Discussion

Follicular mucinosis (FM), first described in 1957 by Pinkus, is clinically characterized by sharply demarcated, infiltrated, erythematous papules or plaques with follicular prominence, scaling and alopecia [1]. Less commonly, nodular lesions, cysts, chronic eczema, follicular spines and acneiform lesions have been described [1, 2]. İn children, lesions resembling lichen spinulosus or pilar keratosis have also been reported in the literature [3].

FM is rare in children. Most cases have a benign prognosis. However, cases which have developed Hodgkin's disease, other lymphomas and leukemias have been reported [4].

Generally, it has been suggested that initial presentation on the head and neck, younger age and the presence of a solitary lesion may be good prognostic signs [1, 5]. Although there is no single histological criterion, reported features of underlying mycosis fungoides include the presence of atypical lymphocytes within the epidermis or follicular epithelium, a band-like lymphocytic infiltrate in the upper dermis, epidermotropism, a paucity of eosinophils and less prominent mucin deposition [1, 5]. In our patient, there was a marked mucinous degeneration of the follicular epithelia without evident features of mycosis fungoides. Considering the age of the patient and the benign clinical and histological features, we did not perform a TCR gene rearrangement study. Currently, the clonal TCR rearrangement in clinically and histologically benign FM is of unknown significance [1]. There is no effective standard treatment for follicular mucinosis. A wait-and-see approach, topical, intralesional and systemic corticoseroids, PUVA, X-rays, dapsone, antimalarials, indomethacine, minocycline, oral isotretinoin, interferon alfa-2b, and UVA1 phototherapy have shown some beneficial effects [6]. In our case, we obtained a complete clinical response with potent topical corticosteroid cream. There was no recurrence at a 2-year follow-up.

In conclusion, we would like to emphasize that FM must be remembered in the differential diagnosis of lesions presenting with follicular hyperkeratosis and spines. Especially in the pediatric age group, it is important to have a high index of suspicion and to take a biopsy in order to make an earlier diagnosis. Although FM is usually considered a benign disease in children, long-term follow-up and biopsy of persistent lesions is mandatory for the detection of possible development of lymphoproliferative malignancies. ■

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1. Bonta M, Tannous Z, Demierra M-F, *et al*. Rapidly progressing mycosis fungoides presenting as follicular mucinosis. *J Am Acad Dermatol* 2000; 43: 635-40.
2. Brown HA, Gibson LE, Pujol RM, *et al*. Primary follicular mucinosis: Long-term follow-up of patients younger than 40 years with and without clonal T-cell receptor gene rearrangement. *J Am Acad Dermatol* 2002; 47: 856-62.
3. Gibson L, Muller S, Peters M. Follicular mucinosis of childhood and adolescence. *Pediatr Dermatol* 1988; 5: 231-5.
4. Lockshin BN, Khachemoune A, Cohen C. Follicular mucinosis in a 4-year-old boy. *Int J Dermatol* 2004; 43: 950-2.
5. Schmid MH, Dummer R, Kempf W, *et al*. Mycosis fungoides with mucinosis follicularis in childhood. *Dermatol* 1999; 198: 284-7.
6. Bolognia JL, Jorizzo JL, Rapini RP. *Dermatology*. 2nd edition. Philadelphia, Elsevier Limited, 2003: 657.

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Is chronic urticaria an atopic condition?

Chronic idiopathic urticaria (CIU) is a common skin disease of unknown etiology [1, 2]. In recent years, we have observed many patients showing coexistence of chronic urticaria and atopic eczema. This led us to look into the relationship between chronic urticaria and atopy. Two representative patients are reported.

Case 1

A 24-year-old man presented in 2002 for a chronic eruption on the face lasting for the past 4 years. This patient had atopic dermatitis (AD) during childhood and allergic rhinitis (AR). His mother also had AD. He complained of "flares" over his face for 2 weeks monthly. During the following months this patient showed either discrete acne or mild eczema over his face. He came back once complaining of severe facial worsening with no lesion over his face but three urticarial papules over his thighs. After questioning, he admitted having for several months had undiagnosed chronic urticaria of the face, which his mild acne and eczema had masked.

Case 2

A 66-year-old patient was referred in June 2003 for chronic urticaria lasting for 30 years. For the past 15 years, he also had hand and foot eczema during the springtime with allergic conjunctivitis. His mother and sister had AD, his son and sister had AR.

Upon examination, this patient had dermographism as well as vesicles on the palms and soles. It was obvious this atopic patient had an association of CIU and eczema.

Cohort report

After seeing several similar patients, we became aware of the possible coexistence of atopy and chronic urticaria. We then prospectively recorded data on personal and familial atopy status in all new and return CIU patients between July 2004 and March 2005: 85 patients were included. The age of patients ranged between 19 and 82. There were 25 male and 60 female patients. We defined atopy as the existence of one or more past or present recurrent item(s) previously diagnosed by a physician: asthma and/or eczema and/or allergic rhino-conjunctivitis. We did not differentiate between extrinsic or intrinsic atopy. We accepted for familial atopy only the same items in first-degree relatives (*i.e.* parents, children and siblings).

Table 1

Past history	Personal	familial
asthma	23 (27%)	20 (23.5%)
eczema	55 (65%)	35 (41%)
Both rhinitis/conjunctivitis	50 (59%)	40 (47%)

Concerning personal atopic status:
– 23 had asthma *i.e.* 27%;
– 55 had eczema *i.e.* 65%;

– 50 had allergic rhinitis/conjunctivitis *i.e.* 59%;
– these figures concern 70 patients who had personal atopy, *i.e.* 82%.
Concerning familial atopic status in the close family:
– 20 had asthma *i.e.* 23.5%;
– 35 had eczema *i.e.* 41%;
– 40 had allergic rhinitis/conjunctivitis *i.e.* 47%;
– these figures concern 60 patients who had atopic close family *i.e.* 70.5%.
Based on the personal and/or familial atopy, out of 85 patients, 82 were atopic, *i.e.* 96.5%.

