Cutaneous Crohn's Disease Causing Deformity of the Penis and Scrotum

Elena González-Guerra, Jorge Angulo, Inmaculada Vargas-Machuca, María del Carmen Fariña, Lucia Martín and Luis Requena*

Department of Dermatology, Fundación Jiménez Díaz, Universidad Autónoma, Avda. Reyes Católicos 2, ES-28040 Madrid, Spain.
**E-mail: lrequena@fjd.es*
Accepted October 3, 2005.

Sir,

Cutaneous Crohn's disease (CD), also known as metastatic CD, first described by Parks et al. in 1965 (1), is a rare complication of CD in which granulomatous lesions involving skin are separated from gastrointestinal lesions by normal tissue. Cutaneous involvement may be the first sign of CD, or more frequently, may appear during the course of the disease (2). The pathogenic mechanism for the development of these skin lesions is uncertain.

We report here a case of a young man with cutaneous CD mostly involving the penis and scrotum without bowel lesions.

CASE REPORT

A healthy 26-year-old man, with no personal or family history of actual interest, was referred to our department from the Urology department, where he had been hospitalized for 1 month due to considerable inflammation and deformity of the scrotal region and penis that prevented retraction of the prepuce. Testicular ultrasound showed oedema of the soft tissue at this level with no other remarkable alterations. A tentative diagnosis of hidradenitis suppurativa was made and treatment with amoxicillin and ibuprofen resulted in mild improvement.

When admitted to the Department of Dermatology he presented with dramatic oedema of the penis and scrotum that caused considerable deformity, along with atrophic scars at the base of the penis, lateral sides of the scrotum and inguinal region (Fig. 1). In the groin, induration papules and pustules were found, with surrounding erythema of 8 months' duration. In the right axilla infiltration and two pustules were found, but no accompanying scarring lesions at that site. One small external haemorrhoid was observed in the perianal region. The patient reported no fever, diarrhoea, pathological products in stools (blood, mucus or pus) or weight loss.

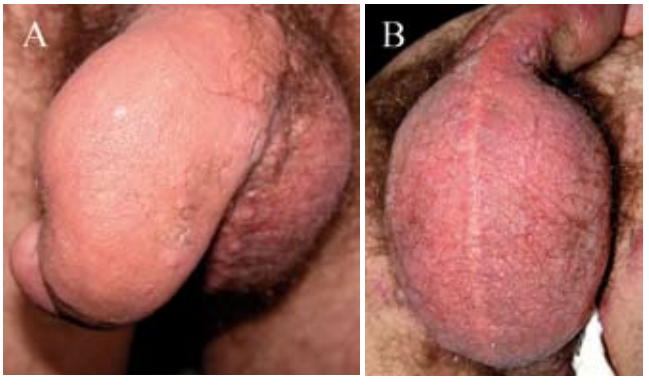

Fig. 1. "Saxophone" deformity of the penis with scars on the skin surface (A) and scrotal oedema with erythema and atrophic scars on the basis of the penis (B).

Histopathological study of a nodular lesion in the suprapubic region showed numerous non-caseating epithelioid granulomas with abundant multinucleate giant cells, with a mostly perivascular distribution, scattered throughout the entire thickness of the dermis and the dermal-subcutaneous border (Fig. 2). Cultures for Mycobacterium species and fungi of a fragment of the cutaneous biopsy yielded negative results.

No macroscopic or microscopic signs of CD were observed on colonoscopy. Histopathogical study of biopsies from the terminal ileum and right colon showed no pathological changes of the intestinal mucosa. The blood test showed leukocytosis (12.380/mm^3), reactive C protein: 0.8mg/dl and negative results HLA-B27, p-ANCA and c-ANCA. On the basis of all previous data, a diagnosis of metastatic CD was established.

The patient received treatment with 30 mg/day of deflazacort (Zamene®) for 7 months, with slight improvement. At present, the patient is receiving corticosteroids at the same dosage, observing slower and incomplete recovery.

DISCUSSION

CD is a granulomatous, idiopathic, chronic inflammatory process that can involve any segment of the gastrointestinal tract (3). The disease is a systemic condition in which the most important extraintestinal manifestation is cutaneous involvement (4). Cutaneous CD is more frequently observed in patients with colon or colorectal involvement than in those with involvement of the small intestine alone (5).

Cutaneous manifestations have been reported in 14–44% of patients with CD, depending on whether or not perianal involvement is included (6). Although dermatological lesions in most patients occur as a clinical symptom during the course of the disease, in 25% of the cases they precede the diagnosis by 3–8 years (7), as occurred in our patient.

Dermatological lesions described in CD can be classified into several groups (8): (*i*) specific lesions of cutaneous CD; (*ii*) reactive dermatosis; (*iii*) cutaneous manifestations secondary to malabsorption; (iv) cutaneous manifestations secondary to treatment; and (*v*) miscellaneous conditions. Cutaneous CD can also be divided into two clinical forms: genital (56%) and extragenital (44%). Two-thirds of children with CD present genital involvement, in comparison with 50% of adults with CD (9).

In patients with cutaneous CD, cutaneous manifestations and intestinal involvement do not always follow a parallel course, and in fact, treatment of the bowel disease is not always followed by improvement of the cutaneous lesions (10).

