entirely solid masses.

When detected at an early stage, the disease carries a favourable prognosis. So that establishing an early and accurate diagnosis is pivotal for better patient outcomes. It is often challenging to accurately diagnose these tumors clinically as well as on histopathology. This is mainly due to the rarity of the tumor and overlapping morphologic features with the other ovarian malignancies. Such as thecoma and small cell carcinoma of the ovary, hypercalcemic type. Especially when these occur in young women [8,9].

Due to the exceptionally low incidence of this condition. The literature about treatment and clinical course of JGCTs is scanty [10,11]. Moreover, the available literature usually considers all OGCTs as a single disease, without any differentiation from AGCTs. Surgery is the first step of treatment in this disease setting. Surgical approach depends on patient's age and disease stage. FSS is recommended to young patients in reproductive age with early-stage disease. Wedge biopsy of the contralateral ovary and lymphadenectomy are not recommended, as these sites are rarely involved. The international guidelines regarding treatment of stage I disease are still controversial. According to the European Society for Medical Oncology (ESMO), fertility sparing surgery (FSS) can be recommended in stage IA disease and for selected stage IC cases. Bleomycin, Etoposide and Cipslatin (BEP) or taxanes and carboplatinum (CT) postoperative chemotherapy is recommended in stage IC JGCTs. For the European Society of Gynaecological Oncology (ESGO), surgery alone is an option in stage IC1, while stages IC2 and IC3 JGCTs should receive adjuvant chemotherapy. National Comprehensive Cancer Network (NCCN) recommends adjuvant

chemotherapy as optional in stage I disease. Given the young age of these patients and the known acute and late side effects of BEP chemotherapy. CT postoperative chemotherapy is recommended [12,13].

Given the limited evidence deriving from retrospective data, further multicenter studies and international collaborations in this setting are strongly needed to confirm the present findings.

4. Conclusion

JGCT is a rare ovarian sex-cord stromal tumor affecting young women, premenarchal adolescent girls and children. The tumor has an overall favourable outcome despite having an aggressive and varied histomorphology. Surgery is the primary modality of treatment, with fertility-sparing surgery being preferred in early-stage tumors. Adjuvant chemotherapy and/or radiotherapy is offered in advanced stage tumors.

5. Conflict of Interest

No potential conflict of interests was disclosed regarding the publication of this paper by all the authors.

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