Comments

Because of these surprising results, we questioned whether chronic urticaria may be part of the atopic diathesis, in addition to asthma, eczema and allergic rhinitis/conjunctivitis.
Looking back into the first definitions of atopy, we were surprised to read that urticaria was part of atopy for at least nine different authors as early as 1892 but also as late as 1930 [3-6]. After 1930, we could not exactly trace when and by whom the term chronic "idiopathic" urticaria was coined in the literature.

Conclusion

Although based on clinical symptoms only, this short study suggests that a high population of patients with CU have an atopic background. Future studies need to confirm this hypothesis by documenting the clinical symptoms of CU together with the immunological criteria of atopy (prick tests, IgE). ■

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1. Kaplan AP. Chronic urticaria: pathogenesis and treatment. *J Allergy Clin Immunol* 2004; 114(3): 465-74.
2. Grattan CE, Sabroe RA, Greaves MW. Chronic urticaria. *J Am Acad Dermatol* 2002; 46(5): 645-57.
3. Wallach D, Taieb A Tilles G. *Histoire de la dermatite atopique*. Chap 1, Masson, Issy les Moulineaux 2004.
4. Special discussion on prurigo, lichenification and allied conditions. *Br J Dermatol* 1912, June 29: 245-267.
5. Talbot FB. Eczema in childhood *Med Clin N Am*, 1918; 1: 985-96
6. Civatte A. *Formes cliniques et pathogénie des prurigos*. IV° Congrès des dermatologistes et syphiligraphes de langue française. Paris: Masson; 1929, 243-308.

Arterial thrombosis with anti-phospholipid antibodies induced by infliximab

Infliximab is a monoclonal antibody directed against TNFα, a major cytokine in the pathophysiology of psoriasis [1]. Infliximab is known to induce the production of auto-antibodies, the role of which in thrombogenicity has not yet been studied in psoriasis. We report a case of secondary arterial thrombosis associated with an anti-phospholipid syndrome induced by infliximab in the treatment of pustular psoriasis.

Case report

A 67-year-old woman was admitted in Spring 2004 for a severe, generalized, pustular psoriasis, resistant to the usual systemic treatments (acitretin, methotrexate, and cyclosporine). Basal assessment of antinuclear antibodies (ANA) was negative. After seven well tolerated injections of infliximab (5 mg/kg, week 0, 2, 6 then every 8 weeks) the patient was in partial remission of the disease, with only persistence of an acrodermatitis continua of Hallopeau. At week 6 of treatment, the ANA increased to 1/2560 along with positive ds-DNA antibodies at 22.3 UI/mL, without any clinical symptom of lupus or other connective tissue disease. Nevertheless we decided to replace infliximab by methotrexate and etanercept, but after 1 month, a generalized pustular (von Zumbusch type) psoriasis relapse occurred. As the patient was known to be resistant to all systemic psoriasis treatments but infliximab, we decide to reintroduce this last treatment. Eight days after the first re-injection, the patient developed a bilateral acute ischemia of both legs. The angio scanner showed a bilateral extended arterial thrombosis, necessitating urgent revascularization surgery followed by oral vitamin K antagonist. Coagulation screening tests revealed the presence of an anti-phospholipid IgM > 200 UI/mL (N < 12.9 UI/mL) associated with an anti-β2glycoprotein I IgM at 490 UI/mL (N < 10 UI/mL). The association of a vascular thrombosis with anti-phospholipid antibodies (aPL) in our patient led us to make the diagnosis of infliximab-induced anti-phospholipid syndrome, and treatment by infliximab was then suspended. Over the next few weeks, as the psoriasis worsened, we decided to reintroduce acitretin, associated with topical steroids and obtained a stabilization of the disease. Six months after the suspension of infliximab and the complete healing of the patient, ANA were dosed at 1/640, with positive ds-DNA antibodies (4.2 UI/mL) and with persistence of anti-cardiolipin antibody at 21 UI/mL. One year and 18 months later (as the patient was still treated by acitretin + topical steroids with a stable PASI score for psoriasis around 40), ANA were dosed at 1/160, ds-DNA were negative, with disappearance of the anti-cardiolipin and the anti-β2glycoprotein I antibodies. 3 years after the occurrence of the thrombosis and the definitive discontinuing of infliximab treatment, the patient's psoriasis remained clinically stable, as did the dosage of auto-antibodies.

Discussion

Infliximab is a very efficient treatment of severe pustular poriasis, as an alternative or in association with methotrexate, acitretin or cyclosporine. The appearance of ANA or ds-DNA antibodies after 6 weeks of infliximab treatment is usual, and is not associated with clinical symptoms in most cases, probably because these antibodies are usually low affinity IgM (the clinical manifestations of lupus are associated with high-affinity IgG auto-antibodies) [2]. The appearance of an anti-cardiolopin IgM has also been described with infliximab therapy, but never – until now – associated with an arterial thrombosis, although a few cases of venous thrombosis have been reported during infliximab therapy [3-5]. Nevertheless, in this case report, the pathogenicity of the anti-phospholipid IgM is highly probable in the genesis of the thrombosis.

Conclusion

Arterial thrombosis secondary to an anti-phospholipid syndrome is a rare but severe complication that can be induced by infliximab. Exploration and follow up of

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