CASE REPORT

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Wide Resection Treatment of Angiomatoid Fibrous Histiocytoma in a 42-Year- Old Female

Anjiyomatoid Fibröz Histiositomaası Bulunan 42 Yaşındaki Hastanın Geniş Rezeksiyon ile Tedavisi

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ABSTRACT

Angiomatoid fibrous histiocytoma is a rare neoplasm with malignant potential that primarily occurs in the subcutaneous tissues or dermis of the extremities in children or young adults. Its characteristic histological appearance involves the nodular growth of histiocytic, epithelioid, or spindle cells surrounded by a fibrous pseudocapsule containing a lymphocytic cuff. It may also feature a prominent myxoid stroma. Unresectable or metastatic tumors may have limited treatment options. Immunotherapy using PD-1/PD-L1 inhibitors could play a role in the future, although it is not yet widely adopted. The standard surgical approach for angiomatoid fibrous histiocytoma involves wide or radical resection of the lesion. In this article, we discuss the case of a 42-year-old female patient who presented to our clinic with shoulder pain and was subsequently diagnosed with angiomatoid fibrous histiocytoma after biopsy.

Keywords: Angiomatoid fibrous histiocytoma, Pd-1/pd-l inhibitors, wide resection

ÖZ

Anjiyomatoid fibröz histiyositoma, çocuklarda veya genç erişkinlerde çoğunlukla deri altı dokularda veya ekstremitelerin dermisinde ortaya çıkan nadir bir malign potansiyelli neoplazmadır. Lenfositik bir manşon içeren fibröz bir psödo kapsül ile çevrili histiyositik, epitelioid veya iğsi hücrelerin nodüler büyümesiyle karakterize edilen histolojik bir görünümü vardır. Belirgin miksoid stroma içerebilir. Rezeke edilemeyen veya metastatik tümörler sınırlı tedavi seçeneklerine sahip olabilir. PD-1/PD-L1 inhibitörleri ile immünoterapi planlaması gelecekte yer alabilir ancak henüz yaygın olarak kullanılmamaktadır. Anjiyomatoid fibröz histiyositomanın yaygın cerrahi tedavisi, lezyonun geniş veya radikal rezeksiyonunu içermektedir. Bu yazıda, kliniğimize omuz ağrısı şikayetiyle başvuran ve biyopsi sonrası anjiomatoid fibröz histiyositoma tanısı konulan 42 yaşındaki kadın hastayı tartıştık.

Anahtar Kelimeler: Anjiyomatoid fibröz histiyositoma, Pd-1/pd-l inhibitörleri, geniş rezeksiyon

Introduction

Angiomatoid fibrous histiocytoma is a rare neoplasm with malignant potential that predominantly occurs in the subcutaneous tissues or dermis of the extremities in children or young adults. most reported cases involve individuals aged between 10 and 30 years (1).

Angiomatoid fibrous histiocytoma belongs to the group of soft tissue tumors with unknown differentiation. Local recurrence has been reported in 15% of the cases, and metastases have been reported in 2-5%. There is no correlation between clinicopathological factors and clinical course (2).

It is most commonly found in the upper and lower extremities. The neck and scalp are also not uncommon locations in reported cases (3). The pelvis is not a common site for angiomatoid fibrous histiocytoma. Although it is not very common, some individuals with pelvic infiltration are treated with wide resection of the lesion (4).

Chemotherapy and radiotherapy can be used in patients with metastasis or widespread dissemination of the disease; however, there is no strict consensus on these treatments. Some centers use PD-1/PD-L1 inhibitors for treatment, but long-term results have not yet been reported for cases of angiomatoid fibrous histiocytoma. The most common surgical treatment is wide resection of the lesion. The

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prognosis heavily depends on the presence of metastasis, with the lungs being the most common site of metastasis reported (5).

Its histological appearance is characterized by histiocytic, epithelioid, or spindle cells surrounded by a fibrous pseudocapsule containing a lymphocytic cuff (6).

Case Report

A 42-year-old female patient was admitted to our clinic with left shoulder pain. Her primary complaint was pain during exertion while trying to perform household tasks. The patient was diagnosed with breast cancer in 2014 and had a history of chemotherapy, radiotherapy, and 1 surgery. Before treatment, consent was obtained from both the patient and the doctors.

On physical examination, there was no asymmetrical view of the shoulders. There was no swelling or redness in the left shoulder. There was a small amount of pain with palpation on the posterior superior edge of the humerus. The patient had a full range of motion in both shoulders, and there was no pain during shoulder joint movement. In her radiological images, a soft tissue lesion around 5x5 cm was identified on the superior posterior aspect of the humerus extending into the deltoid muscle. Magnetic resonance imaging was reported as a soft tissue lesion of a potentially benign nature. A full-body metastasis scan was performed on the patient using positron emission tomography-computed tomography before surgery, and no metastasis was detected.

