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Case Report

Multifocal cutaneous epithelioid hemangiodothelioma on the same lower limb

Mehdi Khallaayoune*, Fatima Azzahra Elgaitibi, Mariame Meziane, Nadia Ismaili, Laila Benzekri, Karima Senouci

Department of Dermatology, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

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***Correspondence:**
Dr. Mehdi Khallaayoune,
E-mail: khallaayounemehdi@gmail.com

ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare low-grade vascular tumour mainly affecting bones, deep soft tissues and exceptionally the skin. Multifocal cutaneous involvement is uncommon and should raise suspicion for metastasizing extracutaneous epithelioid hemangiodothelioma. Hereby we report the case of a young patient with multifocal cutaneous EHE involving the same lower limb. We emphasize the necessity for dermatologists to evoke the diagnosis of EHE in patients presenting with nonspecific tumours of the extremities. Prompt management and diagnosis are essential in this potential aggressive tumour.

Keywords: Cutaneous epithelioid hemangiodothelioma, Vascular tumour, Skin tumour

INTRODUCTION

Epithelioid hemangiodothelioma (EHE) is a rare vascular tumour first described in 1982 as an intermediate malignancy between hemangioma and angiosarcoma.[1] It usually affects lung, liver, bones, deep soft tissues but very occasionally the skin.

CASE REPORT

A 21-years-old patient, with no medical history, was referred to dermatology for skin tumours of the right lower limb evolving for one year. Lesions stated as small papules gradually developing into tumours. He was amputated of the third toe distal phalanx 3 months earlier for a necrotic lesion with underlying osteolysis. Histology found proliferation of large tumour cell with moderate mitotic activity eroding bone tissue and ulcerating the epidermis. Immunochemistry demonstrated negative cytokeratins, HMB45 and PS100. CD34, CD31, INI-1 and vimentine were positive. The histological report concluded to a low-grade vascular tumor proliferation without being able to determine the exact nature. Skin examination revealed two flesh-coloured nodules on the dorsal side of the second and third toe, and a periungueal necrotic lesion on the fourth toe. On the posterior side of the ankle, there were 2 similar tumours discreetly scaly and pigmented (Figure 1). General status was conserved and no lymphadenopathy was found.

Doppler ultrasonography of the lower extremity arteries and veins as well as right lower limb tomography were normal. Skin biopsies with wide-ranging immunochemical analysis were performed. Histology showed spindle cells, and large epithelioid cells embedded in a myxochondroid matrix displaying nuclear atypia with low mitotic activity. Some tumour cells demonstrated cytoplasmic vacuoles (Figure 2). CD31, CD34, ERG and CAMTA1 staining were all positive. Based on this aspect, diagnosis of cutaneous epithelioid hemangioendothelioma (CEHE) was retained.

Lower limb MRI, positron emission tomography and bones scintigraphy were carried out in search of primary

or secondary extracutaneous localisations, showing no suspect lesion. In accordance with the oncologic staff, it was decided to amputate the second and third toe with the last phalanx of the fourth toe. Histology of the amputation segments showed the same morphologic and immunochemical features as the skin biopsies. No bone involvement was found.

The cutaneous lesion of the ankle was removed with broad margins. A close follow up was scheduled.

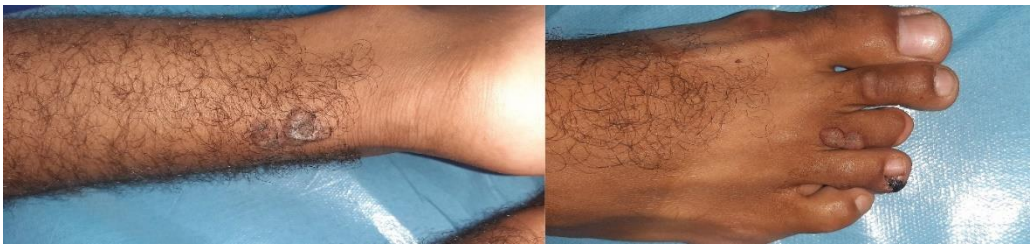

Figure 1: Skin-coloured nonspecific tumours of toes and ankle on the same lower limb.

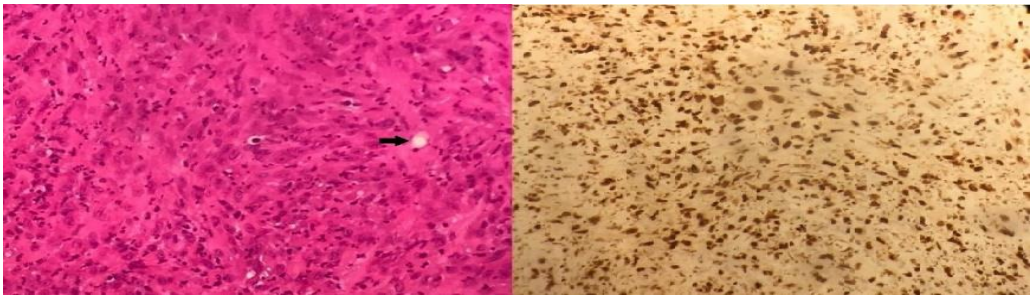

Figure 2: Histological section (HES at 200X) showing spindle cell and epithelioid cell proliferation with nuclear atypia and few mitotic figures, vacuolated tumour cell (arrow), and positive nuclear staining for CAMTA1.

DISCUSSION

EHE may affect all ages but remains rare in childhood.[2] It usually locate to lung, liver, bones, deep soft tissues and very occasionally the skin. Skin-limited EHE are exceptional with only few reports. It typically manifests as a solitary tumour located on the extremities, but may also present as multiple subcutaneous nodules, nonhealing ulcer, scar-like lesion or even a verrucous mass of nose and lips.[3-6] Multiple cutaneous lesions are not common and should raise suspicion for metastatic deep soft tissue or osseous EHE.[2] Multifocal EHE involving the same limb is extremely rare, especially when affecting skin and bones at the same time.[7,8] For our patient, it remains unclear if he displayed skin metastasis of primary osseous EHE of the phalanx or rather multicentric CEHE with secondary involvement of the phalanx bones.

Clinical appearance of skin lesions is often designated as nondescript. Nodules may appear white-grey or skin-coloured not raising suspicion for vascular tumour. Angiocentric tumour proliferation may lead to vessel occlusion occasioning pain, ulceration or necrosis as observed in our patient.[9]

Histology typically shows cords, nests or lobules of epithelioid cells in a myxohyaline stroma. Some spindle cells may occasionally be observed. Vacuolated tumour cells corresponding to attempts of vasoformation are frequent. Mitotic activity and nuclear atypia are usually minimal but may be more significant as in our case presuming an aggressive form. Immunochemistry demonstrates a typical endothelial profile with positivity for CD31, CD34 and ERG. CAMTA1 staining is sensitive and specific to confirm the diagnosis in uncertain cases.[10]

Large surgical excision is the mainstay treatment. Size of the tumour (>3 cm) and mitotic activity are the main risk factors for aggressive course of CEHE.[11]

CONCLUSION

EHE is a rare low-grade vascular tumour that may affect occasionally the skin. Multifocal CEHE is uncommon and should lead to extensive investigations to rule out metastasizing extracutaneous EHE. The treatment of choice is surgical excision with broad margins but close follow up is necessary owing to the potential aggressive course of the tumour.

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