CASE REPORT: INTERESTING TOGETHERNESS OF MACULAR AMYLOIDOSIS AND MARJOLIN'S ULCER

OLGU SUNUMU: MAKÜLER AMİLOİDOZ VE MARJOLİN ÜLSERİNİN İLGİNÇ BİRLİKTELİĞİ

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ABSTRACT

Introduction: In most of the patients, constant friction and vigorous rubbing are the main cause for macular amyloidosis. Malign degenerations and diseases progressing with local irritation and inflammation could cause secondary cutaneous amyloidosis. In our case report, we want to share a Marjolin's ulcer developed on the base of chronically inflamed and irritated macular amyloidosis .

Material and Methods: 88 years old woman patient who has macular amyloidosis on her body suffers from a 9 x 8 x 2 cm ulcerated exophytic, cauliflower like skin lesion with erithematous surrounding on her thigh where macular amyloidosis is more concentrated. Lesion was excised with 2 cm skin margin above the muscle fascia and the defect was closed with split thickness skin graft.

Results: Its pathology result was " low differentiated squamous cell carcinoma" and all the margins were tumor free.

Conclusion: Even though, skin deposition diseases that have chronic inflammation and irritation could provoke malign tumors, we have not seen any togetherness of amyloidosis and Marjolin's ulcer in the litterature.

Keywords: Macular amyloidosis; marjolin's ulcer; squamous cell carcinoma; malign; deposition

ÖZET

Giriş: Sürekli kaşınma ve kuvvetli ovma, hastaların çoğunda maküler amiloidozun ana nedenleridir. Bölgesel tahrişe neden olan ve yangıyla seyreden hastalıklar ve malign dejenarasyon ikincil kutanöz amiloidoza neden olabilir. Bu vaka sunumunda, kronik yangılı ve kaşıntılı maküler amiloidoz zemininde gelişen Marjolin ülser vakasını paylaşmak istiyoruz.

Gereç ve Yöntem: Yıllardır vücudunda maküler amiloidoza ait lekeleri olan 88 yaşındaki bayan hasta lekelerin daha yoğun olduğu uyluğunda bulunan 9 x 8 x 2 cm boyutlarındaki etrafı eritemli, karnabahar görümlü, kaşıntılı lezyon nedeniyle tarafımıza başvurdu. Lezyon, 2 cm cerrahi sınırla kas fasyası üzerinden çıkarıldı ve oluşan doku defekti aynı bacaktan alınan kısmi kalınlıkta deri greftiyle onarıldı.

Bulgular: Patoloji sonucu "Az differansiye yassı hücreli karsinom ve cerrahi kenarlar temiz" olarak raporlandı.

Sonuçlar: Literatürde kronik yangılı ve kaşıntılı cilt depo hastalıklarının malign tümör oluşumunu tetikleyebildiğini gösteren yayınlar olsa da amiloidoz ve Marjolin ülser birlikteliğini vurgulayan yayın olmaması nedeniyle çalışmamız bu anlamda tektir.

Anahtar Sözcükler: Maküler amiloidoz; marjolin ülser; yassı hücreli karsinom; malign; depo hastalıkları

INTRODUCTION

Primary cutaneous amyloidosis is one of the common skin diseases. Its etiology is not fully known but environmental and genetic factors have an important role. It is classified as macular (friction) or lichen (papular) amyloidosis. Macular amyloidosis, which is less frequent than lichen amyloidosis, has prurutic (%82) eruption consisting of small, dusky brown or grayish pigmented, 2-3 mm macules with typical reticulated or rippled appearance and occurs more frequently in individuals with skin phototypes III and IV. It is distrubuted mostly over the trunk (upper back) followed by arms and buttocks respectively. Many risk factors are blamed for etiology such as UVB, EBV, race, genetic predispositions, atopy.[1] It has been already reported that it has relation with progressive systemic sclerosis, primary biliary cirrhosis, systemic lupus erythamatosus, pachyonychia, multiple endocrine neoplasia type 2.[2] However, in most of the patients, its cause might be friction which is first described by Hidano et al. in 1984. Constant friction and vigorous rubbing with cotton towels, horse-hair gloves, bath sponges, brushes, plant sticks and leaves are accused in all over the world. [1-6]

Malign degenerations e.g. squamous cell carcinoma, bowen disease, basal cell carcinoma are discussed in a great number in the litterature that they could cause secondary cutaneous amyloidosis. Also, systemic diseases or diseases triggering chronic irritation and inflammation such as chronic scars, chronic wounds, pilonidal sinus, fistule opening, decubitus ulcer, diabetic ulcer could be the reason for amyloid deposition 7,8 and for malign transformation (Marjolin's ulcer). However, in our case report we want to share a Marjolin's ulcer developped on the base of chronically inflamed and irritated macular amyloidosis lesion.