Following the radiological evaluations (Figures 1, 2), the patient was discussed in the tumor board, and it was deemed appropriate to perform a fine needle aspiration biopsy by the interventional radiology department. The results of the biopsy revealed a low-grade/benign mesenchymal tumor with spindle cells and myxoid stroma. The report also stated that the tumor's myxoid component was predominant. There was no necrosis, and the atypia was mild.

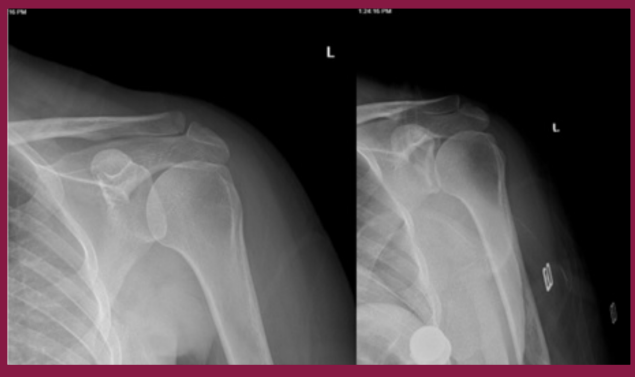

Figure 1. Direct radiological views of the shoulder joint

A few weeks after the fine needle biopsy, a wide resection of the lesion was performed (Figures 3-5) on the patient, and the specimens were sent for histopathological

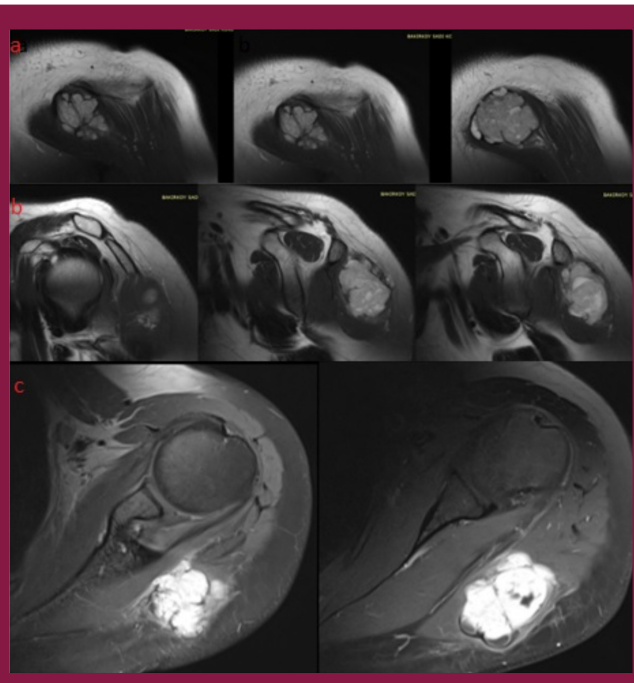

Figure 2. a-c. Magnetic resonance images of the lesion from different views

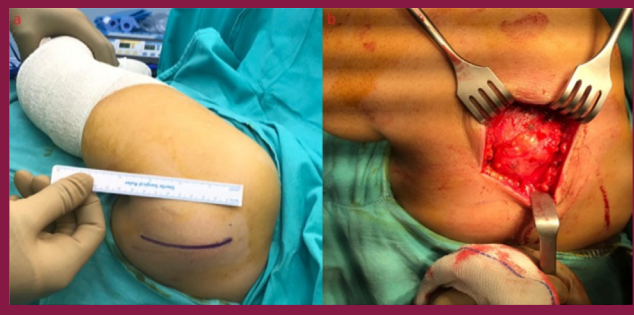

Figure 3. a, b. Peroperative images of the lesion. a) Before surgery, b) During surgery

