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Telangiectasias of the breasts showing diffuse dermal angiomatosis in a patient with diffuse livedo reticularis

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Abstract

The breasts are a common location for diffuse dermal angiomatosis (DDA) in a context of obesity and macromastia. The typical clinical presentation includes erythematous or purplish plaques, reticulated telangiectasias, and sometimes livedo reticularis, often complicated by painful ulcerations of the breasts. Biopsy usually confirms a dermal proliferation of endothelial cells staining positively for CD31, CD34 and SMA α and negatively for HHV8. We report herein a woman with DDA of the breasts presenting as diffuse livedo reticularis and acrocyanosis, both long-standing and considered idiopathic following extensive investigations. Since a biopsy of the livedo did not document DDA features in our case, we suggest that our patient's livedo reticularis and telangiectasias could constitute a vascular predisposition for DDA, as its pathogenesis frequently involves an underlying disease involving ischemia, hypoxia, or hypercoagulability.

Keywords: livedo, proliferative disorder, telangiectasia, vascular

Introduction

Diffuse dermal angiomatosis (DDA) is a subtype of cutaneous reactive vascular proliferation [1,2], most often located on the limbs in a context of atherosclerosis. Diffuse dermal angiomatosis of the breast was first described in 2001 [3]. Since then, numerous cases have been reported, making the

breasts one of the most common locations for DDA, with a total of 38 cases in a review published in 2017 [4], and a few reports since.

On the breasts, DDA presents clinically as erythematous or purplish patches or plaques, surrounded by reticulated telangiectasias and sometimes by localized livedo reticularis. Painful ulcerations can occur as a complication. Concerning DDA of the breasts, patients are mostly in their 50s, and tend to be overweight with large, pendulous breasts (in 75 % of the cases) with a frequent history of breast reduction surgery [4]. Several comorbidities have been noted: coronary artery disease, hypertension, hypercoagulation states (monoclonal gammopathy, anticardiolipin antibodies), and vascular abnormalities (Takayasu arteritis [5]).

Histological analyses typically show an extravascular proliferation of spindle-shaped endothelial cells in the reticular dermis, forming small capillary-like neovessels. Intravascular proliferation is infrequent. Cells have a vacuolated cytoplasm but do not exhibit cytological atypia or mitoses. Immunohistochemistry is positive for CD31, CD34, and ERG. Another characteristic is the presence of α SMA-positive pericytes [2,6]. Human herpes virus type 8 (HHV8) and D2-40 staining are negative, unlike what is observed in Kaposi sarcoma, which shows positivity for HHV8, D2-40, CD34, and CD 31.

There is no consensus for therapeutic strategy: the removal of a hypothetical predisposing factor is sometimes efficacious. In DDA of the breasts, breast

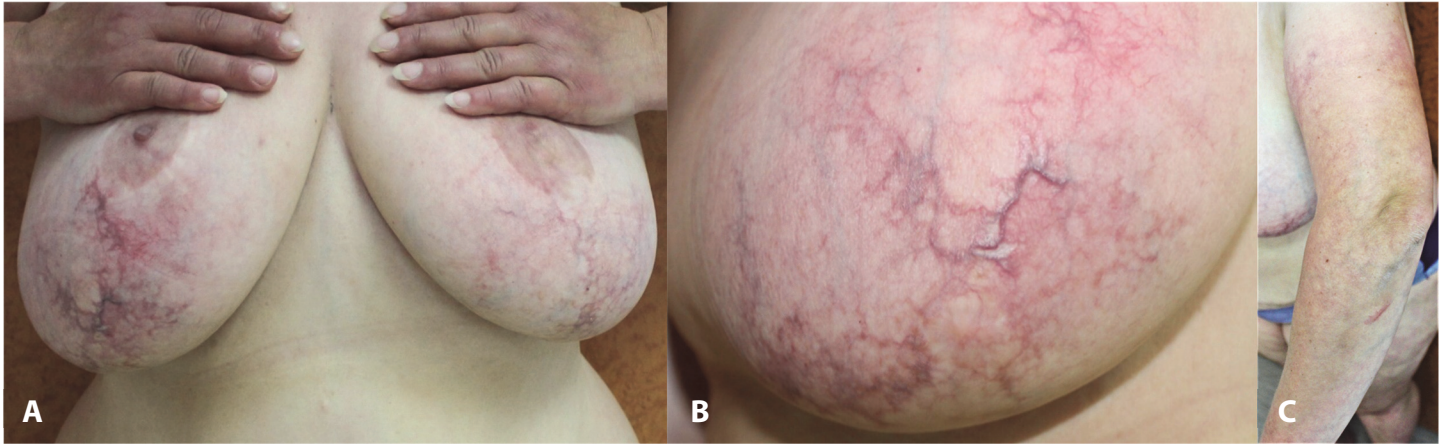


Figure 1. Clinical features of the case: **A, B)** bilateral telangiectasias of the breasts associated with **C)** diffuse livedo reticularis.

reduction surgery has been fully efficacious in 5 cases [4]. Isotretinoin is sometimes successful [3], but with frequent relapses following discontinuation.

Case Synopsis

A 51-year-old-woman presented with a four-year history of bilateral telangiectasias of the breasts. She underwent breast reduction surgery nine years earlier. Three years previously, she presented with a spontaneous non-healing ulceration of the left breast, which resolved only after surgical debridement. A clinical examination showed large pendulous breasts with bilateral telangiectasias on the inferior quadrants (**Figures 1A, B**). She also exhibited diffuse physiological livedo reticularis (**Figure 1C**) and acrocyanosis, both present since her adolescence and unchanged since then. Her work in a food-processing factory required daily exposure to cold and she has been smoking 10 cigarettes a day since her adolescence.

