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Epitheloid Sarcoma Arising from a Burn Scar

- A Case Report -

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= 국문초록 =

화상반흔에서 발생한 상피세포양 육종 - 1예 보고 -

한림대학교 의과대학 병리과학교실, 이화여자대학교 의과대학 해부학교실^{*} 심 정 원·한 후 재*

악성종양의 발생은 중증 회상에서 가장 주의해야 할 합병증이다. 이 악성종양 중에 가장 흔한 것은 편 평세포 상피세포 암이고 기저세포 암, 악성 흑생종 등이 그 뒤를 따르며 육종 발생은 상피세포 암에 비해 현저히 빈도가 낮아, 현재까지 모두 24예만이 문헌에 보고되어 있다. 저자들은 화상흉터에서 발생한 상피세포양 육종 1예를 보고하고자 한다. 46세의 여자환자가 왼쪽 발바닥에 14년 전에 입은 열화상 이후 계속되어온 궤양성 상처와 종괴발생을 주소로 내원하였다. 3년 전에 다른 병원에서 같은 종괴에 대한 절제술을 받은 후 연부 조직 악성 종양을 의심 받았으나 별다른 치료 없이 지내왔다. 본원에 전원되어 시행한 광범위 절제술 후, 종괴는 현미경상 등근 상피세포모양의 종양세포들이 육아종 모양을 구성하고 있었고 핵의 이형성은 낮으나 세포분열이 흔하게 관찰되었다. 면역조직화학염색에서 종양세포 들은 vimentin에 강한 양성반응을 보임과 동시에 keratin, EMA 등의 상피세포 기원 항체에도 강한 양성반응을 보여 상피세포양 육종 진단을 가능케 하였다.

중심 단어: 화상 · 상피세포양 육종.

Introduction

It is well known that the development of malignant tumor in a chronic burn scar is one of the long-term complications of a severe burn. Most of these tumors are squamous cell carcinomas, and other carcinomas such as basal cell carcinoma and malignant melanoma have been reported to a lesser degree. However, sarcomas are much rarely seen in chronic burn scars. In the previous literature, 24 cases of burn scar sarcomas were reported ¹⁾, and epitheloid sarcoma has not been reported yet. The authors reported the first case of epitheloid sarcoma arising in a chronic, severe burn scar.

Case

A 46-year-old woman with tumor mass in the left sole was referred to this hospital. The patient had a history of severe thermal burn injury involving whole left foot and

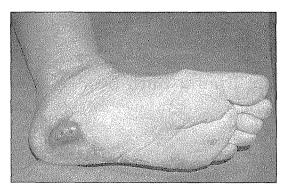


Fig. 1. Preoperative view of left sole showed burn scar with ulcer over the protruding soft tissue mass.

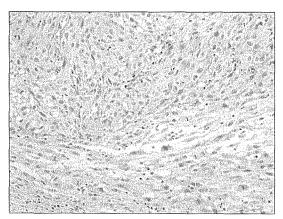


Fig. 2. Section showed round epitheloid tumor cells with bland nuclei and frequent mitoses (H&E, ×400).

ankle at the age of 31. The sole part of the injured foot had been repeatedly healed with scar and ulcerated with infection since that burn injury. Three years ago, firmly palpable mass began to grow on the sole lesion. Excinional biopsy was done at other hospital and malignant soft tissue neoplasm was suspected. She has been without any further treatment and presented to this hospital. On physical examination, on the left sole, there was a burn scar lesion with central reddish ulcer, measuring 4cm in diameter (Fig. 1). Beneath this lesion, round, firm, nontender mass, measuring 3cm in diameter, was palpated. There was no evidence of palpable lymph node along the left leg or distant metastasis to other organ. The red, firm, soft tissue tumor, measuring 3×2 cm, including the surrounding burn scar tissue, was widely excised.

Histological examination of the specimen revealed that soft tissue tumor mass was composed of monotonous population of epitheloid cells in vague granuloma like

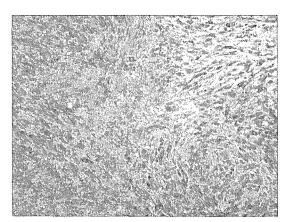


Fig. 3. Immunohistochemistry for EMA showed strong positive reaction (EMA, ×400).

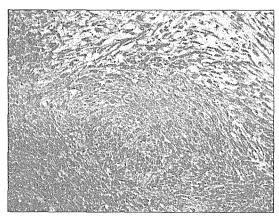


Fig. 4. Immunohistochemistry for vimentin showed strong positive reaction (vimentin, $\times 400$).

fashion (Fig. 2). The tumor cells had bland round nuclei and abundant acidophilic cytoplasm. Mitoses were frequently observed. There were little stromal tissue and rare necrotic areas. The tumor mainly located in reticular dermis and subcutaneous fat tissue, and skeletal muscle and aponeuroses were not involved. Immunohistologic examination showed positive staining with keratin, EMA (Fig. 3), CD34 and vimentin (Fig. 4), and negative staining with factor XIII, CD 31, S-100 protein, HMB 45, neuron specific enolase, smooth muscle actin, and CD99. These results supported the diagnosis of epitheloid sarcoma.

Discussion

This case was diagnosed histologically and immuno-

histochemically as epitheloid sarcoma, making it the first case report of epitheloid sarcoma arising in a burn scar. Of burn scar neoplasm, the most common malignancy is reported as squamous cell carcinoma, involving up to 71% and the second one is basal cell carcinoma, up to 12%, and the third one is malignant melanoma, up to 6%²⁾. Squamous cell carcinoma and basal cell carcinoma could be ruled out because of absence of epidermal lesion and positive reaction for vimentin and CD 34. Malignant melanoma could be ruled out because there was no reaction for S-100 protein and HMB 45. There have been 24 cases of burn scar sarcoma reported in the English literature²⁾. In these cases, 12 were diagnosed as malignant fibrous histiocytoma³⁾, the most common sarcoma arising from burn scar, and 4 were diagnosed as fibrosarcoma4). These two types of the major burn scar sarcomas should be ruled out at first. Malignant fibrous histiocytoma could be ruled out because there was no storiform pattern, no inflammatory cell infiltration, and the presence of keratin and EMA positivity. Fibrous sarcoma could be ruled out because there was granuloma pattern, not fascicular or herringbone pattern, and presence of keratin and EMA positivity.

The reason for the malignant degeneration in chronic wounds is not clear; the loss of skin elasticity causing from repeated trauma can produce ulceration and provoked malignant transformation⁵⁾, and restricted immunologic mechanisms in scars with avascular and obliterated lymphatic vessels could allow primary tumor growth⁶⁾. Considering that burn scar epithelial cancers occasionally behave more aggressively than usual⁷⁾, burn scar sarcomas may have a more aggressive character than those arising in unscarred tissue. In this patient, three years after the first excision, local recurrence on the same site was occurred. Continuous careful follow-up is

Radical excision appears to be the treatment of choice for epitheloid sarcoma⁸. Local recurrence occurred in 2 of 23 patients treated with wide excision in recent series. and skin flap taken from left groin area was covered⁹.

In conclusion, we report the first case of epitheloid sarcoma arising from a burn scar and we should recognize the possibility of the occurrence of various malignant tumors including epitheloid sarcoma arising from burn scars.

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