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From the Clinic

Renal amyloidosis secondary to hidradenitis suppurativa

Hidradenitis suppurativa (HS), also known as Verneuil's disease [1] or acne inversa, is characterized by hyperkeratosis of the follicular epithelium, resulting in obstruction of the follicular stoma and chronic perifollicular inflammation with remissions and relapses, usually involving skin overlying the axillary, inguinal, perianal, perineal and inframammarian regions. It usually presents in the second and third decades of life, has a female preponderance, no racial predilection and a prevalence of 4%. Some causing factors might be obesity and related hormonal changes, hyperandrogenism, immunologic and genetic status, diet and smoking [2]. The diagnosis is based on the presence of relapsing lesions in typical anatomical regions. HS may be localized with chronic, relapsing, superficial papulopustular lesions, as it may be associated with deep, widespread abscesses, sinus tracts, fistulas, malignancies like squamous cell carcinoma and systemic complications like arthropathy or anaemia [3]. Besides topical and surgical care, therapy with antibiotics, steroids, retinoids and anti-TNF agents is usually considered. Rarely, amyloidosis may complicate the cutaneous disease [4].

A 42-year-old male was referred to our hospital for nephrotic syndrome. Three years before he was diagnosed with *hidradenitis suppurativa* when papulopustular lesions over the inguinal, perineal, perianal and axillary regions appeared. Over these 3 years, he had relapses with fever, extensive perineal tenderness and partial remissions leaving fibrotic scar tissue following treatment with topical steroids, keratolytics and antibiotics. His temperature was 37.1°C, blood pressure 140/90 mmHg and heart rate 76/min. He had bilateral pretibial pitting oedema and active bilateral inguinal, perineal, perianal and left axillary papulonodular and pustular, hyperaemic, tender lesions of 2–3 cm diameter, some of which fluctuating and draining, scar tissue, sinus tracts of the older lesions and bilateral inguinal and left axillary tender lymphadenopathies, the largest of which being 2 cm in diameter (Figure 1).

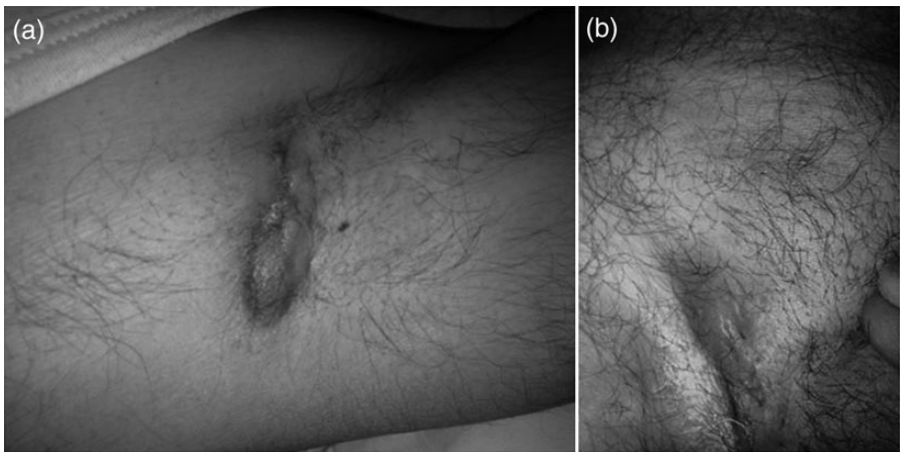

Fig. 1. Left axillary active lesion (**a**) and sinus opening on the right lateral scrotal wall (**b**).

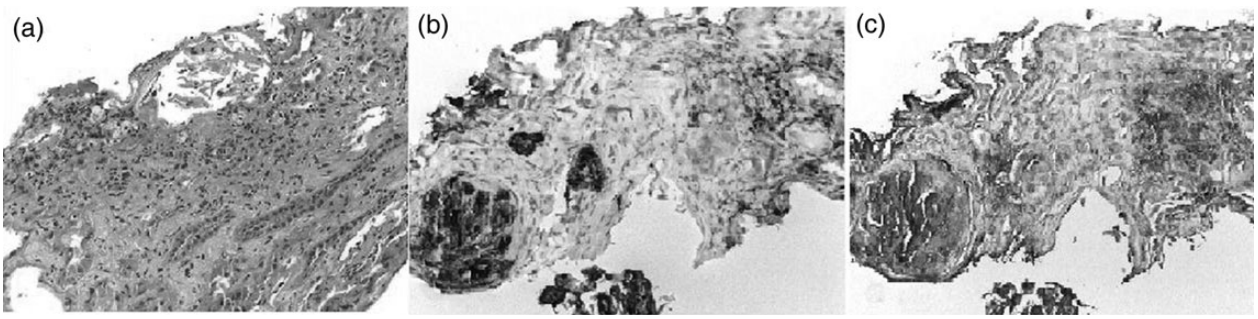

Fig. 2. Interstitial eosinophilic amorphous amyloid deposition on haematoxylin and eosin stained section (**a**), strong positive glomerular and perivascular staining with amyloid A (**b**) and P component positivity (**c**).

No pathogen was isolated in the cultures of sinus openings and in his blood. Topical keratolytic and anti-inflammatory drugs combined with roxithromycin, rifampicin and clindamycin treatment for 1 month resulted in partial remission of the active lesions. A biopsy of the perineal skin revealed a non-specific inflammatory reaction without amyloid deposition. He was found to have a heterozygous M680I mutation of the exon 10 of the *MEFV* gene. Kidney biopsy revealed glomerular, perivascular and interstitial deposits that stained with Gentian violet, compatible with amyloidosis that was identified immunohistochemically as AA amyloidosis (Figure 2). Despite treatment with colchicine, kidney failure and proteinuria showed progression.

Amyloidosis and nephrotic syndrome are rare complications of HS with only four such cases in the literature [4]. One had rectal biopsy-proven symptomatic gastrointestinal amyloidosis, another one had infiltrative cardiomyopathy, assumed to be caused by amyloidosis.

In our case, there was no amyloid deposition in skin biopsy and the abdominal computed tomography scan was normal with no hepatosplenomegaly. The patient did not have diarrhoea or gastrointestinal bleeding. His echocardiography was normal and he had no neuropathy.

Although seen prevalently in endemic regions, carriers of MEFV gene mutations may have an increased risk of inflammatory diseases resulting in amyloidosis. The clinical course of inflammatory diseases like rheumatoid arthritis may be more severe in *MEFV* gene mutation carriers [5].

Despite its benign nature, hidrosadenitis must be early and actively treated as any chronic suppuration to avoid the development of AA amyloidosis, especially in carriers of the *MEFV* gene mutation.

Conflict of interest statement. None declared.

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