Case Report
Olgu Sunumu

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Cutaneous leiomyosarcoma misdiagnosed as a keloid: A case report with dermatoscopy

Keloid olarak yanlış tanı alan kutanöz leiomyosarkoma: Dermatoskopik bulgularla bir olgu sunumu

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Abstract

Cutaneous leiomyosarcoma (CLMS), with an annual incidence of 0.2/100,000, is a very rare non-melanoma skin malignancy. CLMS has two subtypes: Dermal (primary) and subcutaneous; when diagnosed in the early period, the prognosis in the dermal type is better. CLMS is generally seen in the 6th and 7th decades of life and more frequently in the lower extremities and head and neck. Trauma, radiation, chemicals, and sun rays have been reported as the factors involved in its etiology. Epidermal cysts, skin metastases, dermatofibrosarcoma pro-tuberans, and keloid should be considered in the differential diagnosis of CLMS, which usually manifests as a nodular lesion. In the presented case, the lesion, which developed over scar tissue was diagnosed as keloid and treated accordingly for four years. The pathologic examination of dermal CLMS (d-CLMS) in this patient, in whom diagnosis was delayed, revealed invasion of the subcutaneous fat tissue, which is among the poor prognostic factors. Herein, we present a case with d-CLMS, in which early recognition is vitally important, who was treated for an extended period as keloid, and we emphasize the differential diagnosis in CLMS and pathological and dermatoscopic features that, to our knowledge, are being reported here for the first time.
Keywords: Cutaneous leiomyosarcoma, keloid, dermatoscopy

Öz

Yıllık insidansı 0,2/100.000 olan kutanöz leiomyosarkoma (KLMS) çok nadir görülen bir melanom dışı deri kanseridir. KLMS'nin dermal (birincil) ve subkutanöz iki alt tipi vardır; erken dönemde teşhis edildiğinde dermal tipte prognoz daha iyidir. KLMS genellikle 6. ve 7. dekatta ve sıklıkla alt ekstremite, baş ve boyunda görülür. Travma, radyasyon, kimyasallar ve güneş ışınları etiyolojide rol oynayan faktörler olarak bildirilmiştir. Genellikle nodüler bir lezyon olarak ortaya çıkan KLMS ayırıcı tanısında epidermal kistler, deri metastazları, dermatofibrosarkom protuberans ve keloid düşünülmelidir. Olgumuzda da skar dokusu üzerinde gelişen nodüler lezyon 4 yıl boyunca keloid olarak tedavi edilmişti. Histopatolojik incelemede dermal KLMS (d-KLMS) tanısı alan bu olguda, tanıda gecikme sebebiyle kötü prognoz kriterlerinden yağ doku invazyonu da görüldü. Biz de keloid olarak yanlış tedavi edilen, erken tanının hayati öneme sahip olduğu d-KLMS olgumuzla, KLMS ayırıcı tanı, literatürde ilk olarak dermatoskopik ve patolojik özelliklerini ortaya koymayı ve vurgulamayı amaçladık.
Anahtar Kelimeler: Kutanöz leiomyosarkoma, keloid, dermatoskopi

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Introduction

Cutaneous sarcomas, non-melanoma skin cancers, are a group of malignancies with mesenchymal origin[1]. The incidence of cutaneous leiomyosarcoma (CLMS) is 0.2/100,000/year, and it is most commonly seen in the 6th-7th decades[2,3]. The lesions are most commonly reported on the trunk, upper extremities, head, and neck[1]. CLMS often presents as a slow-growing, erythematous nodular lesion, sometimes accompanied by pain and itching[1]. There are two subtypes of CLMS: the primary (dermal) type (d-CLMS) originates from the erector pili muscle and smooth muscle cells surrounding the sweat gland, and the subcutaneous type (s-CLMS) arises from the vascular smooth muscle cells. The two subgroups are clinically similar[3].

Early diagnosis is very important in CLMS. CLMS can be confused with some diseases due to its rarity and non-specific clinical presentation. We herein present a case of d-CLMS, who was treated for an extended period as keloid, and we emphasize the differential diagnosis of CLMS with its dermatoscopic features.

