Acroangiodermatitis (Pseudo-Kaposi Sarcoma) from Chronic Congestive Cardiac Failure Secondary to Dilated Cardiomyopathy in a Nigerian: A Case Report

Adekanmbi O.A[1], Ayanlowo O[1], Akinkugbe A[1], Akinde R[2], Otrofanowei E [1], Okesola O[1]

[1] Department of Medicine, Lagos University Teaching Hospital , Idi-Araba, Lagos

[2] Department of Morbid Anatomy, Lagos University Teaching Hospital, Idi Araba, Lagos

INTRODUCTION

Acroangiodermatitis (pseudokaposi sarcoma) is an uncommon benign disease of the blood vessels, characterized by a proliferation of pre-existing small blood vessels. The term was first used by Mali et al, who described their findings of mauve-coloured macules and plaques on the extensor surfaces of the feet of 18 patients with chronic venous insufficiency. Since then it has been described in a number of clinical conditions including vascular malformations such as Klippel-Trenaunay syndrome, Prader-Willi Syndrome and Stewart-Bluefarb synfrome; presence of iatrogenic AV-fistulas; amputees and patients with paralyzed legs; and in association with Hepatitis C virus. It is clinically characterized by the finding of violaceous macules, papules, and nodules, lesions which can ulcerate and cause bleeding, on the extensor surfaces of the distal lower limbs. These lesions are similar to those seen in Kaposi sarcoma which is a close differential, and can be difficult to differentiate clinically.

It is important to recognize this condition and exclude Kaposi sarcoma. Hence, this report aims to highlight the clinical and histologic features of acroangiodermatitis and differentiate this from Kaposi Sarcoma.

CASE

A 56 year old man with biventricular failure secondary to dilated cardiomyopathy presented with bilateral leg swelling up to the knees, with associated hyperpigmentation on lower limbs; stasis dermatitis; violaceous nodules, plaques and papules on the anterior part of the leg and the lateral malleolus bilaterally (Figure 1). An initial diagnosis of Kaposi sarcoma was entertained.

The nodules around the shin were biopsied and histology showed numerous small vessels in the deep dermis with deposition of red blood cells. The blood vessels were lined by endothelial cells and there were lymphoplasmacytic infiltrates consistent with acroangiodermatitis (Figures 2-4). His blood parameters were essentially normal and HIV test was negative. He was placed on erythromycin and asked to limit his mobility and keep his feet elevated as much as possible. At his next visit one month later, the lesions improved significantly as the nodules were flatter and the stasis dermatitis was significantly healed and not discharging. He was however admitted about two months later because his heart failure condition progressed with no identifiable precipitant and he died of cardiogenic shock complicating the heart failure while on admission.

DISCUSSION

Acroangiodermatitis (AAD) is a benign condition easily confused with Kaposi sarcoma as they share certain clinical and histological features. Furthermore, it is often misdiagnosed because it is not commonly seen : less than hundred cases have been reported worldwide. Lesions of AAD usually occur bilaterally on the lower limbs when due to chronic venous insufficiency, but can be unilateral, or occur elsewhere when due to other causes as exemplified in patients with arteriovenous fistula undergoing hemodialysis in whom lesions can involve the upper limbs. Four types have been described: the Stewart-Bluefarb type accompanying chronic venous malformations, the Mali type accompanying stasis dermatitis, a type accompanying first gestation, and a type accompanying arteriovenous shunts in patients with chronic kidney disease. The Stewart-Bluefarb type is usually unilateral and starts at an early age. The Mali type is seen in elderly patients and is usually bilateral, occurring due to failure of the venous pump of the lower limbs. The patient presented is most likely the Mali type. Acroangiodermatitis can also be confused with lymphangiosarcoma and basal cell carcinoma.

The HIV epidemic and associated epidemic Kaposi Sarcoma has made Kaposi Sarcoma an important differential of AAD. The usual violaceous plaques seen in AAD are similar to those seen in KS and these two may be difficult to distinguish clinically. However,