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CASE REPORT

Eighty eight-years-old woman patient has macular, pruritic, hyperpigmented lesions covering whole body except her face since her childhood. The lesions are more concentrated on her shoulders, chest and the extensonsor side of arms. She did not seek any medical treatment for these lesions previously. She doesn't have any family history for any type of skin lesions. In 2003, patient noticed that there are more dark-brown lesions on the posterior side of her left thigh that she frequently scratches. Patient did not have any medical treatment for her concentrating macules. In 2009, she noticed a papular skin lesion which is growing by time and bleeds temporarily. Patient always wears traditional long underwear which covers the part of her body from umbilicus to thigh. This underwear's lower part irritates her thighs and that could be the reason why she has concentrated macules on this area. Moreover, due to her age she spends most of her time sitting that increases the pressure on the posterior side of her thighs. We think that these conditions increased the local irritation on her left thigh already made by scratching. Patient had several skin biopsies from her macular and papular lesions and their results were "macular amyloidosis."

When patient presented in our clinic in 2010, she had a 9 x 8 x 2 cm ulcerated exophytic, cauliflower like skin lesion with eritematous surrounding on the posterior side and inferior half of her left thigh where macular amyloidosis was more concentrated than other parts of her body (Figure 1). We performed left inguinal and popliteal ultrasound and 17 x 9 mm lymphadenopathy with malign characteristic (irregular margins and hilus could not be seen) was observed in the inguinal area. Patient was operated under general anesthesia. Lesion was exicised with 2 cm skin margin above the muscle fascia and the defect was closed with split thickness skin graft taken from supero-lateral side of the same thigh (Figure 2). Patient's dressing was opened on the postoperative fifth day. Graft seemed healthy and taken. Patient was discharged on the seventh postoperative day. Its pathology result was "low differentiated squamous cell carcinoma" and all the margins were tumor free (Figure 3). After being had the pathology report, we did punch biopsies under local anesthesia from operation margin, surrounding tissues close to the operation area and pigmented area that is 10 cm further from the operation area; to double check previuos biopsy results of the skin lesions. Their results were respectively chronic inflammation, amyloid positive in small focal area and amyloid diffusely positive. Then, we informed patient about her condition and suggested her to remove left inguinal lymphadenopathy seen on the USG but she did not accept it. Patient was followed in outpatient clinic with dressings on alternate days while body scan (lung, abdomen and head CT and MRI) was performed and its result came clean. After all, patient was consulted to the medical and radiation oncology for further evaluation.

