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Primary Ovarian Synovial Sarcoma: The Second Report

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Abstract

Introduction: Synovial sarcoma of the ovary is a very rare tumor reported only once in the past. It is the second softest tissue mass after rhabdomyosarcoma in adults but its usual site is extremities not ovary.
Case Presentation: Here we describe a 53-year-old woman with primary synovial sarcoma of the ovary with insufficient treatment and lung metastasis of the tumor.
Conclusions: Because of harmlessness symptoms, it is usually missed and correct treatment is delayed. When facing this type of tumor, referring to well-equipped centers with experienced surgeons in this field is recommended for sufficient treatment and best results.

Keywords: Synovial Sarcoma, Ovarian Tumor, Lung Metastasis

1. Introduction

Synovial sarcoma is a well-defined and malignant soft tissue neoplasm especially in young patients although it may occur in any age (1). 5% - 10% of soft tissue sarcomas are synovial sarcomas, which are high-grade tumors ranking second after rhabdomyosarcoma in adolescents and children (2). Epidemiologically, the overall prevalence has been reported 7.25 per 100.000 (3). It occurs in women more than in men (4). Trans location(X; 18), (p11; q11) is known as the etiology and it is reported that this genetic abnormality exists approximately in 90% of patients with the disease. In this translocation, SYT gene on chromosome number 18 fuses to either SSX1 or SSX2 on the X chromosome. The name of its tumor never expresses its origin and it is not supposed to be raised on synovial tissue of the joints but can occur anywhere in the body. In fact, the reason of this naming is the similarity between tumor cells and synoviocytes (4). Histologically, it is divided into three subtypes: Biphasic, monophasic, and poorly differentiated. Local recurrence and distant metastasis are common in up to 70%, and there is some evidence of tumor spreading (5, 6). Given the low incidence and harmlessness symptoms, exact diagnosis often is delayed leading to referring to non-third level hospitals, insufficient treatment, and undesirable results. Our case is one of them. Meanwhile to our knowledge, our presentation is the second case reported.

2. Case Presentation

We describe here a 53-year-old patient who came to our joint oncology committee at Imam Khomeini hospital affiliated to Tehran University of Medical Sciences, Tehran, Iran, with post-menopausal bleeding. She had undergone left ovarian cystectomy and right oophorectomy via laparotomy due to a solid mass with 15-centimeter dimensions 3 years earlier in one of the country's border cities. Her histopathological diagnosis was synovial sarcoma of the ovary and left luteal cyst confirmed with a second opinion of another pathologist. Immunohistochemistry staining included the fallowing markers: positive for CK and S 100, negative for CD 34, HMB 45, Desmin, and Ki-67. In fact, the management of the patient was at a setting with insufficient experience in this field and thus, she did not have complete surgery following this diagnosis. Of course, her surgeon stated that according to the history of four times cesarean sections as well as due to the recent surgery and the potential risks of complex laparotomy, she was advised to go to a Referral center, but she refused. Inevitably, she gave combined regimen of chemotherapy composed of Mesna+ Adriamycin+ Iphosphamid in five days every

three weeks for six cycles. At that time, her metastasis workup was negative. She did not have any symptoms in her follow-up period for three years until she presented due to suffering from post-menopausal bleeding. There was nothing special in her examination except for vaginal atrophy. We performed vaginal sonography, pipelle biopsy, thoracic and abdominopelvic CT scan with and without contrast for detection of potential metastasis according to her history of ovarian synovial sarcoma. Endometrial biopsy was normal. We discovered tree nodules in left lower lobe of the lung from 5 millimeters to 8 millimeters. In the joint committee, a second opinion of an experienced pathologist was scheduled to confirm synovial sarcoma and if the diagnosis was the same, complete surgery and chemotherapy would be considered for the patient. Microscopic examination revealed neoplastic proliferation denoted by sheets and nests and some arrangements of atypical cell with glandular, vesicular structures and pleomorphic nuclei with foci of necrosis (Figure 1). In immunohistochemistry, staining positivity was seen in the markers CKAE/AE3, EMA, Vimentin, and Bcl2 while the following markers showed negative staining: Calretinin, Inhibin, H-Caldesmin, CD10, and Ki 67 was 10% - 15%. Thus, Biphasic Synovial sarcoma was endorsed. Nevertheless, no part of the healthy ovarian tissue was seen. Biopsy of her lung nodules reveled tumor spreading, too. She underwent complete surgical staging and hysterectomy, unilateral oophorectomy, omentectomy and pelvic and para aortic lymph nodes dissection. During operation, we did not detect any structure as right ovarian tissue. This was because the whole ovarian tissue probably had been tumorized and resected 3 years ago. Her post operational period was uneventful. In addition, there was no tumoral tissue in all of the specimens in the pathological findings. She advised to perform 3 - 4 cycles of chemotherapy as neo adjuvant treatment before lung metastasectomy. However, despite our explanations and insistences, the patient did not agree and came back to her city. It has been 5 months since then and in her new CT scan of the chest, her responsible surgeon reported no change in lung nodules size. She is asymptomatic now and is under follow up.