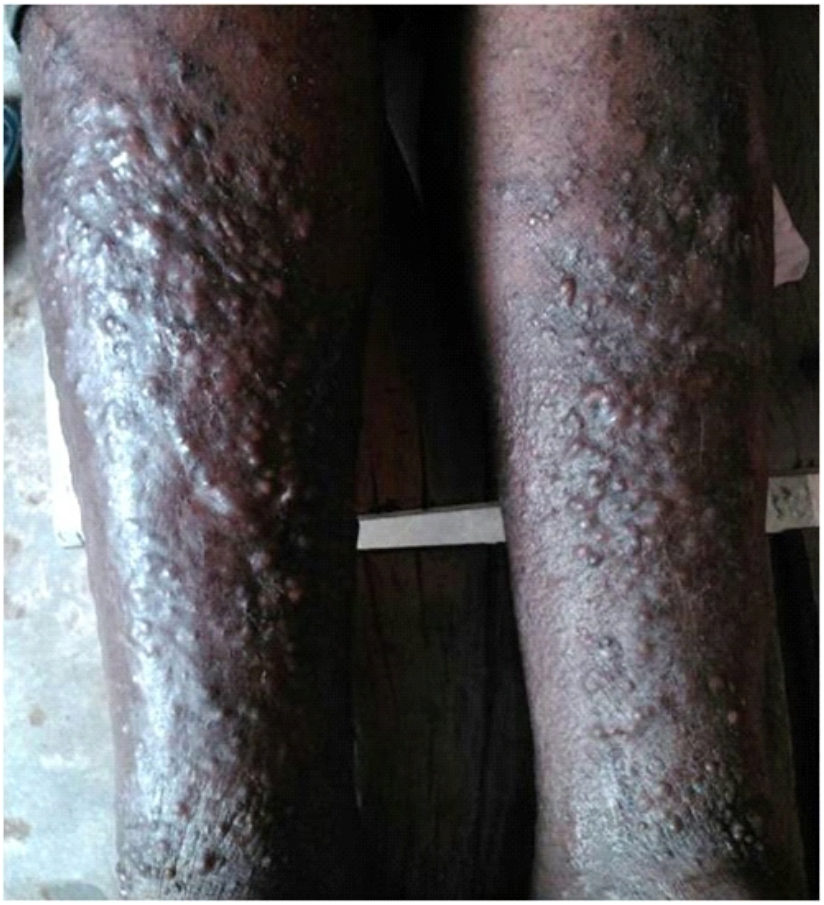

Figure 1: Hyperpigmented nodules in the anterior aspect of both lower limbs of patient with acroangiodermatitis

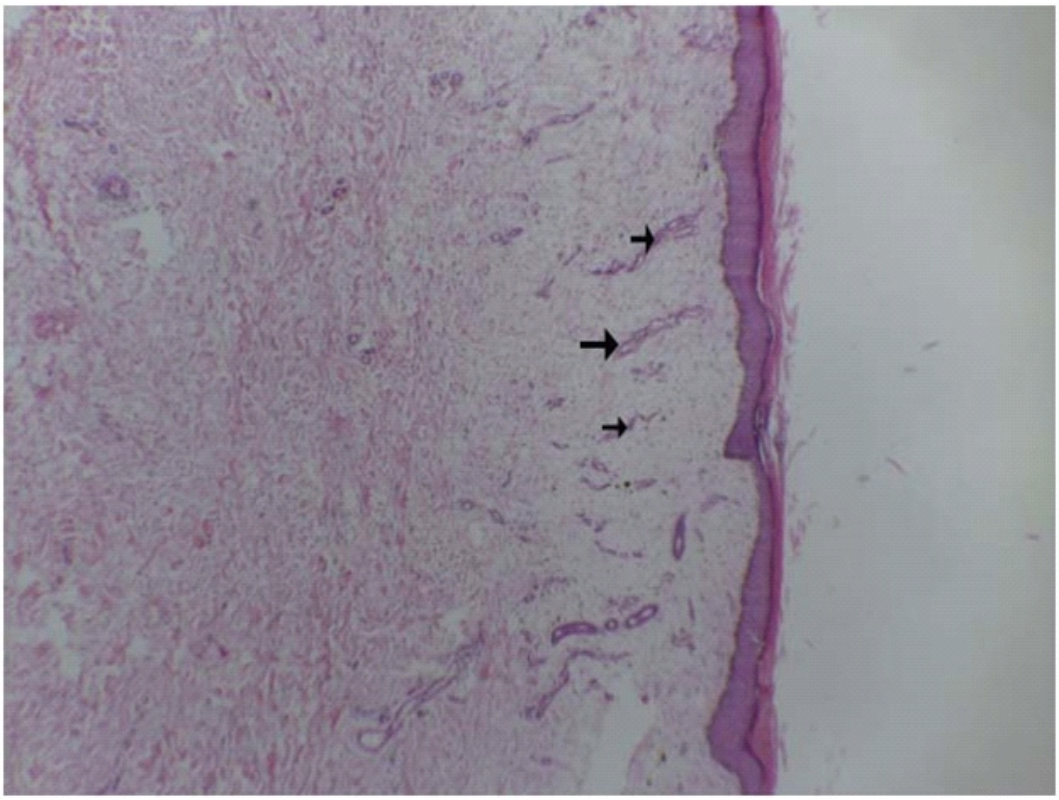

Figure 2: Proliferating thin walled blood vessels within the papillary dermis. The stroma is loose and oedematous (H&E x40).

