

ACROANGIODERMATITIS IN A LEG AMPUTEE USING A SUCTION-SOCKET PROSTHESIS: CLINICAL, HISTOLOGICAL AS WELL AS HHV-8 AND CD34 IMMUNOHISTOCHEMICAL STUDY

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Acroangiokeratosis (AAD) is a rare, vascular phenomenon of unclear pathogenesis. Itchy, lichenoid, purple/violaceous/yellowish/brownish papules/nodules, plaques/patches mainly on lower limbs occasionally evolve into verrucous lesions and recurrent painful ulcerations. Elevated vein and capillary pressure due to the sub-atmospheric suspension system seems to be the triggering factor for angioproliferation in the amputation stump. A middle-aged male amputee, a suction-socket prosthesis user, showing combined clinical, histological and immunohistochemical (HHV-8 negative; CD34 and CD31 expressed in endothelial, but not perivascular, cells) features of AAD is presented. Dermatologists, orthopedic surgeons, pathomorphologists, but also prosthesis makers and amputees themselves, should be aware of AAD as suction-socket prostheses become increasingly popular.

Key words: acroangiokeratosis, prosthesis, amputation stump.

Introduction

Acroangiokeratosis (pseudo-Kaposi sarcoma, AAD) is a rare, chronic vascular phenomenon of unclear pathogenesis described by Mali *et al.* in 1965 [1, 2]. The disease might be congenital, yet in acquired form chronic venous insufficiency and vascular anomalies seem to be an underlying background for proliferation of pre-existing blood vessels. The AAD was reported in patients with paralyzed limbs [3], hemodialysis patients with arteriovenous shunts [4] or after minor trauma [5] as well as in amputees with suction-socket prostheses [6, 7].

Although vascular hyperplasia, blood extravasation leading to hemosiderin deposition and inflammatory infiltration are features of both AAD and Kaposi sarcoma (KS), both entities remain unrelated, despite the

fact that a relationship might be suspected on the basis of misleading nosology proposed by Earhart *et al.* [2]. Kaposi sarcoma is a progressive cancer of endothelial origin that may imitate AAD clinically and even histopathologically. Regardless of histopathological features that may be utilized to distinguish these angioproliferative disorders (a lobular arrangement of blood vessels with thick walls and “chubby” endothelial cells, absence of cell atypia and lesser inflammatory infiltrate that is generally poor in plasma cells in AAD), immunostaining for the CD34 antigen as well as tissue, most importantly, and blood studies for Kaposi's sarcoma-associated herpesvirus (HHV-8) were adopted to differentiate these entities of significantly distinct prognosis [8-10]. Other methods of distinction may be CD31-immunolabelling, endothelial cell markers

UEA-1 and factor VIII, or radiological imaging (ultrasonography, Doppler ultrasound, magnetic resonance tomography, computed tomography, angiography and phlebography) [4, 11-13]. The first suction-socket prosthesis was invented by Dubois D. Parmelee in 1863. In the fourth decade of the twentieth century that type of prosthesis was reintroduced for war amputees. The prosthesis holds the amputation stump by anatomical adjustment and by suction created by an air-tight socket that prevents rotation. Elevated vein and capillary pressure due to the sub-atmospheric suspension system seems to be the triggering factor for angioproliferation.

We report a case of a male amputee, using a suction-socket prosthesis, with combined clinical, histopathological and immunohistochemical features of AAD.

Case report

A 50-year-old male truck driver presented with itchy blue-purple papulo-erythematous lichenoid lesions on the amputation stump (Fig. 1A). Above-knee amputation was performed a year before due to crushing trauma of the left lower limb. The lesion appeared primarily as an erythematous macule after half-year usage of a suction-socket prosthesis. The patient was consulted by the angiologist two weeks before admission and had Doppler ultrasound of limb vessels performed, finding neither vascular nor blood flow disorder. Laboratory tests were normal (including HBV and HCV testing). Epicutaneous patch tests with European Standard Series and Extended Orthopedic Series (a total of 63 allergens) were performed to rule out allergic contact dermatitis to prosthesis material, yet all were negative at 48 and 72 hours. Histopathology of the lesional skin (hematoxylin and eosin staining) (Fig. 1C) revealed a lobular arrangement of proliferating blood vessels with thick walls and "chubby" endothelial cells, marked erythrocyte extravasation and hemosiderin deposits in the upper half of the dermis with inconspicuous inflammatory cells. Blood study for HHV-8 DNA as well as immunostaining on a paraffin block slide with mouse monoclonal antibody (13B10) against HHV-8 latent nuclear antigen-1 (LNA-1) (Fig. 1D) was negative. The vessels showed regularity and there was neither endothelial nor epithelial atypia present. CD34 (Fig. 1E) and CD31 (Fig. 1F) expression in plump endothelial cells, but not in perivascular cells, in blood vessels of the papillary dermis was revealed with immunohistochemical staining. Thus, the patient presented clinical, histopathological and immunopathological features which considered together enabled the diagnosis of suction-socket prosthesis-associated AAD. The patient was advised to have his suspension type prosthesis replaced with a strap-suspension system retaining other functional modules and treated topically with clobetasol pro-