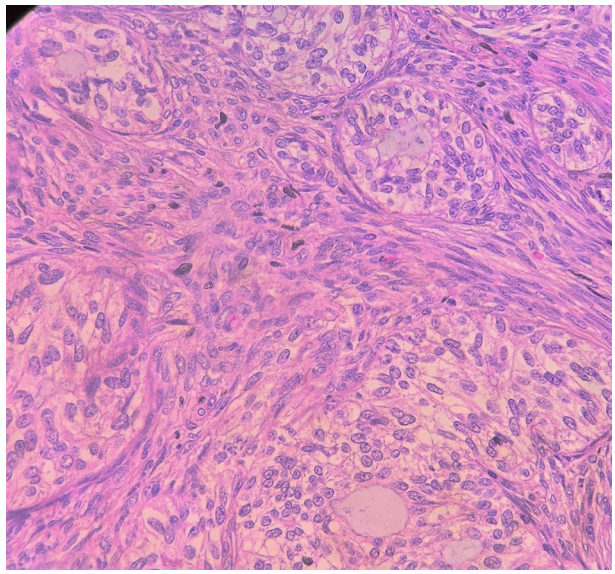

Figure 1. Microscopic examination shows neoplastic tissue composed of two cell populations, spindle cells with long spindle nuclei and eosinophilic cytoplasm and clusters of epithelioid cells with occasional glandular structures.

3. Discussion

Synovial sarcoma is one of the most common soft tissue sarcomas in adults and approximately 30% of cases occur in the two first decades of life. The average age at diagnosis is 30 years (7), but it can develop at any age. Our patient was 50 years at the initial diagnosis time. It is usually presented in lower and upper proximal extremities but can occur at any location such as stomach, kidney, uterus, pelvic, and ovary (1, 8-10). Typically, synovial sarcomas exist for a long time and are suddenly symptomatic by rapid growth. Our case had a large ovarian mass that caused abdominal pain. There is no consensus about prognostic factors but previous studies have reported Biphasic subtype, SYT-SS1 fusion, location in extremities, tumor size less than 5 centimeters, female gender, age below 50, and negative surgical margin (11). The presented patient had only two positive prognostic factors including female gender and Biphasic pathologic subtype. Nevertheless, she had almost all of the negative factors mentioned above. The most site of metastases is lung and it can occur late. Therefore, long-term surveillance should be considered. Other common sites are liver and bone, in sequence. Recurrence has been reported up to 69 months. So, long follow up is recommended to discover local or distant relapse of the tumor. Rarity of this tumor leads to a challenge in the management as well as in the treatment that is controversial. Radical surgery followed by radiotherapy and chemotherapy has been described (12). In our case, it was necessary to complete surgical staging before chemotherapy but she refused the correct treatment option. We completed the surgery and according to thorax surgeons' recommendation stating that neo adjuvant chemotherapy could be helpful, we advised her to receive neo adjuvant. She had a special character not to accept surgeon's opinion, leading to incomplete treatment in this stage similar to the previous steps, as described before. In lung involve-

ment of sarcoma, metastasectomy even if nodules are numerous is done that may increase overall survival in some patients (13). Of course, this option was not carried out for her. To determine whether the primary origin of the tumor is ovary, it is better to detect at least a small part of the normal ovarian tissue. Although we did not find it, there was no remnant tissue neither in follow-up imaging after the first surgery nor in pathological examination of second radical surgery. Therefore, it was a primary ovarian synovial sarcoma that based on our knowledge is reported for the second time.

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