A Case of Acral Cutaneous Leiomyoma

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Abstract: Cutaneous leiomyomas are smooth muscle tumours which compose the majority of non-uterine leiomyomas. Majority of cutaneous leiomyomas are associated with synchronous uterine leiomyomas. Ten percent of the cutenous leiomyoma patients were reported to have renal cell cancer. These three entites can ocur together in the Reed Syndrome. Here we present a 47 years old female who applied to the outpatient clinic with a painful subcutenous nodule on the right foot. The histopathology revealed cutenoue leiomyoma and the patient was screened for uterine leiomyomas and renal cell cancer, which were remarkable. We presented this case because it is a rarely observed cutaneous pathology and has great importance due to its association with renal cell cancer.

Keywords: Cutaneous, Leiomyoma, Painful, Reed, Syndrome.

1. INTRODUCTION

Cutaneous Leiomyomas are rare tumours that originate from smooth muscle cells. This is a pathology that is seen slightly more in females. Although these are rarely observed tumours, they compose the majority of non-uterine leiomyomas. Depending on the smooth muscle of origin and the cliniopathological features, there are three types of cutaneous leiomyomas. These are piloleiomyomas, genital leiomyomas, and angioleiomyomas [1]. Previous studies have showed that 70-98% of the female cutaneous leiomyoma patients also have synchronous uterine leiomyomas [2]. Furthermore, the synchronous appearance of uterine and cutaneous leiomyomas have been associated with an increased risk of renal cell cancer of papillary subtype [3].

2. CASE REPORT

A 47 years old female patient applied to the out-patient clinic with a painful nodule on the lateral aspect of right foot. The lesion first appeared 5 years ago with a tingling sensation and has increased in size within the years. A firm, mobile, 3mm subcutaneous nodule was palpated during the physical examination and the nodule was tender upon palpation.

The past medical history was remarkable for Behçet's Disease which was diagnosed 23 years ago. The patient has first applied to the rheumatology department due to joint stiffness, joint pain and oral aphtous ulcers. The patient has experienced 2 anterior uveitis attacks since the diagnosis. For the last 3 years, she has been using colchicine 1 mg/day, prednisolone 5 mg/day and azathiopurine 50 mg/day. Furthemore, she has been diagnosed with pulmonary hypertension 8 years ago; has experienced intra-cardiac thrombi formation and has had several attacks of lower extremity deep venous thrombi. For that reason, she has been under the survelliance of cardiologists and pulmonologists for 8 years and has been under anticoagulation.

Complete blood count, liver function tests, kidney function tests and electrolytes were requested; all were within the normal limits. Deep excisional biopsy was performed with the clinical diagnosis of ecrine spiradenoma due to its typical acral localization and pain upon palpation. The lips of the wound were approximated and primary healing was intended. The histopathological examination has revealed cutaneous leiomyoma. The tumour cells were positive for smooth muscle antigen (SMA) and were negative for ERG, CD34, S100, p40 and CK7. The Ki67 proliferative index was reported as 0-4%. The patient was referred to obstretrics for the survellience of uterine leiomyomas, upon transvaginal ultrasonography no uterine leiomyomas were visualized. The patient was screened for renal cell cancer by nephrologists as well. She did not complain of hematuria and the renal ultrasonography was unremarkable.

3. DISCUSSION

Cutaneous leiomyomas are smooth muscle tumours with a slight female predominance. They compose the majority of non-uterine leiomyomas. Cutaneous leiomyomas present with mobile, dermal or subcutaneous nodules that may be single or multiple.

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The clinical subtypes of cutaneous leiomyomas are piloleiomyomas, genital leiomyomas, and angioleiomyomas [1]. Piloleiomyomas originate from the arrector pili muscle of the pilosebaceous unit. It is seen between the ages 20 and 40. The usual localization is extremities and trunk [4,5]. Piloleiomyomas usually present with pain [6]. Genital leiomyomas originate from the labial/vulvar, dartoid or mamillary muscles. The ages of presentation of this unusal tumour has been reported as 32 to 65 years. The usual localizations are nipple, areola, scrotum, penis and vulva [1]. The lesion is not associated with pain [7]. Angioleiomyomas originate from the tunica media of the small and medium sized arteries. It is observed between the ages 20 to 60 years [5]. The usual localization of this tumour is the lower extremities for females, and the upper extremities for males [1]. Angioleiomyomas usually present with pain [4].