pionate ointment initially and then tacrolimus ointment. After 6-month follow-up, an improvement of the clinical picture was noted (Fig. 1B).

Discussion

The renaissance of suction-socket prostheses began in the fourth decade of the previous century. The dermatological problems of amputees may be caused by contact dermatitis, poor hygiene of the stump, mechanical microtrauma and, last but not least, the prosthesis suspension mechanism. Despite the embarrassing fart-like sound during readying for use and poor fit (probably due to maladaptation), our patient opted for this suspension type, as it seemed to him comfortable for everyday use. Interestingly, the price of the expensive modular prosthesis bought by the patient was equivalent to about eight mean monthly incomes in Poland at the end of 2011 and Poland's National Health Fund covered just slightly above 10% of the costs. Recently, there has been some debate whether sub-atmospheric socket prostheses can be used in patients with vascular amputation. Production of a thigh-fit socket is difficult artisan-like work as acquiring proper adjustment frequently means repetition of the whole process. Unnecessary air chambers and poor fitting favor impairment of blood supply of the stump that results in bruising/violaceous discoloration. In the case reported here, the prosthesis using the sub-atmospheric suspension system plausibly led to development of AAD by evoking superficial venous stasis in the amputation stump, resulting in pathological reactive angiogenesis and fibrosis.

In the context of AAD ruling out KS is essential since both entities have different prognosis and treatment methods. Obviously, the expression of CD34 and CD31 in endothelial cells of blood vessels is not an extraordinary finding; nevertheless, the expression pattern of these proteins [14] considered not separately but in conjunction with remaining clinical (particularly improvement of lesions after prosthesis change), microscopic and laboratory data, together with tissue and blood HHV-8 evaluation [15], enabled the diagnosis of AAD in the case presented here.

Over 100 cases of AAD have been described in adults, making this disease rare, yet not exceptional. Nonetheless, the association with suction-socket prostheses is mentioned in just a tiny fraction of published AAD cases, making such an association still infrequent. As some authors recommend suction-socket prostheses also in children as young as 5 years of age [16], it seems imaginable that AAD lesions might appear in both pre-adolescent and adolescent amputees as the child's growing body should be taken into consideration. In both children and adults neovascularization is mediated by intertwined molecular pathways mediated by VEGF, bFGF, angiopoietins, ephrins, and class 3

