Ana Cláudia Mendes do Nascimento[1]
Tatiana Mimura Cortez[1]
Daniela Barros Cortez Gaspardo[1]
Hélio Amante Miot[2]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20142615

CASE REPORT

A 40-year-old man had, since 1996, presented arthralgia of the knees, ankles, and wrists; recurrent oral and genital ulcers; papulopustular lesions on the upper limbs and upper front torso, and painful nodules on the lower limbs.

A diagnosis of Behçet's disease was established from clinical criteria and histopathological exam. Treatment was initiated with dapsone, deflazacort, calcium carbonate, and Vitamin D with lesion remission.

In 2010 he began recurrent crises of oedema, erythema, and painful nodules on the ear auricles, more accentuated on the left side, which abated after increased corticosteroid treatment (Figure 1).

A smoker and drinker, he presented femural neck osteopenia and hepatic steatosis of alcoholic aetiology. Annual ophthalmological exam was performed without ocular compromise from Behçet's disease, although he presented hypertensive retinopathy.

Laboratory exams had shown normal hepatic enzyme levels, serum negativity for hepatitis B & C, hemogram, and electrolytes. Inflammatory tests were altered: VHS 28 mm and PCR 36.0.

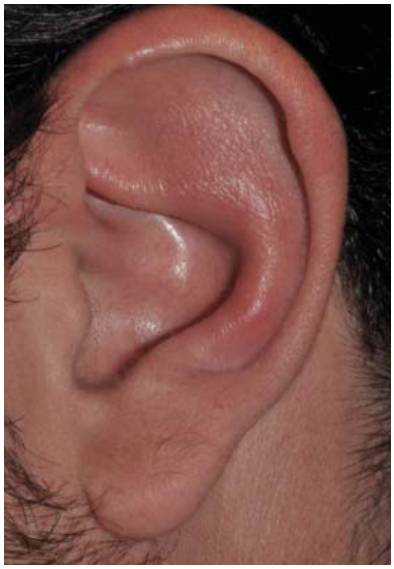

FIGURE 1: Painful erythematous nodules of the ear auricle preferring the antihelix without affecting the lobe

Received on 13/3/2013
Approved by the Advisory Board and accepted for publication on 01/4/2013
* Work performed at the Department of Dermatology and Radiotherapy, Botucatu School of Medicine - "Júlio de Mesquita Filho" Paulista State University (FMB-UNESP) – Botucatu, SP, Brazil.
Financial Support: None.
Conflict of Interests: None.

[1] Doctor – Resident of the Dermatology and Radiotherapy Department, Botucatu School of Medicine - "Júlio de Mesquita Filho" Paulista State University (FMB-UNESP) – Botucatu, SP, Brazil.
[2] PhD – Assistant Professor of the Dermatology and Radiotherapy Department, Botucatu School of Medicine - "Júlio de Mesquita Filho" Paulista State University (FMB-UNESP) – Botucatu, SP, Brazil.

DISCUSSION

MAGIC syndrome (mouth and genital ulcers with inflamed cartilage).

Relapsing Polychondritis (RP) and Behçet's disease (BD) are different multisystem inflammatory diseases. Manifestations of both diseases occurring in the same patient was reported by Firestein (1985) in a series of five cases, suggesting the existence of a common pathogenic mechanism.[1]

BD can be diagnosed from criteria which were proposed by the International Study Group for Behçet´s Disease in 1990 and supported by findings of vasculitis and thrombosis subjacent to mucosal ulcers (Chart 1).[2]

RP diagnosis is based on clinical criteria by McAdam: (1) bilateral auricular chondritis; (2) serum negative non-erosive inflammatory polyarthritis; (3) nasal chondritis; (4) ocular inflammation, including conjunctivitis, keratitis, scleritis, episcleritis, or uveitis; (5) respiratory tract chondritis; and (6) audiovestibular lesion; at least three of these criteria should be present.[3]

The name MAGIC syndrome is used to designate an overview of BD and RP manifestations, implied by a common physiopathogenic mechanism of probable autoimmune origin. This association is rare with few cases described in literature.[4-6]

According to Firestein, patients with MAGIC present specific findings for BD and RP, respectively, orogenital aphthous ulcers and chondritis (auricular, nasal, or respiratory tract), can also present less specific manifestations such as audiovestibular dysfunction, large artery thrombosis and aneurisms, as well as

CHART 1: Diagnostic criteria for Behçet's disease. The presence of one major and at least two minor criteria are necessary to establish diagnosis[2]

Major Criteria
Recurrent oral ulceration – minor or major aphthae or herpetiform ulceration observed by the doctor or reliably reported by the patient, which has recurred at least three times in a twelve month period.