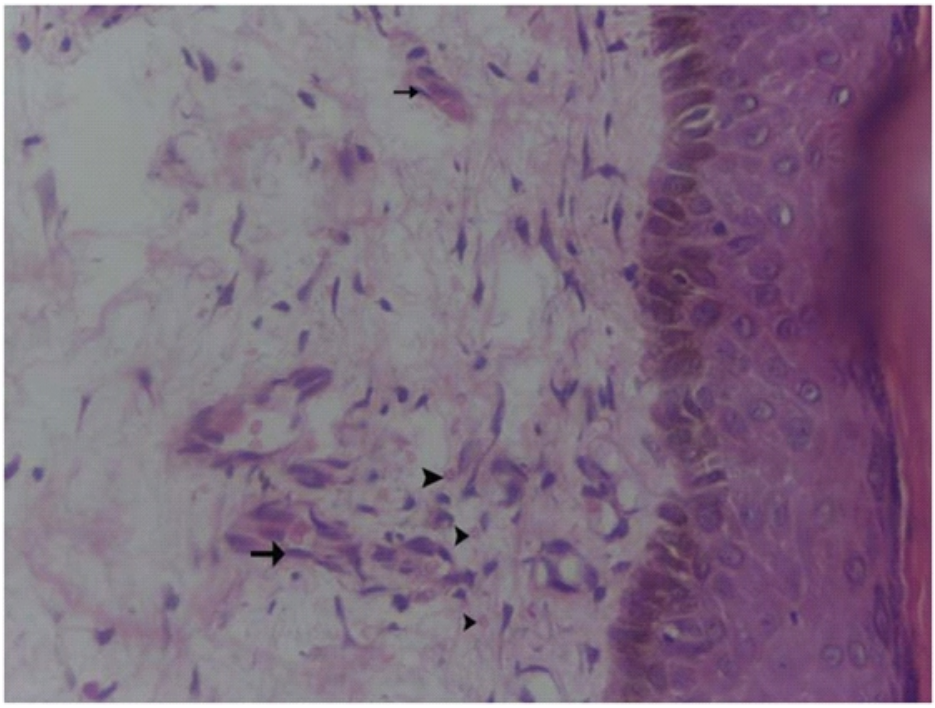

Figure 3: Higher power showing proliferating vessels (arrows) and oedematous stroma with sprinkling of chronic inflammatory cells (curved arrows) (H&E x100).