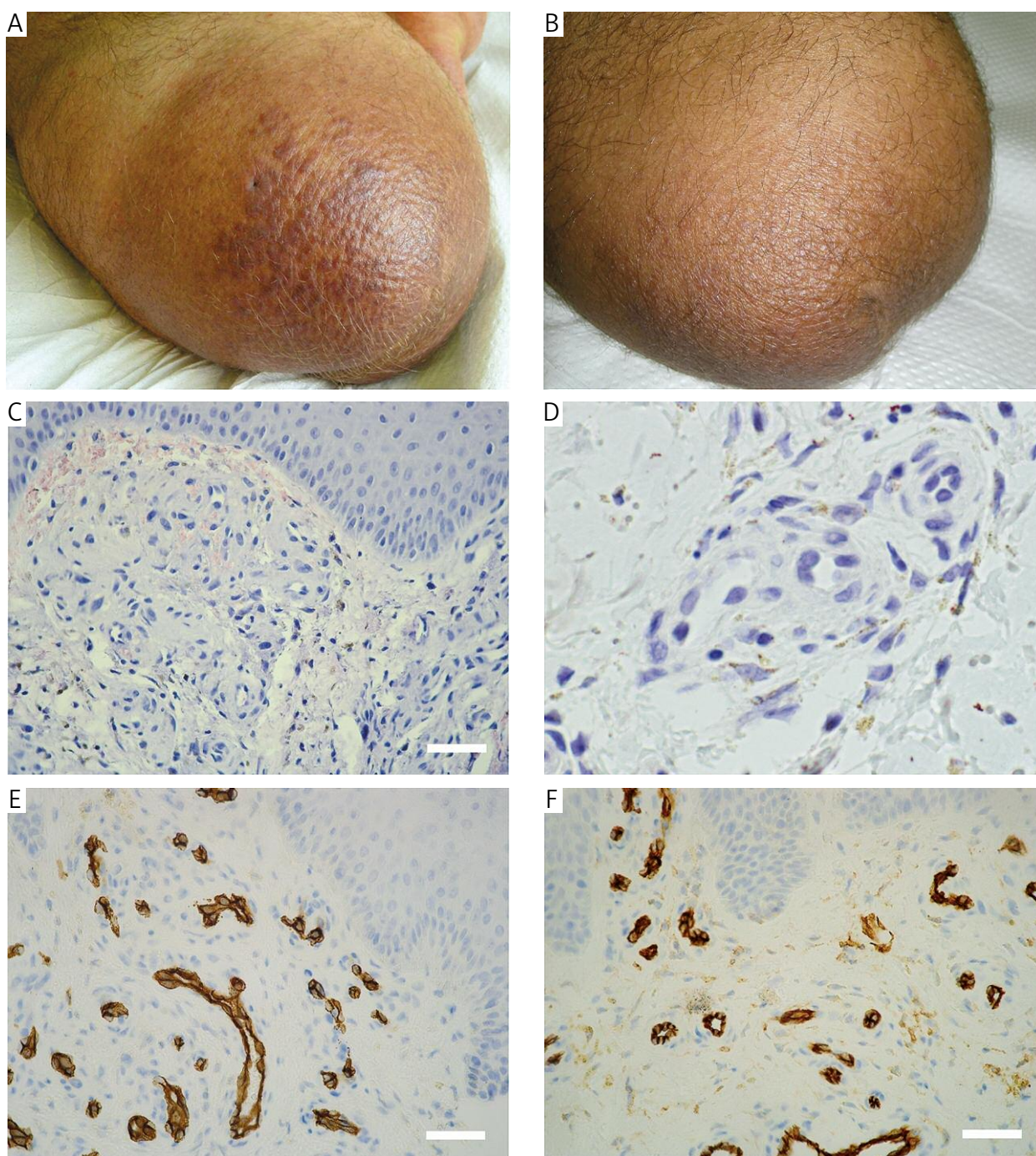


Fig. 1. Violaceous/brownish lichenoid lesions on the amputation stump of the leg. Biopsy site on the medial/frontal aspect of the thigh. The patient was referred by Leszek Bartoszak MD/PhD (A). Remission of stump lesions after 6-month follow-up period (B). Proliferation of blood vessels, in lobular pattern, with thick walls and “chubby” endothelial cells and marked erythrocyte extravasation and hemosiderin deposits in the papillary dermis without either endothelial or epithelial atypia and inconspicuous inflammatory infiltrate (HE staining, original magnification 400 \times , bar = 50 μ m) (C). Immunohistochemical evaluation for HHV-8 – negative result (mouse monoclonal antibody (13B10) against HHV-8 latent nuclear antigen-1, LNA-1, primary objective magnification 40 \times) (D). CD34 (E) and CD31 (F) expression in “chubby” endothelial cells, but not in perivascular cells, in blood vessels of the papillary dermis (immunohistochemical staining, original magnifications 400 \times , bar = 50 μ m)

semaphorins [17], which conceivably might be targets for AAD-oriented therapeutic intervention.

As realities in former Soviet bloc countries were unfavorable for both international scientific exchange and

availability of foreign language dermatology textbooks, AAD was neglected in Polish dermatological literature until the end of the 20th century [18, 19]. Hence, middle-aged and elderly Polish dermatologists who had ac-

cess to just native literature might misdiagnose and presumably underdiagnose AAD. Ironically, many international journals continue to require purchasing online access, and open access is still uncommon, making accessibility to professional knowledge difficult even nowadays, especially for junior physicians. Thus, this case report was conceived within a framework of the continuing medical self-education to increase awareness of AAD among dermatologists, orthopedic surgeons, pathomorphologists, but also prosthesis makers and amputees themselves, regardless of their age and nationality, as suction-socket prostheses become increasingly popular.

The authors declare no conflict of interest.

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