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Rare Fit on Ankle Dermatofibrosarcoma Protuberans

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Abstract

Dermatofibrosarcoma protuberans is a rare, slow-growing mesenchymal tumor of soft tissue with low-to-moderate malignancy potential, high local recurrence, and low distant metastatic activity. Although it is frequently seen in the trunk and proximal upper extremities and head and neck region, there are a few cases in the literature in localizations such as lower extremities and breasts. Therefore, it is not among the first diagnoses that come to mind in lower extremity lesions and is diagnosed after histopathological examination. In this case, a 41-year-old female patient who had thought to be a callus on the posteromedial left ankle for approximately 3 years and therefore neglected was reported as having low-grade dermatofibrosarcoma protuberens after excision and had a suspicious lesion in the left breast for the same diagnosis on detailed examination.

Keywords: Dermatofibrosarcoma protuberans, ankle, callus, soft tissue tumor

Introduction

Dermatofibrosarcoma protuberens is a very rare malignant soft tissue tumor with an incidence of 0.1% among all cancers and less than 2% among soft tissue sarcomas.[1] Although it is more common in the third and fourth decades, it can be seen at any age; it has an extremely low rate of distant metastasis and slow growth but a high local recurrence rate.[2,3] Therefore, wide local excision of at least 2-3 cm of the resection margin should be planned in these patients, and a multidisciplinary approach and long-term follow-up are required to achieve optimal results.[4,5] Magnetic resonance imaging (MRI) is used for surgical planning and follow-up for recurrence, and computed tomography (CT) and ^{18}F-fluorodeoxygl ucose-positron emission tomography (^{18}F-FDG PET/CT) are used to evaluate metastasis and response to treatment.[6] In the presence of close surgical margins, reexcision or neoadjuvant or adjuvant radiotherapy is the treatment method applied to reduce the risk of local recurrence.[3,5,7]

Dermatofibrosarcoma protuberans is frequently seen in the subcutaneous tissue of the trunk and upper extremities; there are very few cases in the literature in which it is observed in the breast and rarely in the lower extremities.[7-9] In this case, a patient with a rare localization of dermatofibrosarcoma protuberens is presented.

Case Presentation

A 41-year-old female patient. She applied with a request for excision of the lesion known as callus, which has been present for approximately 3 years in the posteromedial region of the left ankle. The patient had a known fibroadenoma in the right breast; there was no other diagnosed disease, previous surgery, or history of drug use. Although it is painless, it causes more discomfort as time passes while wearing shoes. The smooth-surfaced nodular lesion, approximately 1 × 0.5 cm, with partially hyperkeratotic skin on it, was excised under local anesthesia (Figure 1). Pathological examination, low-grade dermatofibrosarcoma protuberens, and basic immunohistochemical findings were reported as CD34 diffuse positivity and Ki-67 1-5% in tumor cells. The tumor was 0.5 cm from the base surgical margin and was considered adjacent to the margin. Therefore, a wide reexcision was performed, and the defect was repaired with a partial thickness skin graft taken from the medial right thigh. No tumor cells were found in the pathological examination of the reexcision. Chemotherapy and radiotherapy were not considered necessary, and follow-up was recommended. Foot MRI performed 2 months later revealed no local recurrence. A hypoechoic solid lesion with partially smooth borders and coarse calcifications was observed, with a size of 2 × 1 cm, 3 cm away from the areola, at the 2 o'clock position on the right breast, which did not differ significantly from previous imaging studies, and upon the patient's refusal, it was accepted as calcified fibroadenoma, and annual follow-up was recommended without histopathological diagnosis. Written informed consent was obtained from the patient who agreed to take part in the study.

Discussion

Dermatofibrosarcoma protuberans is a rare, locally aggressive, rarely metastasizing, malignant soft tissue sarcoma originating from the dermis. However, 10-year mortality is reported to be less than 3% in the literature.[10] Although dermatofibrosarcoma protuberans is rare, it has been reported to have rare localizations with the introduction of a small number of cases in the literature. Although it is mostly seen in the trunk and upper extremities, it is less common in the lower extremities, and the number of reported cases in which it is seen in the toes as the distal lower extremity is extremely low. This case, which was found to have a tumor in the left ankle, made this case valuable.[7] In addition, this lesion, which he neglected for approximately 3 years, thinking it was a callus, is in parallel with dermatofibrosarcoma protuberans being a painless and slowly growing tumor; however, it suggests that it may be beneficial for patients to plan as wide an excision as possible due to the

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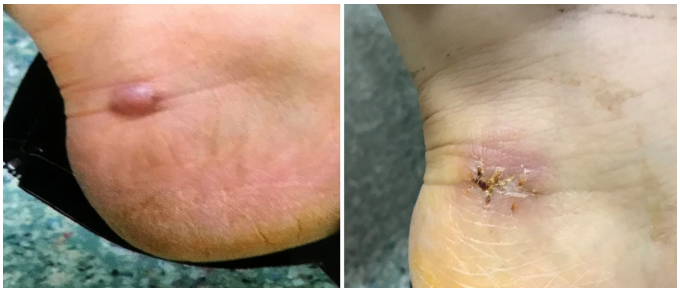

Figure 1. Dermatofibrosarcoma protuberens in the posteromedial left ankle. (A) Appearance during the first application. (B) View 14 days after excision.

possibility of malignancy in clinically suspected benign soft tissue lesions. Because it is known that surgical margins play an important role in local recurrence and metastases, the safe surgical margin is accepted as 4-5 cm.[10] In our case, the 0.5 cm surgical margin was accepted as adjacent, and reexcision was performed. Widespread expression of CD34 is extremely important in the histopathological differential diagnosis of dermatofibrosarcoma protuberans.[11,12] In our case, pathological examination, low-grade dermatofibrosarcoma protuberens, and basic immunohistochemical findings were reported as CD34 diffuse positivity and Ki-67 1-5% in tumor cells. After this diagnosis, the patient was re-evaluated in terms of fibroadenomas, since there are cases in the literature that were followed as fibroadenoma and histopathological examination resulted as dermatofibrosarcoma protuberans. Considering the slow growth feature of the tumor, it was decided to follow up with the patient's request. Magnetic resonance imaging was used for follow-up in terms of surgical plan and recurrence, and CT and ^{18}F-FDG PET/CT were used to evaluate metastasis and response to treatment[13] in our case who was followed up with control MRIs for recurrence. No recurrence was observed at the end of the first year.

Conclusion

Although dermatofibrosarcoma protuberens is a very rare malignant soft tissue tumor, it is important that the treatment approach and follow-up are extremely effective due to its locally aggressive nature. This is expected to develop together with the specific cases reported in the literature.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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