Journal of Pakistan Association of Dermatologists. 2023;**33(4)**:1717-1720.

Case Report

The tale of rare skin condition of the breast: A case series

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Abstract In clinical practice, one may come across clinical conditions, the diagnosis of which, is either overlooked or missed because it is rare so it is not considered by the treating dermatologist or it present with unusual modes regarding the presenting odd features, site affected, patient's age, predominant sex and time of occurrence, so that, all these points should be considered in order to pick up the proper diagnosis. The ingenuity in clinical practice lies in the diagnosis of challenging unusual cases. Herein we present a case series of female patients presenting with chronic odd skin breast lesions with poor response to treatments to increase the awareness of the dermatologist to such rare condition. To the best of our knowledge, this series is the first one in our country and the region and the fifth worldwide.

Key words
Breast; Rare condition; Unusual.

Introduction

Diffuse dermal angiomatosis (DDA) is a rare, benign, reactive diffuse angioendotheliomatosis to many associated risk factors like obesity, smoking, underlying vaso-occlusion, and hypercoagulability, pressure induced trauma and hypoxia to the underlying fatty tissue.[1] Krell *et al.* first observed it at 1994.[2] The lower extremities are the site of predilection where there was six cases reported previously in literature at the extremities. DDA is presented clinically as erythematous to violecious reticulated plaques with ulcers and secondary infection at some areas.[3-5] Rarely, DDA may affect the breast especially in obese female with pendulous breasts.[6]

Herein we present a case series of female patients presenting with breasts DDA with poor response to treatments to increase the awareness of the dermatologist to such condition. To the best of our knowledge, this series is the first one in our country and the region and the fifth worldwide.

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Case 1

A 43- year- old female patient presented with chronic tender skin lesions on right breast with ulcers for 1 year, which was chronic, non-healing, and progressive with time. By history, she was healthy, non-smoker, no chronic drug use with negative family history of the same condition. On examination, the patient looked normal, well, and obese (BMI 33) with large pendulous breasts. All vital signs were normal. The skin examination exhibited tender violecious reticulated nonblanchable plaques with multiple (three) ulcers on inner medial aspect of the right breast **(Figure 1)** with no skin lesions on the other breast or the rest of the body. All the relevant investigations were within normal. Skin biopsy showed diffuse proliferation of capillary vessels in superficial

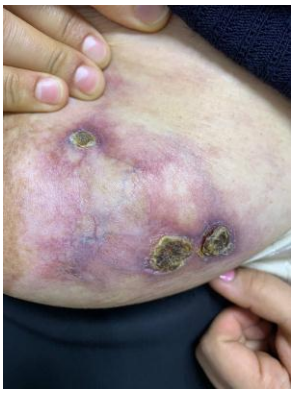


Figure 1 reticulated erythematous to violecious plaque with three ulcers on the middle inner surface of the right breast of fist case (author's photo).

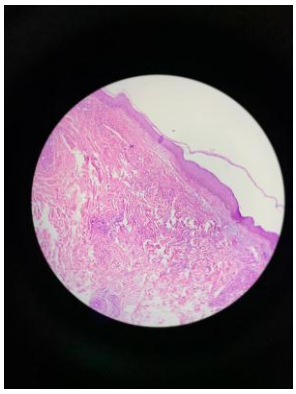


Figure 2

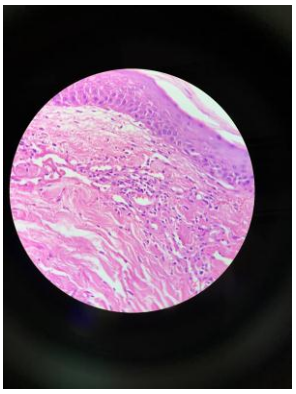


Figure 3

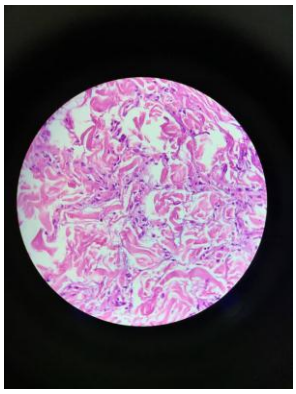


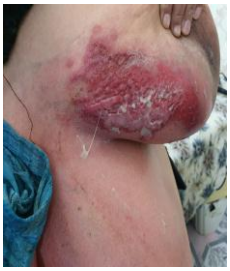
Figure 4

Figure 2-4 histopathotogy of the first case, showed diffuse proliferation of capillary vessels in superficial and deep dermis in between the collagen bundles with dense proliferation of endothelial cells without nuclear atypia (author's photo).

and deep dermis in between the collagen bundles with dense proliferation of endothelial cells without nuclear atypia **(Figures 2-4)**, which is consistent with diffuse dermal angiomatosis. The CD31 immunostaining was positive. The test for herpes simplex virus 8 was negative. The angiogram for both subclavian arteries were normal. The patients was advised to reduce weight and change life style(do regular exercise, avoid passive smoking, avoid high caloric food) and informed that mastoplasty is the treatment of choice, meanwhile she was kept on isotretinoin 10mg per day with topical steroid plus antibiotics with good results

Case 2

A 38- year-old female patient came to the private clinic complaining from chronic non-healing bilateral breast ulcers with draining pus for 10 months. She was healthy, non-smoker with negative past medical and drug history with negative family history of the same condition. On examination, the patient was well, healthy, and obese (BMI 31) with large pendulous breasts with normal vital signs. The skin of the breasts showed erythematous indurated plaques with cribriform ulcers with yellowish pus bilaterally on the inner medial aspects of both breasts **(Figure 5,6)** with no lesions elsewhere in the body. All the relevant investigations were within normal. Skin biopsy showed diffuse proliferation of capillary vessels in superficial and deep dermis in between the collagen bundles with dense proliferation of endothelial cells without nuclear atypia **(Figures 7-9)**, which is consistent with diffuse dermal angiomatosis.