 ISSN 0001-5555
DOI: 10.2340/00015555-0030

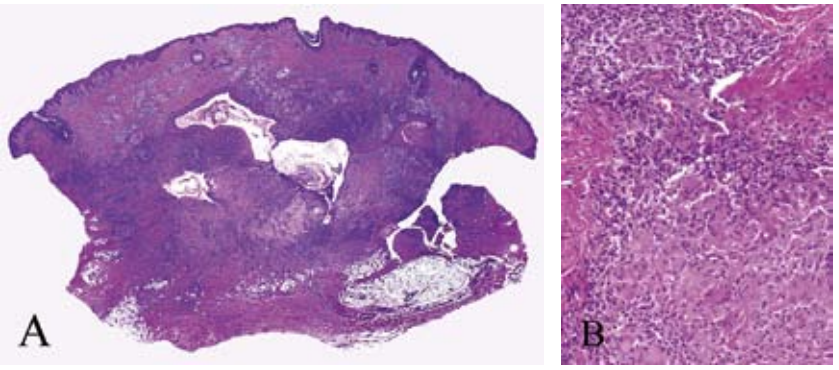

Fig. 2. Scanning power view of the cutaneous biopsy showing cystic spaces and granulomatous infiltrates involving the full-thickness of the dermis (A) (original magnification ×10). Higher magnification showing non-caseating granulomas with epithelioid histiocytes and abundant number of multinucleated giant cells (B) (original magnification ×200).

In rare instances (less than 100 cases reported in the literature), cutaneous lesions are found in areas anatomically distant from the gastrointestinal tract, separated by normal skin. This entity is known as metastatic CD and comprises a high clinical variability of cutaneous lesions including ulcers, papules, nodules, hard plaques and crusts. It can affect any area of the skin, but the lower extremities, skin folds and external genitals are the most commonly involved sites (11).

Histopathologically, the findings in cutaneous lesions of CD are not characteristic, but the most frequent features include non-caseating epithelioid granulomas with some multinucleate giant cells, mainly of perivascular distribution, scattered throughout the entire thickness of the dermis and, sometimes, extending to subcutaneous fat (9).

Differential diagnosis of cutaneous CD should be established with other granulomatous disorders, including sarcoidosis, tuberculoid leprosy, tuberculosis, infections caused by opportunistic mycobacteria, some fungal infections, granuloma annulare, venereal lymphogranuloma, foreign body granuloma and pelvic or retroperitoneal neoplastic processes (12). Most of these infectious granulomatous processes may be ruled out by cultures and microbiological investigations.

Most cutaneous manifestations of CD tend to become chronic (13). Treatment with prednisolone and methylprednisolone have been first-drug regimens. However, the response has not been satisfactory in all cases, and other drugs have been administered with variable response, including sulfasalazine, azathioprine, 6-mercaptopurine, metronidazole, ciprofloxacin, cyclosporine and infliximab. There are also patients that have been treated by dermatological and intestinal surgery (14).

Involvement of the penis and scrotum by CD is very rare. In our literature review we found only one similar case of a man with deforming oedema of the penis, without intestinal involvement (15).

REFERENCES

1. Parks AG, Morson BC, Pegum JS. Crohn's disease with cutaneous involvement. Proc R Soc Med 1965; 58: 241–242.
2. Chalvardijan A, Nethercott JR. Cutaneous granulomatous vasculitis associated with Crohn's disease. Cutis 1982; 30: 645–655.
3. Crohn BB, Ginzburg L, Oppenheimer GD. Regional ileitis. JAMA 1932; 99: 1323–1329.
4. Bel S, García-Patos D, García G. Linfedema vulvar: una manifestación infrecuente de la enfermedad de Crohn metastásica. Gastroenterol Hepatol 2001; 24: 297–299.
5. Biancone L, Geboes K, Spagnoli G. Metastatic Crohn's disease of the forehead. Inflamm Bowel Dis 2002; 8: 101–105.
6. Ciubotaru V, Tattevin P, Carton-Savin L. Manifestations métastatiques cutanées de la maladie de Crohn. Rev Med Interne 2003; 24: 198–201.
7. Fenniche S, Mokni M, Haouet S, Ben Osman A. Maladie de Crohn vulvaire. Ann Dermatol Venereol 1997; 124: 629–632.
8. Enguidanos J, Sánchez JM, Fortea JM. Vasculitis granulomatosa cutánea en la enfermedad de Crohn: Crohn metastásico. Actas Dermosifiliogr 1998; 89: 339–348.
9. Anderson KM, Sedghizadeh P, Allen CM, Camisa C. Oral disease. In: Bolognia JL, Jorizzo JL, Rapini RP, eds. Dermatology. London: Mosby, 2003: 1079–1098.
10. Lebwohl M, Fleischmajer R, Janowitz H, Present D, Prioleau PG. Metastatic Crohn's disease. J Am Acad Dermatol 1984; 10: 33–38.
11. Acker S, Sahn E, Rogers H. Genital cutaneous Crohn disease: two cases with unusual clinical and histopathologic features in young men. J Am Acad Dermatol 2000; 22: 443–446.
12. Handfield-Jones SE, Prendiville WJ, Norman S. Vulval lymphangiectasia. Genitourin Med 1989; 65: 335–337.
13. Berkowitz E, Lebwohl M. Cutaneous manifestations of inflammatory bowel disease. J Eur Acad Dermatol Venereol 2000; 14: 349–350.
14. Borrego L, Hernandez J, Islas D, Peña L, Baez O, Hernández B. Enfermedad de Crohn con afectación vulvar. Presentación de un caso. Actas Dermosifiliogr 1993; 84: 447–450.
15. Macaya A, Marcoval J, Bordas X, Moreno A, Vázquez S, Peyri J. Crohn's disease presenting as prepuce and scrotal edema. J Am Acad Dermatol 2003; 49: 182–183.