Case Report

A 37-year-old male presented with a tender nodular mass in his abdominal region. The patient reported a previous history of expansive scar tissue over an abdominal incision after surgery in 1987. In 2013 a nodular mass arose on the left lateral side of the scar and grew over time. The lesion was totally excised in 2015 and histopathological diagnosis was reported as keloid. Immediately after the excision, the lesion grew rapidly, with accompanying pain. Monthly intralesional corticosteroid injections were performed 5 times. During admission to our department for the first time, his physical examination revealed a 3x1 cm, slightly erythematous, painful nodular lesion which was firm on palpation, on the left superolateral side of the umbilicus (Figure 1). There was an atrophic scar tissue concomitant to this nodular mass. There were no palpable lymph nodes.

Polarized dermatoscopy revealed a central white structureless area, serpentine vessels, brown-pink structureless areas, and delicate brown reticular lines at the periphery (Figure 2). No sign of metastasis was detected on positron emission tomography/computed tomography. On abdominal magnetic resonance imaging, a 16x34 mm mass was detected on the left side of the anterior abdominal wall, involving the skin and the subcutaneous tissue. Excisional biopsy revealed infiltrative tumoral growth occupying the entire dermis and invading the subcutaneous adipose tissue (Figure 3). There were 8 mitotic figures per 10 high-power fields. Immunohistochemical stains demonstrated positive reactions for smooth muscle markers including smooth muscle actin and desmin. Based on these results, the diagnosis of d-CLMS was made.

Informed consent was obtained.

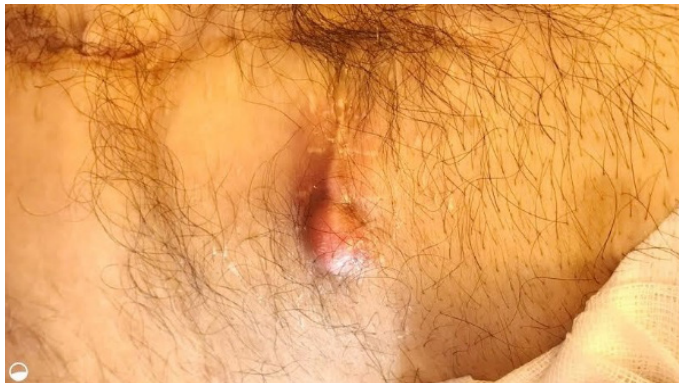

Figure 1. Nodular lesion (3x1 cm) located at the left superior lateral of the umbilicus

Discussion

CLMS is an extremely rare malignant superficial soft tissue sarcoma and is more commonly seen in the 6th and 7th decades of life[2,4]. Our case was younger than the reported cases. The majority of the patients

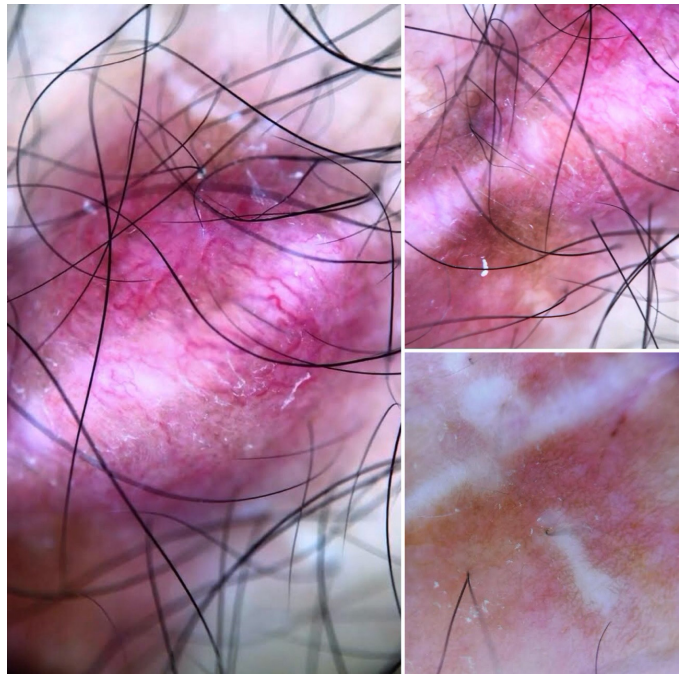

Figure 2. White structureless areas and irregular, serpentine vessels in the central zone. Brown-pink structureless areas and delicate brown reticular lines at the periphery