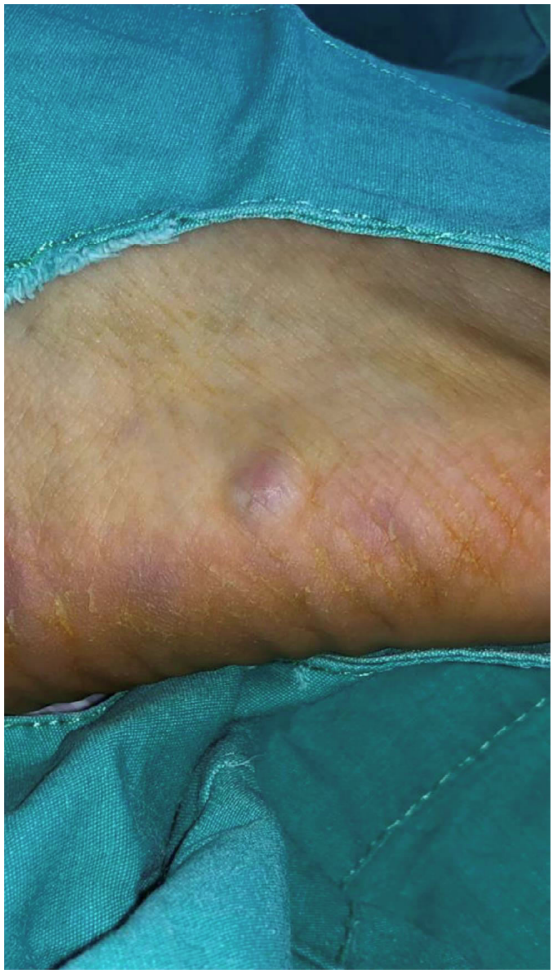

Figure 1: The pre-operative picture of the lesion: a firm, mobile, slightly violaceus nodule that was painful upon palpation.

To our knowledge, there are only 28 cases of acral leiomyomas that have been reported, all of which were angioleiomyomas; of these 7 were calcified and majority were painful [8-13]. The largest series of acral leiomyomas consisted of 21 patients and was reported by Hammond *et al*. in the year 2017. According to their series, 62% of the patients were females. The most common age range was 51 to 60 years. The most common site was the heel and 14% of the patients presented with a lesion on the lateral aspect of the foot similar to our patient. Sixty-seven percent of the patients with acral leiomyomas complained of pain. The previous cases held glomus tumour in the differential diagnosis; however ecrine spiradenoma has never been considered before, which was our initial clinical diagnosis [8]. Malik *et al*. consider ecrine spiradenoma as a differential diagnosis in their review, though [1]. Unfortunately, the subtype of our case has not been specified by our pathology department; therefore we cannot conclude whether or not our case is a angioleiomyoma definetly. On the other hand, the clinical presentation fits with the previous literature. Yet, the only definitive diagnostic method for cutaneous leiomyomas is histopathological analysis [1].

Beyond being a rare cutaneous tumour, an important aspect of cutaneous leiomyomas is their association with uterine leiomyomas and renal cell cancer, which is also known as the Reed Syndrome. Cutaneous leiomyomas are often the first manifestation of this syndrome, usually during the 4th decade and are often associated with pain, similar to our case. Ten percent of the patients with cutaneous leiomyomas have been reported have developed renal cell cancer. The most commonly seen subtype of renal cell cancer is papillary type 2 and the most common age of presentation is 44 years of age [14].

If left untreated, cutenous leiomyomas have a tendency to increase in size. The most commonly used treatment modality for cutaneous leiomyomas is excision, which is also a diagnostic method [1]. Excision is accepted as a gold standard treatment method [4]. Electrocauterisation, cryotherapy and ablative lasers (eg. Er:YAG and carbondioxide) are modalities that may be used in smaller lesions; however, they are not as curative as excision [1,15]. The recurrence rate of cutaneous leiomyomas are high, therefore negative surgical margins are of great importance [1]. The surgical margins were negative in our case.

4. CONCLUSION

Cutaneous leiomyomas are rare tumours, especially when it is located acrally. Furthermore, due to its

association with Reed Syndrome, cutaneous leiomyomas are of utmost importance, they are the inital manifestation of the syndrome which includes a mortal disease: renal cell cancer. For that reason, patients who are diagnosed with cutaneous leiomyomas should be rapidly screened for uterine leiomyomas and renal cell cancer. In conclusion, we presented this case since it was a rarely observed cutaneous pathology which has great importance due to its association with renal cell cancer.

DISCLOSURE OF INTEREST

The authors report no conflict of interest.

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