**Case Report**

**Breast Metastasis of Alveolar Soft Part Sarcoma**

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**Abstract**

Alveolar soft part sarcoma is a rare tumor which presents less than 1% of all mesenchymal tumors [1]. It mostly occurs in young adults with a slightly female dominance [2]. Common places for metastasis of alveolar soft part sarcoma, like other sarcomas, are lungs, bones and brain but, other regions such as vagina, orbita, colon and breast can be involved very rarely [1,2]. In this study we present an alveolar soft part sarcoma with breast metastasis.

**INTRODUCTION**

Alveolar soft part sarcoma is a rare tumor which presents less than 1% of all mesenchymal tumors [1]. It mostly occurs in young adults with a slightly female dominance [2]. Common places for metastasis of alveolar soft part sarcoma, like other sarcomas, are lungs, bones and brain but, other regions such as vagina, orbita, colon and breast can be involved very rarely [1,2]. In this study we present an alveolar soft part sarcoma with breast metastasis.

**CASE PRESENTATION**

Female patient of 32 years old presented with multiple painful masses in both breasts. Physical examination showed numerous mass lesions of various sizes. There were a few lymphadenopathies in both axillary regions. Her history revealed left inguinal mass lesion surgery 7 years ago, and adjuvant oncologic treatment with the diagnosis of soft tissue sarcoma. She had breast ultrasonography, abdominal CT scan, PET/CT and percutaneous breast biopsy. PET/CT demonstrated malignant-like hypermetabolic multiple nodular lesions in bilateral breasts, metastatic nodular lesions in both lungs with 2 cm diameter of the biggest, and hypermetabolic malignant-like lesions in paraspinal and right upper extremity muscles. Pathology of the percutaneous core biopsy was interpreted as pleomorphic soft part sarcoma metastasis. The patient was then referred to medical oncology clinic for advanced metastatic disease, and she had 3 cycles of doxorubicin and ifosfamide. Afterwards, the patient received 30 Gy palliative radiotherapy. Chemotherapy was cancelled for a year after the follow-up examination due to low performance of the patient, and then a second regime of docetaxel and gemcitabine was given as three cycles. But, no regression was found in the lesions and significant pain and exulceration developed in her breasts (Figure 1). One month later, the patients were referred to general surgery clinic and palliative mastectomy was planned. Pre-operative preparations were completed and bilateral mastectomy was performed. The patient was discharged in the next day with no complication. The pathological evaluation and Immunohistochemistry of resection material showed pancytokeratin (-), HMB45(-), S-100(-), desmin (rare +), synaptophysin (-), chromogranin (-), PAS (+), d-PAS (+) and the nodular lesions of both breasts were interpreted as alveolar soft part sarcoma metastasis. All surgical margins were tumor free (Figures 2,3).

**ABBREVIATIONS**

ASPS: Alveolar Soft Part Sarcoma, CT: Computerized Tomography, MRI: Magnetic Resonance Imaging

**Keywords**

- Alveolar soft part sarcoma
- Breast
- Metastasis

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Alveolar soft part sarcoma constitutes only 0.2-0.9% of all soft tissue sarcomas [3]. It is more common in young patients, and is usually diagnosed between 15-35 years of age [3]. Localized disease is two times more in women, however general disease prevalence did not differ among sexes in a study of 70 patients in 2001 [4]. It has been reported that the most common locations of the primary tumor are lower extremities, but it can also be found in trunk, head and neck, like synovial sarcomas [4,5]. Retroperitoneum, tongue, bladder, orbita, braid and uterus are atypical regions of the tumor involvement [5,6].

The sites of metastasis are frequently lungs, bones and brain [7]. The possibility of synchronous metastasis is nearly 100% in the presence of lung metastasis [1]. It is recommended to perform cranial imaging of all patients diagnosed with alveolar soft part sarcoma due to frequent intracranial involvement. Moreover, in a study of Portera et al., the incidence of brain metastasis is 19% in stage 4 disease and they recommended selective imaging rather than screening [4]. Metastasis is observed in 65% of the cases at the time of diagnosis [4], however breast metastases are rare [8]. Literature search showed only 9 case reports similar to this case [9]. In all these nine cases, the primary lesions of breast metastasis are located in the thigh. Five year-survival rates are over 80% for localized disease and drop 20% in the presence of metastasis and the median survival is 40 months for these cases [1]. Prognosis is largely dependent on the initial presentation (whether it is metastatic or not), tumor size and age [4,10,11]. Patients with a large tumor in first presentation are most likely to have a metastatic disease at time of diagnosis [11]. Childhood alveolar soft part sarcomas have a better prognosis, particularly, the survival is near 100% for head and neck originated tumors [12]. The reason behind this is not completely understood, however, the differences in tumor biology are held responsible as possible explanation [3]. Nevertheless, in children, the masses are assessed at earlier size with advanced imaging and pathological evaluation, so that the disease might be diagnosed earlier and this would be contributory. Local recurrence occurs in 20% to 30% of cases [13].

The radical treatment is resection in localized disease. If metastatic disease is developed, conventional chemotherapy and radiotherapy are not satisfactory. It has been demonstrated in molecular and genetic studies that T(X;17) (p11.2;q25.3) translocation causes ASPSCR1-TFE3 fusion protein to arise, and that protein acts in MET signal pathway and induces tumoral cell proliferation and angiogenesis [14]. This condition is a milestone for the usage of MET kinase inhibitors and antiangiogenic therapies for targeted treatment of this disease. In a study performed in China with 66 alveolar soft part sarcoma patients, a tyrosine kinas receptor inhibitor, sunitinib, was used and showed promising results to increase disease free survival as well as total survival [15,16]. More studies are needed to better understand the nature and potential treatments of this condition.

REFERENCES