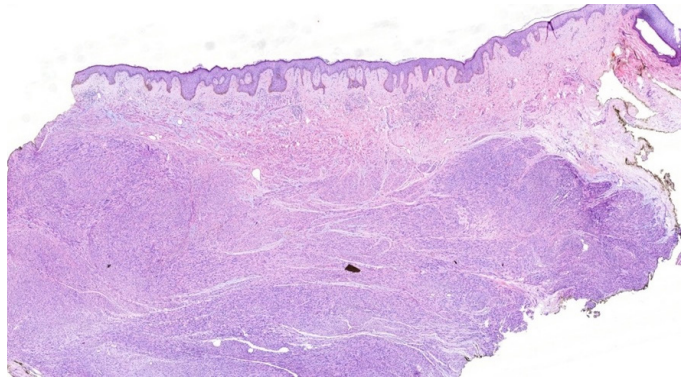

Figure 3. Histopathological features of cutaneous leiomyosarcoma: The tumor is composed of spindle cells intersecting each other and forming long bundles, tumoral growth occupying the entire reticular dermis and subcutaneous tissue starting from the surface (hematoxylin & eosin stain x40)

with CLMS presented with a solitary nodule with a median size of 1.8 cm at presentation. Pain may be present at presentation in one-fourth of the patients or could be elicited on pressure[5]. The tumor was very painful in our patient.

Trauma, ionizing radiation, sunburn, and chemicals may play a role in the etiology of CLMS[2]. There are also reported cases of CLMS arising on a scrofuloderma or a smallpox scar[6,7]. A history of possible trauma was reported by our patient as well and the d-CLMS developed on a surgical scar. However, there is no case of CLMS developing from keloid in the literature. In our case, we think that the lesion was CLMS from the very beginning and it was misdiagnosed as keloid.

The differential diagnosis of CLMS includes epidermal cysts, granulomas, skin metastases, dermatofibrosarcoma protuberans, and keloid. Because the development of the lesion over the scar tissue and histopathologic findings were consistent with keloid, the diagnosis was delayed for four years, and the patient was treated as if it was a keloid. The previous pathology preparations of the patient could not be reached. Immunohistochemical examination is very important in the diagnosis of CLMS. We think that the correct diagnosis could not be made because an immunohistochemical examination was not performed in the previous pathological examination. However, rapid growth after total excision and unresponsiveness to repeated intralesional corticosteroid injections, painful, very firm, and infiltrated nature of the mass, and the dermatoscopic findings were not compatible with keloid. The reported dermatoscopic findings of keloid include curved, linear, and branched vessels[8]. Brown reticular lines, and white and brown structureless areas are not reported in keloid as observed in our case. These dermatoscopic findings in combination with the clinical presentation suggested dermatofibrosarcoma protuberans which is the most common type of cutaneous sarcomas, in our differentials[9]. Central serpentine vessels might have been either the result of intralesional corticosteroid injections or caused by the pressure of the tumor under the dermal plexus. To our knowledge, the dermatoscopic features of CLMS have not been reported in the literature previously.

s-CLMS has a mortality rate of 30-40%, and a poorer prognosis than dermal type[3]. Subcutaneous invasion of d-CLMS worsens the prognosis[2]. The presence of infiltrative margins extending to the subcutaneous fat tissue in our patient with d-CLMS and diagnosis after four years may worsen the prognosis. There are studies in which early diagnosis of CLMS and extensive excision with a clear surgical margin of at least 1 cm have been shown to increase survival in the long term[1,3]. Mohs surgery, chemotherapy, and radiotherapy are other treatment options[1,3]. Our case was successfully treated with complete excision and no recurrence or complication was observed after one year of follow-up.

CLMS should be considered in the differential diagnosis of painful, slow-growing lesions with atypical clinical features. In addition to pathological examination, dermatoscopic examination may be helpful in the early diagnosis.

Ethics

Informed Consent: It was obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: M.A., Design: M.A., Data Collection or Processing: S.B., B.N.A., M.A., A.H.O., Analysis or Interpretation: S.B., B.N.A., M.A., A.H.O., Literature Search: S.B., B.N.A., M.A., A.H.O., Writing: S.B., B.N.A., M.A., A.H.O.

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