Cutaneous Manifestation of Alpha-1 Antitrypsin Deficiency: A Case of Panniculitis

Sofia Lopes¹, Carla Damas², Filomena Azevedo¹, Alberto Mota¹,³

From the ¹Department of Dermatology and Venereology, Centro Hospitalar São João EPE, ²Department of Pneumology, Lung Transplant Unit, Centro Hospitalar São João EPE, ³Faculty of Medicine, University of Porto, Porto, Portugal.
E-mail: sofialopes88@gmail.com

Indian J Dermatol 2018;63(4):355-7

Sir,

Alpha-1 antitrypsin deficiency (AATD) is a genetic disorder usually associated with hepatic and pulmonary manifestations.¹,² In rare and severe situations, a panniculitis may be present.¹,² It may present before the onset of other systemic manifestations giving a clue to an earlier diagnosis and adequate monitoring.³

A 49 year-old man was referred to our department due to painful and debilitating ulcers of the lower limbs evolving in the last few months, without previous trauma. He had medical history of severe AATD, with biliary pulmonary transplantation in 2010 complicated with bronchiolitis obliterans syndrome in 2015, as well as immunosuppressive drug-related diabetes mellitus. Physical examination revealed ulcerated lesions with a copious oily discharge and a pronounced edema of the lower limbs suggestive of AATD-related panniculitis [Figure 1]. Histologic examination of a skin biopsy showed mild acanthosis in the epidermis and extensive necrosis with a predominant neutrophilic inflammatory infiltrate and cytoesteatonecrosis throughout the dermis and hypodermis, confirming the suspected diagnosis [Figure 2]. Serum level of α-1 antitrypsin was low (51.8 mg/dL).

The patient started treatment with topical fusidic acid two times daily with little improvement. A few weeks later, a cellulitis associated with panniculitis was diagnosed, requiring hospitalization due to elevated C-reactive protein and leukocytosis. He was treated with ertapenem plus vancomycin and silver dressings. The patient improved significantly in the following days.

AATD-related panniculitis usually appears around the age of 40 years and is a debilitating manifestation of the disease.³ There are several possible precipitating factors for these skin lesions including trauma, surgery, debridement, and cryosurgery, but in our patient, none was found.¹ Tender plaques and nodules are usually present, but ulceration is the feature that helps to distinguish this panniculitis from other entities.¹,³,⁴ The presence of an oily discharge is also characteristic.⁴
Lesions may be present not only in the lower extremities but also in the trunk or proximal extremities and usually heal with atrophic scars.\textsuperscript{[1,2,4]}

Histologic examination reveals a predominantly lobular neutrophilic panniculitis with a mixture of necrotic fat lobules and areas without lesions.\textsuperscript{[2,4]}

There is no gold standard treatment for this panniculitis since only a few cases have been reported to date and no clinical trials have been previously conducted.\textsuperscript{[3]} Several options may be considered, including corticosteroids, colchicine, doxycycline, among others.\textsuperscript{[1,2,5]} Dapsone is an alternative and effective drug probably by reducing neutrophils migration and inhibiting oxidative reaction.\textsuperscript{[1]}

Replacement of AAT seems to be a successful approach to these patients, although the costs of this treatment are extremely high.\textsuperscript{[1,3]} Pulmonary transplantation is an option in some systemic manifestations of AATD, but not in the case of panniculitis, since no clinical improvement has been noted in previous cases.\textsuperscript{[5]} In our patient, antibiotics contributed to the significant enhancement of the skin lesions probably due to their anti-inflammatory effect.

The recognition of AATD-related panniculitis is crucial since it may be the first manifestation of this systemic disease. Its high risk of complications and associated mortality explain the need of early treatment and close monitoring of these patients.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Acknowledgment**
We would like to thank Ana Rodrigues-Pereira, MD, for her contribution in the histological diagnosis of the disease.
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. de Oliveira P, Paz-Melgar L, Takahashi MD, Nico MM, Rivitti EA, Mendrone A Jr., et al. Alpha-1-antitrypsin deficiency associated with panniculitis treated with plasma exchange therapy. Int J Dermatol 2004;43:693-7.
2. Laureano A, Carvalho R, Chaveiro A, Cardoso J. Alpha-1-antitrypsin deficiency-associated panniculitis: A case report. Dermatol Online J 2014;20:21245.
3. Cardoso JC. Panniculitis associated with alpha-1 antitrypsin deficiency: From early descriptions to current targeted therapy. Br J Dermatol 2016;174:711-2.
4. Geraminejad P, DeBloom JR 2nd, Walling HW, Sontheimer RD, VanBeek M. Alpha-1-antitrypsin associated panniculitis: The MS variant. J Am Acad Dermatol 2004;51:645-55.
5. Ortiz PG, Skov BG, Benfeldt E. Alpha1-antitrypsin deficiency-associated panniculitis: Case report and review of treatment options. J Eur Acad Dermatol Venereol 2005;19:487-90.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Access this article online
Quick Response Code:
Website: www.e-ijd.org
DOI: 10.4103/ijd.IJD_421_17

How to cite this article: Lopes S, Damas C, Azevedo F, Mota A. Cutaneous manifestation of alpha-1 antitrypsin deficiency: A case of panniculitis. Indian J Dermatol 2018;63:355-7.

Received: September, 2017. Accepted: March, 2018.