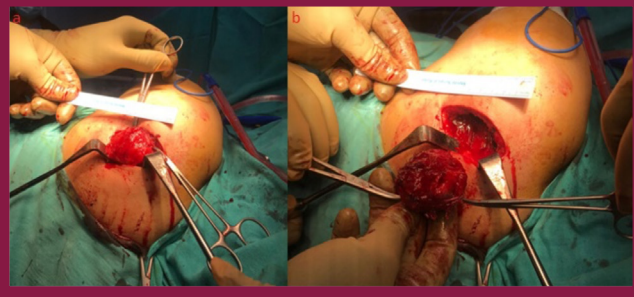

Figure 4. Peroperative images of the lesion during removal

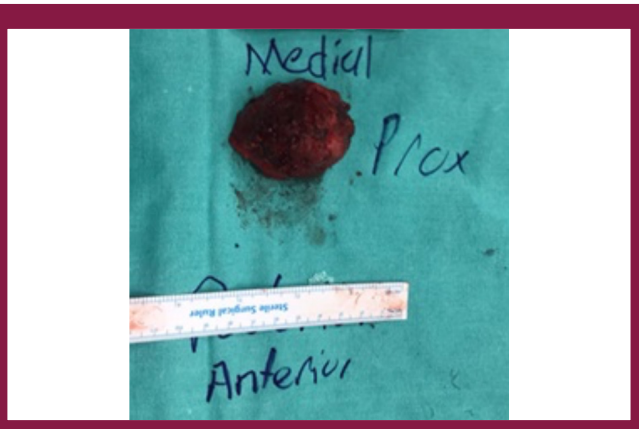

Figure 5. Peroperative pictures of the lesion after removal

investigations. The result of the wide resection material was "angiomatoid fibrous histiocytoma, myxoid form". During surgery, pathological examination revealed a negative surgical margin. The immunohistochemical markers were as follows:

- CD99: Cytoplasmic positive
- Desmin: Generalized diffuse positive
- EMA: Positive
- SMA: Focal positive
- S100: Negative
- BCOR: Negative
- STAT-6: Cytoplasmic positive
- CD34: Negative
- Ki67: 3-4%

The patient was discharged from the hospital on the day of surgery. Before discharge, the patient was taught pendulum and passive elbow exercises. The wound site was clear, and there was no leakage. The patient was then asked to visit for weekly check-ups in the polyclinics. During the check-ups, the patient was content and relieved of pain symptoms. The patient's shoulder range of motion was complete and without limitation.

After the pathological results were released, further molecular rearrangement of EWSR1 was studied and found to be positive. The report stated that no mutations, copy number increases, or rearrangements have been detected in the studied genes (7).

During the 2 years of postoperative checkups, there were no signs of recurrence or metastasis of the lesion in the control magnetic resonance images. No PD-1/PD-L1 inhibitors were used before or after treatment. The patient was recommended to undergo annual contrast-enhanced MRI follow-up.

Discussion

Most of the reported cases comprise female individuals between the ages of 10 and 30. However, our case was a 42-year-old female patient with a lesion occurring in a relatively common site. Some studies have shown that age has no prognostic value in angiomatoid fibrous histiocytoma (8,9).

Angiomatoid fibrous histiocytoma is classified as a soft tissue tumor with unknown differentiation. The patient's pathology report states, "the tumor's myxoid component is predominant. There is no necrosis, and the atypia is mild". In cases of angiomatoid fibrous histiocytoma with low atypia and either no or minimal necrosis, metastasis is uncommon, and the tumor does not exhibit aggressive behavior. These findings are consistent with the clinical presentation of our patient (9).

Among the options of fine-needle aspiration biopsy, core biopsy, and surgical biopsy, surgical biopsy is the most effective and definitive method for making a diagnosis. To assess the possibility of medical treatment, a fine-needle aspiration biopsy was initially performed. Surgical excision was considered appropriate following the diagnosis of angiomatoid fibrous histiocytoma.

The most notable treatment for angiomatoid fibrous histiocytoma is surgical-wide resection of the lesion. We also performed wide resection surgery, and there were no complications during or after the surgery. The lesion was encapsulated and limited to the soft tissue, with no spread to bone or neurovascular bundles. Resection of the lesion was easily accomplished during surgery (7).

The time between diagnosis and surgery was approximately 1 month. During the surgery, the size of the lesion was approximately the same as that in the magnetic resonance image, and there was no growth between the two periods [the magnetic resonance imaging (MRI) was taken about 2 and a half months before the surgery].

We did not use any biological drugs or PD-1/PD-L1 inhibitors during the patient's treatment. There may be beneficial results in postoperative follow-ups, but the long-term effects are still uncertain (7). The most commonly used PD-1/PD-L1 inhibitors are pembrolizumab, nivolumab, and cemiplimab, and atezolizumab, avelumab, and durvalumab. They have been predominantly employed in diseases such as melanoma, lung cancer, and urothelial cancer. (10). Reviews conducted to date indicate that in most cases, wide resection of the lesion is sufficient as treatment, and further chemo/radiotherapy or biological agents are not required. Similar to the literature, our patient has been successfully treated with only-wide excision without receiving chemotherapy or radiotherapy.

Conclusion

Angiomatoid fibrous histiocytoma is a rare neoplasm that predominantly occurs in subcutaneous tissues of the extremities in children or young adults and can be successfully treated with surgical removal. Chemotherapy and radiotherapy are generally not recommended for treatment. Wide resection is the preferred approach. The use of PD-1/PD-L1 inhibitors is still under discussion and may play a primary role in future treatment.

Informed Consent: Before treatment, consent was obtained from both the patient and the doctors.

Peer-review: Externally and internally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: A.C., M.K.Ö., H.B., Concept: A.C., M.K.Ö., H.B., Design: A.C., Data Collection or Processing: A.C., M.K.Ö., Analysis or Interpretation: A.C., M.K.Ö., H.B., Literature Search: A.C., M.K.Ö., Writing: A.C.

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