CASE REPORT

Two cases of hidradenitis suppurativa exacerbated by ambulatory aides for myelomeningocele

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INTRODUCTION

Hidradenitis suppurativa (HS) is a chronic inflammatory and recurrent disease of the hair follicle, characterized by nodules, abscesses, fistulas, and scars on the intertriginous areas. The prevalence rates range between 1% and 4%.1 Many factors contribute to the inflammatory state: genetic predisposition, smoking, obesity, hormonal factors, and the microbiome. In addition, the skin barrier dysfunction in friction areas could play a role in the HS pathogenesis.1

Myelomeningocele (MMC) is a congenital defect caused by a primary failure of either neural tube or mesenchymal closure at the caudal neuropore in the embryonic period that is characterized by protrusion of the meninges and spinal cord through open vertebral arches leading to lifelong paralysis. In addition, MMC patients are often limited by other disabilities, the severity of which depends on the magnitude and location of the neural defect. The incidence is 1 to 2 cases per every 2,000 live births.2

To the best of our knowledge, the association of HS with MMC has not been reported. We describe 2 cases of HS in patients with MMC, and we postulate possible physiopathogenic mechanisms for its development.

CASE 1

A 35-year-old man, with a body mass index (BMI) of 22, presented with progressive worsening of HS lesions during the last 6 months that caused intense pain related to the use of crutches. The patient had a history of MMC at the lumbar level, flaccid paraplegia that required the use of reciprocator and crutches, neurogenic bladder, and axillary HS since the age of 25, and no family history of HS. He received several oral antibiotics with little clinical response. In addition, he had 4 surgical drainages with temporary improvement of pain. Physical examination found inflammatory nodules, abscesses, fistulas, and contracted scars in both axillae with healthy interlesional skin (Hurley stage II; Figs 1 and 2). He was treated with clindamycin, 300 mg twice a day, plus rifampicin, 600 mg once a day for 10 weeks with resolution of the acute inflammatory component and pain, with a sustained response at 6 months of follow-up.

CASE 2

A 30-year-old woman, with a BMI of 21.6, presented with inflammatory nodules and abscesses in the groin, inflammatory nodules, abscesses, fistulas and contracted scars in the axillae (Hurley stage III; Fig 3). The patient had a medical history of MMC at the lumbar level, flaccid paraplegia (walks 1 hour daily with reciprocator and crutches and the rest of the day she uses a wheelchair), neurogenic bladder, hypothyroidism, severe acne, and HS since the age of 17, and no family history of HS. Twelve years ago, she was treated with isotretinoin for 18 months with improvement of the acne but not of HS. She also had treatment with trimethoprim-sulfamethoxazole several times. Three years ago, she received

Abbreviations used:

BMI: body mass index
HS: hidradenitis suppurativa
MMC: myelomeningocele
infliximab 5mg/kg during weeks 0, 2, and 6 and then every 8 weeks until it was stopped after 12 months because of the complete resolution of the inflammatory lesions. Then, with the objective of removing fistulas and contracted scars, a wide excision surgery with a rotary flap of both axillae was performed. Two years later, the patient no longer has active lesions of HS (Fig 4).

**DISCUSSION**

We describe 2 patients with MMC and HS, and we postulate paraplegia and the use of crutches or other supporting orthopedic devices as triggers or aggravating factors of the latter.

Mechanical stress is a risk factor for HS development. Some patients with MMC may present a permanent occlusion of the inguinal folds according
to the degree of paraplegia. In addition, axillae are areas of constant friction, either by the use of crutches or as a support for mobilization assisted by third parties.

The paradigm for explaining the role of friction in HS development is obesity, where lesions are located in areas of skin friction beyond the classic location in large folds, such as redundant abdominal skin.1-5 Moreover, there is a positive correlation between the severity of the disease and the BMI.6 Furthermore, some studies show that weight loss (either by a nutritional approach or after bariatric surgery) could improve the symptoms of HS.5 There are some explanations for why obesity can contribute to the development of HS: (1) there is more skin-to-skin contact, leading to an increase in maceration and friction; (2) the mechanical stress contributes to hyperkeratinization and follicular occlusion, with its consequent dilation, microtears and follicular rupture1,4; and (3) in these skin folds, a warm humid and occlusive microclimate develops, which favors microbial growth.1,3,6 On the other hand, the adipose mass itself is associated with a low-grade systemic inflammatory state, capable of producing adipokines, cytokines, and chemokines.1

The role of mechanical stress in HS is also supported by the report of a pediatric case that had HS-like lesions on an inguinal nevus comedonicus when the child began to walk.7 de Winter et al3 describes a case of a man with a posttraumatic amputation of a leg, who had HS-like lesions with fistulas on the stump after using a prosthesis. Moreover, results of a survey suggest that tight clothing or friction aggravated the symptoms of HS.9 Other investigators add that HS can be caused by a defective follicular support. It has been found that the sebofollicular junction of the skin with HS has a thinning of the basal membrane, which may explain the fragility of this union. It can be hypothesized that, in individuals with such a predisposition, exposure to mechanical stress easily leads to damage and follicular rupture.9

Interestingly, the role of mechanical stress as a trigger factor is seen in other diseases owing to follicular occlusion, such as acne mechanica and pilonidal cysts.10

We report 2 cases with HS and MMC, and we propose possible physiopathogenic pathways for its coexistence, such as mechanical occlusion and frictional stress. Perhaps this warns us about the need to take precautionary measures in paraplegic patients. As a limitation, it is a retrospective report of only 2 cases. Further studies with larger numbers of patients should confirm if it is an association or a coincidence.

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