Exhaustive biological investigations, including serum protein electrophoresis, cryoglobulinemia, anti-nuclear antibodies, and antiphospholipid panel did not evidence any abnormalities. Transthoracic ultrasound and chest X-ray investigations were normal. Nail capillaroscopy showed only minor capillary dystrophia.

A skin biopsy of telangiectasias on the right breast evidenced a proliferation of endothelial cells with moderate hemorrhagic suffusions clustered in cellular areas in the papillary and reticular dermis, forming small vascular channels (**Figure 2A, B**). The nuclei were oval and regular without mitoses. Immunohistochemistry investigations showed that the cells were Erg positive, CD31 and CD34 positive, and HHV8 negative (**Figure 2C**). Actin-positive myopericytes were also found. The histological examination of the livedo pattern skin biopsy was normal.

On the basis of this clinical-pathological correlation, dermal angiomatosis of the breasts was diagnosed.

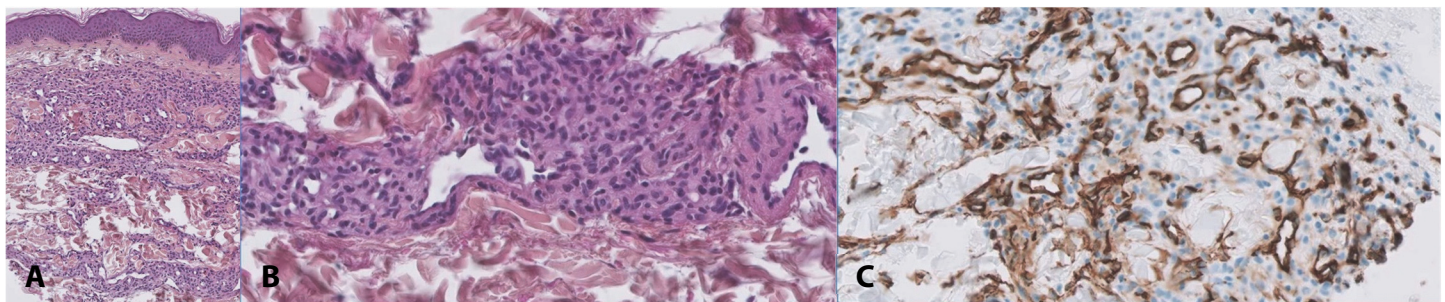


Figure 2. Vertical sections of 4mm punch biopsies taken from the right breast. **A, B)** Proliferation of endothelial cells with regular nuclei, forming small vascular channels across the entire thickness of the dermis, dissociating collagen bundles. H&E, **A)** 100 \times ; **B)** 400 \times . **C)** Positive CD31 staining, 400 \times .

The livedo reticularis and the acrocyanosis were considered idiopathic given the normality of the para-clinical examinations.

Case Discussion

Histological differences between DDA and other cutaneous reactive angiomatoses, especially reactive angioendotheliomatosis, are quite subtle and positive diagnoses rely on clinical-pathological consistency. Hemorrhagic suffusions, hemosiderin deposits, diffusion across the whole thickness of the dermis, and association with inflammatory infiltrates, as in our case, give evidence in favor of DDA, rather than angioendotheliomatosis [1]. Localization on the breasts is typical of DDA and this led us to diagnose DDA clinically in our case.

Diffuse dermal angiomatosis pathogenesis is as yet poorly understood and remains hypothetical. Local hypoxia due to ischemia appears to be a key factor initiating neo-angiogenesis and endothelial proliferation, whereas cardiovascular risk factors, obesity, and smoking could lead to general endothelial inflammation. The increased venous hydrostatic pressure due to macromastia in DDA of the breasts could lead to local hypoxia. The traumatic factor of breast reduction surgery could also be involved [4].

Interestingly, an association between diffuse livedo reticularis, DDA of the breast, and acrocyanosis, as found in our patient, has hitherto not been described in the literature. It raises the issue of the existence of an underlying microcirculation disorder, which in our case remained unidentified, despite exhaustive investigations.

To our knowledge, the association between livedo reticularis and DDA has mostly been described locally in certain cases of DDA of the breasts [6], DDA in a context of arteriovenous fistula [7], and DDA

associated with calciphylaxis [8]. In these cases, a livedo pattern appeared concomitantly specifically around the DDA areas. The only other case of diffuse livedo reticularis associated with DDA was reported in a very particular situation of trabectenin and pelfigrastim injection for liposarcoma [9]. In this case it appeared de novo as a specific clinical presentation of DDA. In our case, primary diffuse livedo reticularis had already been present for a long time: rather than a specific localization of DDA, this suggests a hypothetical vascular predisposition to DDA. The only other case of livedo prior to DDA was described in a context of cutis marmorata telangiectatica congenita [10]; both pathological manifestations occurred on the same arm, suggesting a possible causal link.

Conclusion

The case described sheds some light on the pathogenesis of DDA of the breasts, which is still not completely understood but most certainly implicates vascular insufficiency or inflammation. Indeed, our patient presented three different vascular abnormalities, suggesting a common underlying mechanism, hitherto unknown in this setting. We therefore suggest that diffuse and primary livedo reticularis, could be associated with DDA of the breasts and could be a clinical feature enabling this rare diagnosis.

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Potential conflicts of interest

The authors declare no conflicts of interest.

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