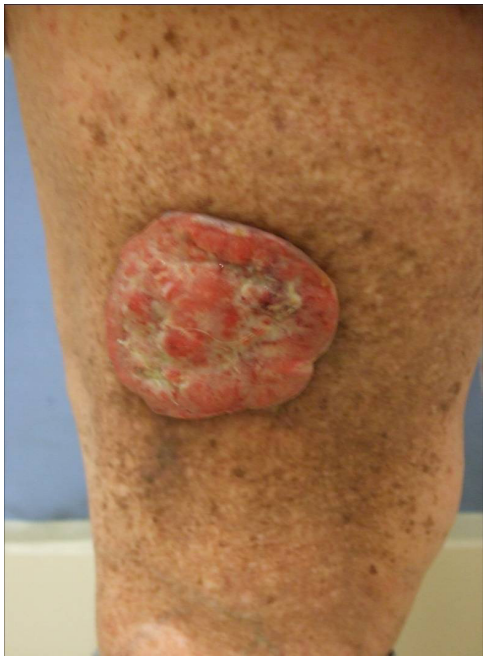

Figure 1. Preoperative view of cauliflower like skin lesion in thigh region

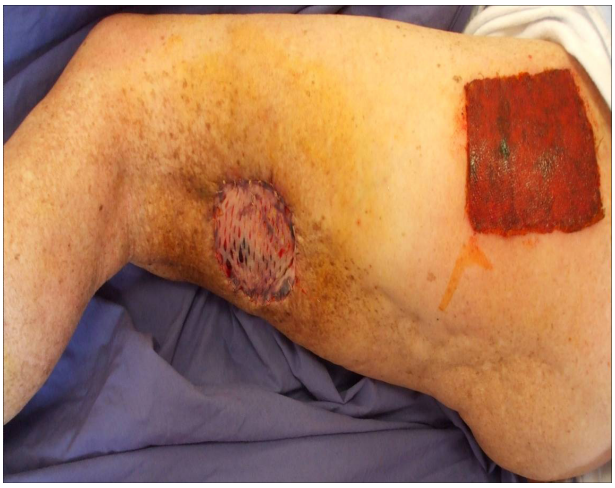

Figure 2. Postoperative fifth day view of grafted area

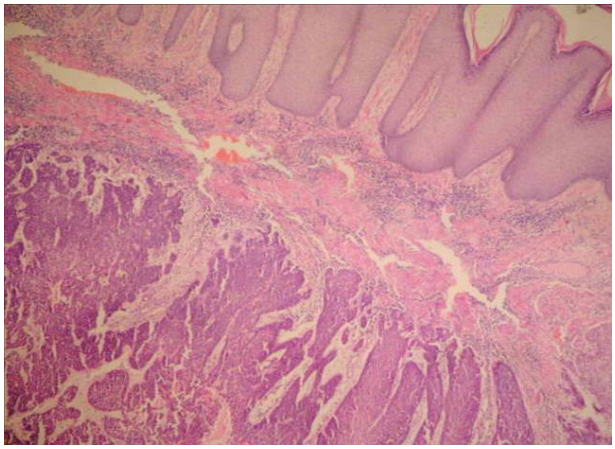

Figure 3. Hematoxylene-Eosine stained, stratified squamous epithelium originated squamous cell carcinoma on the surface (x100)

DISCUSSION

Amyloid seen in primary cutaneous amyloidosis, could be originated from keratinocytes. In 1979, Kumakiri et al. first observed fibroblasts and macrophages near amyloid islands but further they demonstrated the ultrastructural transformation of basal keratocytes into amyloid (Figure 4).[9] Hashimoto et al. proposed that necrotic epidermal cells are transformed into amyloid by dermal macrophages and fibroblasts by a process called filamentous degeneration.[10] Yamagihara proposed another theory that the amyloid is secreted by disrupted basal cells and is accumulated at the dermoepidermal junction.[11] Other theory is basal keratinocytes placed on the dermoepidermal junction produce precursor proteins and these are transformated into amyloid by inflammatory cells.[9,11] Moreover, apoptosis of basal keratinocytes could lead to amyloid formation and deposition.[12,13]

Regarding to the litterature, amyloid deposition due to malign tumors are not rare but we have not seen any report about Marjolin's ulcer developed on the ground of macular amyloidosis. Classically the term Marjolin's ulcer is used for the malignant degeneration secondary to burn injuries and diseases advancing with chronic inflammation and irritation that skin loses its property e.g. Bowen's disease,[14] chronic scar, chronic fistule, pilonidal sinus,[7] chronic venous ulcer, osteomyelitic sinus, thermal burn scar, stasis ulcer, physical and chemical frostbite, etc. The incidence of malignant skin tumors on scarred tissues is %0,1-2.5 and squamous cell carcinoma compose the majority (others are basal cell carcinoma, malignant melanoma and mezenchymal malignancies).[15,16] It is already known that skin deposition diseases like macular amyloidosis that have chronic inflammation and irritation could provoke malign tumors. Frequent irritation because of the traditional underwear and repetitive minor trauma caused by scrathing the lesions of macular amyloidosis might result with malign degeneration.

In conclusion, our case report is unique in the litterature by presenting a Marjolin's Ulcer grew on the ground of skin deposition dermatose presenting with pruritus.

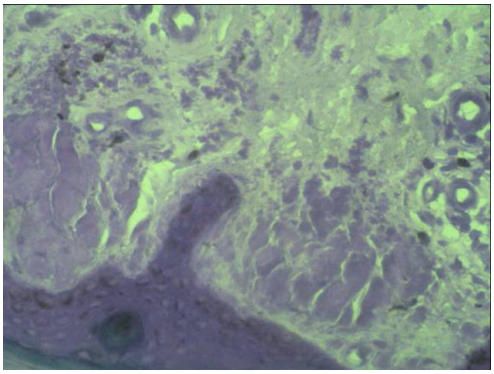

Figure 4. Methylene violet stained amyloid islands. (x200)

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