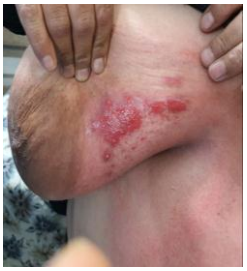
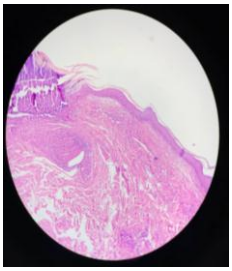
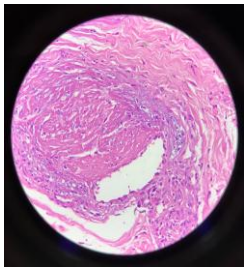


Figure 5,6 Erythematous plaque with cribriform ulcer and draining pus at the inner middle surface of the left breast of second case (author's photo).





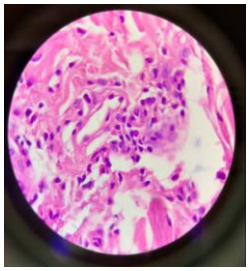


Figure 7-9 Histopathology of the second case, , showed diffuse proliferation of capillary vessels in superficial and deep dermis in between the collagen bundles with dense proliferation of endothelial cells without nuclear atypia (author's photo).

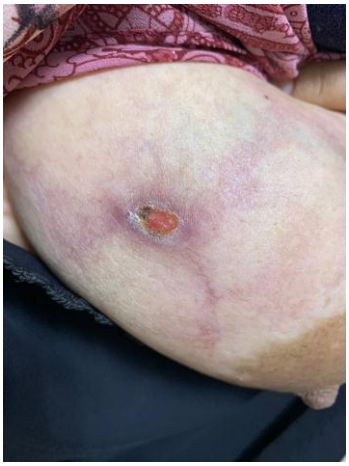


Figure 10 reticulated erythematous to violecious plaque with ulcer on the middle inner surface of the left breast of third case (author's photo).

The CD31 immunostaining was positive. The test for herpes simplex virus 8 was negative. The angiogram for both subclavian arteries were normal. The patient was treated as the treatment plan in the previous case with addition of systemic antibiotics and topical drying agent.

Case 3

A 46 - year-old female patient presented with tender chronic recurrent breast ulcer on the left breast for 1.5 year with poor response to treatment and extension with time. The patient had no chronic diseases or chronic drug use, but she was smoker with negative family history of the same condition. On examination, the patient looked in good health, obese (BMI 35) with large pendulous breasts. All vital signs were normal. The skin examination revealed tender violecious reticulated nonblanchable plaques with small ulcer on inner medial aspect of the left breast **(Figure 10)** with no skin lesions on the other breast or the rest of the body. All the relevant investigations were within normal. Skin biopsy showed diffuse proliferation of capillary vessels in superficial and deep dermis in between the collagen bundles with dense proliferation of endothelial cells without nuclear atypia, which is consistent with diffuse dermal angiomatosis. The CD31 immunostaining was positive. The test for herpes simplex virus 8 was negative. The angiogram for both subclavian arteries were normal .The patients was advised to reduce weight and change life style(do regular exercise, avoid smoking, avoid high caloric food) and informed that mastoplasty is the treatment of choice, meanwhile she was kept on isotretinoin 10mg per day with topical steroid plus antibiotics with good results.

Discussion

DDA is rare reactive, benign inflammatory angioendotheliomatosis that is mostly affecting the lower extremities particularly in those who are suffering from peripheral vascular diseases, hypercoagulable state like anti-phospholipid syndrome, atherosclerosis.[1,7,8] There were only six reported cases of peripheral DDA worldwide. On the other hand, until now, only three cases of breast DDA were reported all over the world, which means that this condition is possibly underestimated, overlooked or really is a rare condition. Breast DDA mostly affects middle-aged obese females with large pendulous breasts.[1] Other risk factors include subclavian artery stenosis, smoking, hyercoagulable state arteriovenous fistula and calciphylaxis.[1,7-9] It may be clinically inflamed, irritated or secondarily infected such as in our second case.[1] Although, the exact pathogenesis of DDA is not well understood, but it is thought that obesity (high BMI) with large pendulous breast lead to chronic focal repeated compression, torsion and pressure effect on deep fat breast tissue resulting in hypoxia with reactive new blood vessels formation in response to up-regulation of vascular endothelial growth factor, which is the basic pathology of this disease.[1,5] Moreover, many factors affect wound healing process like smoking,[7] pressure effect of the large size breast that acts not only as a causative agent, but also as risk factor in persistent delayed wound

healing in addition to the occlusion of the folded area by the bra. The histopathology of DDA is a proliferation of endothelial cells and capillaries between collagen bundles within whole layers of the dermis.[10] Immunostaining CD31, CD34, and ERG are positive for DDA, while Human herpes virus 8 testing is negative.[11,12] The curable treatment of DDA is directed toward correction of the risk factors, excision of the lesion and reduction mammoplasty.[6,13] Other medical treatments like isotretinoin, aspirin, systemic steroid and pentoxifylline has been used with variable results.[5,14] DDA may resolved spontaneously. In our series, the patients were completely normal with normal all relevant investigations. The histopathology was consistent with DDA, (CD31) immunostaining was positive; they were treated with isotretinoin with partial improvement.

Conclusions

DDA should be considered in any obese female with large pendulous breast particularly those running chronic course with poor response to treatments, in order to avoid misdiagnosis or underestimation of such rare condition.

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