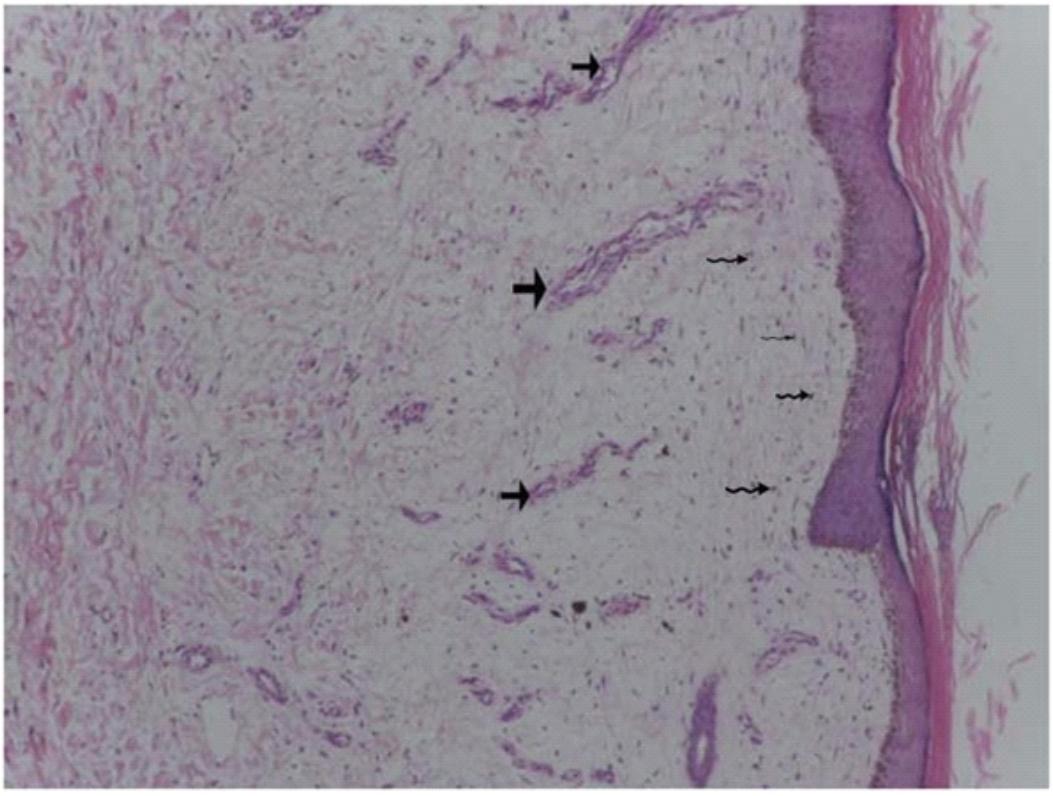

Figure 4: Blood vessels lined by flattened endothelial cells (arrows) with extravasated red blood cells (arrow heads) within the oedematous stroma (H&E x400)

lesions in KS can involve other body areas not usual for AAD. An important distinguishing feature is the mucosal involvement which occurs in KS.

Histology of AAD lesions reveal marked proliferation of vessels in the dermis, mild edema, hemosiderin deposition, fibrosis, areas of patchy lymphocytic infiltrates, and fibrosis. Vessels are usually dilated, regular and lack atypia. On the other hand, histology of KS shows vascular slits, proliferation of atypical cells and vascular hyperplasia independent of existing vasculature.. There may however be histologic similarities between AAD and KS, a situation that may warrant the use of imunohistochemistry with labeling of CD34, a cell adhesion molecule present in endothelial cells of high endothelial venules. CD34 binds to CD62L which is expressed on lymphocytes, monocytes, and granulocytes. Immunopathologic analysis of KS lesions shows CD34 positivity in both endothelial and perivascular cells while in AAD it shows CD34 positivity in endothelial cells of hyperplastic vessels but negativity in perivascular regions . Also, using antibody active against latent nuclear antigen (LNA-1) of the human herpes virus 8 can detect the presence of herpes virus in Kaposi sarcoma tumour cells, thus differentiating Kaposi sarcoma from its histological variants. In cases where clinical features and histology are inconclusive , electron microscopy has a role where signs of degeneration of vascular walls and infiltrating cells are demonstrated in KS.

Treatment for AAD can be conservative or surgical. Success has been reported with the use of erythromycin and dapsone, as well as with cryotherapy and intermittent pneumatic compression therapy. The patient reported was treated with a course of erythromycin and this resulted in improvement of the lesions. Acroangiodermatitis is not primarily associated with death; however, death could occur from the underlying clinical condition as occurred in our patient.

It is important to diagnose acroangiodermatitis, a benign lesion, early and differentiate it from Kaposi sarcoma. This is because appropriate treatment for AAD will lead to regression of lesions; whereas, incorrect therapies can further lead to complications from chemotherapy. Histology and immunohistochemistry are important investigations for diagnosis.

REFERENCES

1. Mali JW, Kuiper JP, Hamers AA. Acro-angiodermatitis of the foot. Arch Dermatol [Internet]. 1965 Nov [cited 2016 Jan 30];92(5):515–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/5844394
2. Lyle WG, Given KS. Acroangiodermatitis (pseudo-Kaposi's sarcoma) associated with Klippel-Trénaunay syndrome. Ann Plast Surg [Internet]. 1996 Dec [cited 2016 Jan 30];37(6):654–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/8988782
3. Wei-Min Tan A, Lee JS-S, Pramono ZAD, Chong W-S. Acroangiodermatitis of Mali in protein C deficiency due to a novel PROC gene mutation. Am J Dermatopathol [Internet]. 2012 Apr [cited 2016 Jan 30];34(2):e19–21. Available from: http://www.ncbi.nlm.nih.gov/pubmed/22441373
4. Güçlüer H, Gürbüz O, Kotiloglu E. Kaposi-like acroangiodermatitis in an amputee. Br J Dermatol [Internet]. 1999 Aug [cited 2016 Jan 30];141(2):380–1. Available from: http://www.ncbi.nlm.nih.gov/pubmed/10468837
5. Bernardes Filho F, Martins G, Nery JA da C, Andrade CV de, Kac BK. Acroangiodermatitis (pseudo-Kaposi's sarcoma) in an HIV sero-positive patient with syphilis and hepatitis C virus coinfection: clinical and dermatopathological features. An Bras Dermatol [Internet]. Jan [cited 2016 Jan 30];89(5):792–5. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=4155958&tool=pmcentrez&rendertype=abstract
6. Singh SK, Manchanda K. Acroangiodermatitis (Pseudo-Kaposi sarcoma). Indian Dermatol Online J [Internet]. 2014 Jul [cited 2016 Jan 30];5(3):323–5. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=4144224&tool=pmcentrez&rendertype=abstract
7. Varyani N, Thukral A, Kumar N, Gupta KK, Tandon R, Tripathi K. Nonhealing ulcer: acroangiodermatitis of mali. Case Rep Dermatol Med [Internet]. 2011 Jan [cited 2016 Jan 30];2011:909383. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=3504217&tool=pmcentrez&rendertype=abstract
8. Pimentel MIF, Cuzzi T, Azeredo-Coutinho RBG de, Vasconcellos É de CFE, Benzi TSCG, Carvalho LMV de. Acroangiodermatitis (pseudo-Kaposi sarcoma): a rarely-recognized condition. A case on the plantar aspect of the foot associated with chronic venous insufficiency. An Bras Dermatol [Internet]. Jan [cited 2016 Jan 30];86(4 Suppl 1):S13–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/22068760
9. Mehta AA, Pereira RR, Nayak CS, Dhurat RS. Acroangiodermatitis of mali: a rare vascular phenomenon. Indian J Dermatol Venereol Leprol [Internet]. Jan [cited 2016 Jan

