

Title page

• *Title*

Metastatic cutaneous Crohn's disease of the face - a successful case treated with ustekinumab

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• *Short title*

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L.M.F.: conception and design of the case and writing up of the first draft of the paper

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Extraintestinal manifestations are common in Crohn's disease (CD), with an incidence of 22%–44%, and its clinical spectrum is very extensive.¹ Metastatic Crohn's disease (MCD) is one of the most uncommon cutaneous extraintestinal manifestations.² It is a rare granulomatous inflammatory process, similar to the pathogenic mechanism of CD that occurs in sites discontinuous from the gastrointestinal tract.^{1,3,4} The face is the second rarest location.^{2,4}

The diagnosis can be confirmed by a histopathological examination which characteristically shows sterile, noncaseating granulomas composed of Langerhans giant cells, epithelioid histiocytes, lymphocytes, and occasional plasma cells, localized to the superficial papillary and deep reticular dermis.^{3,4}

MCD should be managed in close collaboration with a dermatologist². First-line recommendations include topical steroids or topical tacrolimus, followed by oral metronidazole, and if they are still symptomatic, prednisolone 30 mg/d can be initiated. Immunomodulators can be considered for treatment-resistant disease. Finally, case reports have shown success with tumor necrosis factor-alpha inhibitors.¹

A 40-year-old male patient with a 10-year history of Crohn's disease, only with jejunal involvement, under infliximab 5mg/kg every eight weeks, in clinical and endoscopic remission, started to complain about symmetrical peripheral polyarthralgias (wrists, ankles and elbows) that began one month after infliximab and resolved immediately after its administration. There was a need to shorten infliximab administrations to every six weeks and, after a Rheumatology evaluation, oral and topical steroids were associated, with no response. So, it was decided to add methotrexate 10mg weekly, with symptomatic control.

Three months later, progressive dermatological alterations have appeared: erythematous plaques on the elbows suggestive of psoriasis and exuberant scaling rash on the face (more exuberant on malar and forehead locations) (Figure 1A) that was biopsied and which histology revealed interstitial granulomatous dermatitis. Multidisciplinary evaluation concluded that it was psoriatic arthritis and Metastatic Cutaneous Crohn's Disease of the face. It was decided to stop infliximab and swap to ustekinumab. Now, after two administrations, there is no joint complaint and there is a clear regression of the dermatological lesions (Figure 1B).

There are some aspects that makes this an extremely rare case: this disorder is more commonly seen in patients with colonic or rectal involvement^{2,3} and our patient has only jejunal involvement. A slight female preponderance had been observed in a few studies⁴ and the age of onset of this entity ranged from 29 to 39 years in adults – the diagnostic of our patient was made later.⁴ The malar region, the chin, and the perioral area are the most usual locations on the face, whereas the temporal region, the forehead, and the submandibular area are the most infrequent² – he had more exuberant rash on malar (most usual) and forehead (less frequent) areas.

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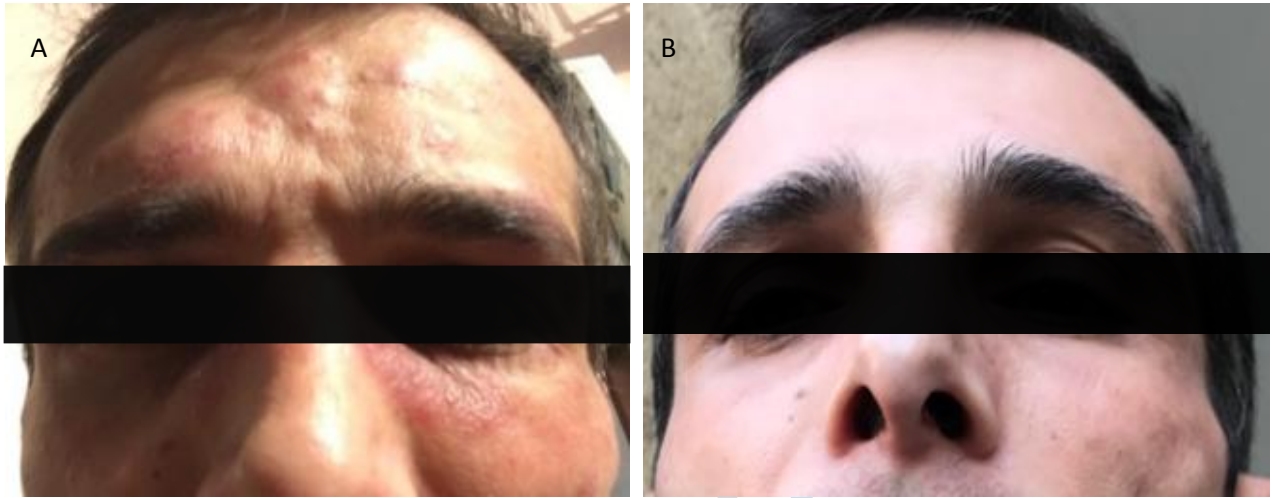
Figure Legend

Figure 1 A: Scaling rash on the face. B: Regression of the dermatological lesions after two ustekinumab administrations

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Figure

Figure 1



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