30];76(5):553–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20826998

10. Strutton G, Weedon D. Acro-angiodermatitis. A simulant of Kaposi's sarcoma. Am J Dermatopathol [Internet]. 1987 Apr [cited 2016 Jan 30];9(2):85–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/3504113

11. Bulat V, Lugovic L et al. Acroangiodermatitis (Pseudo-Kaposi Sarcoma) as part of chronic venous insufficiency. Acta Clin Croat 2007; 46: 273-277 - Google Search [Internet]. [cited 2016 Jan 30]. Available from: https://www.google.com.ng/?gfe_rd=cr&ei=Ua2sVrLAMu7U8geu75Bg&gws_rd=ssl#q=11.%09Bulat+V%2C+Lugovic+L+et+al.+Acroangiodermatitis+(Pseudo-Kaposi+Sarcoma)+as+part+of+chronic+venous+insufficiency.+Acta+Clin+Croat+2007%3B+46:+273-277

12. Lugović L, Pusić J, Situm M, Buljan M, Bulat V, Sebetić K, et al. Acroangiodermatitis (pseudo-Kaposi sarcoma): three case reports. Acta Dermatovenerol Croat [Internet]. 2007 Jan [cited 2016 Jan 30];15(3):152–7. Available from: www.ncbi.nlm.nih.gov/pubmed/17868541

13. Pereira PF, Cuzzi T, Galhardo MC. Immunohistochemical detection of the latent nuclear antigen-1 of the human herpesvitus type 8 to differentiate cutaneous epidemic Kaposi sarcoma and its histological simulators. An Bras Dermatol 2013; 88; 243-246 - Go [Internet]. [cited 2016 Jan 30]. Available from: https://www.google.com.ng/?gfe_rd=cr&ei=Ua2sVrLAMu7U8geu75Bg&gws_rd=ssl#q=13.%09Pereira+PF%2C+Cuzzi+T%2C+Galhardo+MC.+Immunohistochemical+detection+of+the+latent+nuclear+antigen-1+of+the+human+herpesvitus+type+8+to+differentiate+cutaneous+epidemic+Kaposi+sarcoma+and+its+histological+simulators.+An+Bras+Dermatol+2013%3B+88%3B+243-246

14. Samad A, Dodds S. Acroangiodermatitis: review of the literature and report of a case associated with symmetrical foot ulcers. Eur J Vasc Endovasc Surg [Internet]. 2002 Dec [cited 2016 Jan 30];24(6):558–60. Available from: www.ncbi.nlm.nih.gov/pubmed/12443758

15. Grayson W, Pantanowitz L. Histological variants of cutaneous Kaposi sarcoma. Diagn Pathol [Internet]. 2008 Jan [cited 2016 Jan 30];3:31. Available from: http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2526984&tool=pmcentrez&rendertype=abstract

16. Secher L, Weismann K, Kobayasi T. Pseudo-Kaposi sarcoma of the feet: an electron microscopic investigation. Acta Derm Venereol [Internet]. 1984 Jan [cited 2016 Jan 30];64(3):246–9. Available from: http://www.ncbi.nlm.nih.gov/pubmed/6204488