Minor Criteria:

- Recurrent genital ulceration – genital aphthous ulceration or scarring, especially in men observed by a doctor or reliably reported by the patient.
- Ocular lesions - (1) anterior uveitis, posterior uveitis, cells in the vitreous humour and in slit lamp exam, or (2), retinal vasculitis seen by ophthalmologist doctor
- Skin lesions - (1) erythema nodosum-like lesions, observed by a doctor or reliably reported by the patient, pseudo folliculitis, papulopustular lesions, or (2) acneiform nodules consistent with Behçet's disease, observed by a doctor and in patients who have not received cortiosteroids.
- Positive Pathergy test – erythematous papule larger than 2 mm at prick site, checked by a doctor 48 hours after obliquely pricking the avascular part of the skin to a depth of 5mm with a 20 to 22 gauge sterile needle.

TABLE 1: Frequency of clinical manifestations in MAGIC syndrome.[9]

Oral Ulcers	100.0%
Bilateral Auricular Chondritis	100.0%
Genital Ulcers	83.3%
Polyarthritis	83.3%
Nasal Chondritis	58.3%
Scleritis, episcleritis, conjunctivitis, keratitis	58.3%
Pseudofolliculitis	58.3%
Thrombosis	41.6%
Uveitis	25.0%
Audiovestibular	25.0%
Cutaneous Vasculitis	25.0%
Gastrointestinal	25.0%
SNC	16.6%
Orchiepididimitis	16.6%
Erythema nodosum	16.6%
Pathergy	8.3%
Respiratory Tract Chondritis	8.3%

other signs and symptoms common to many rheumatic disorders, such as ocular changes, pseudofolliculitis, migratory polyarthritis, glomerulonephritis, gastrointestinal tract involvement, and central nervous system involvement (Table 1).[4-7]

Therapeutic experience with MAGIC syndrome is limited to case reports with very successful treatment using pentoxifyllin, corticosteroids, dapsone, and infliximab.[4,5,8,9]

In this case, treatment was based on clinical manifestations and histological findings characteristic of BD; it was initially systemic corticosteroid and dapsone, which after the appearance of chondritis, was changed to a higher dose oral corticosteroid with maintained dapsone.[10]

BD and RP are long lasting and have a morbimortality potential due to multisystemic compromise. Dermatologists should be alert to manifestations of chondritis in BD patients and the early identification of MAGIC syndrome. ❑

Abstract: The authors present a male 40-year-old patient with established diagnosis of Behçet's disease which had evolved to recurrent bilateral auricular polychondritis crises. MAGIC syndrome (mouth and genital ulcers with inflamed cartilage) is rare and groups together patients with this clinical picture without necessarily fulfilling the clinical criteria for Behçet's disease or relapsing polychondritis, demonstrating an independent disorder.
Keywords: Behcet Syndrome; Polychondritis, relapsing; Stomatitis, aphthous; Vasculitis

REFERENCES

1. Firestein GS, Gruber HE, Weisman MH, Zvaifler NJ, Barber J, O'Duffy JD. Mouth and genital ulcers with inflamed cartilage: MAGIC syndrome. Five patients with features of relapsing polychondritis and Behçet's disease. Am J Med. 1985;79:65-72.
2. Criteria for diagnosis of Behçet's disease. International Study Group for Behçets Disease. Lancet. 1990;335:1078-80.
3. McAdam LP, O'Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: prospective study of 23 patients and review of the literature. Medicine (Baltimore). 1976;55:193-215.
4. Orme RL, Nordlund JJ, Barich L, Brown T. The MAGIC syndrome (mouth and genital ulcers with inflamed cartilage). Arch Dermatol. 1990;126:940-4.
5. Imai H, Motegi M, Mizuki N, Ohtani H, Komatsuda A, Hamai K, et al. Mouth and genital ulcers with inflamed cartilage (MAGIC syndrome): a case report and literature review. Am J Med Sci. 1997;314:330-2.
6. Kötter I, Deuter C, Günaydin I, Zierhut M. MAGIC or not MAGIC--does the MAGIC (mouth and genital ulcers with inflamed cartilage) syndrome really exist? A case report and review of the literature. Clin Exp Rheumatol. 2006;24:S108-12.
7. Gamboa F, Rivera JM, Mayoral L, Grilo A. Behçet's disease and relapsing polychondritis (MAGIC syndrome) associated with antiphospolipid syndrome. Med Clin (Barc). 1998;110:678-9.
8. Geissal ED, Wernick R. A case of severe MAGIC syndrome treated successfully with the tumor necrosis factor-alpha inhibitor infliximab. J Clin Rheumatol. 2010;16:185-7.
9. Neves FDS, Gonçalves DP, Lage LV, Gonçalves CR. Síndrome de Behçet e policondrite recidivante: síndrome MAGIC. Rev Bras Reumatol. 2006; 46:157-60.
10. Bonamigo RR, Razera F, Olm GS. Neutrophilic dermatoses - Part I. An Bras Dermatol. 2011; 86:11-25.

MAILING ADDRESS:
Hélio Amante Miot
Departamento de Dermatologia e Radioterapia da Faculdade de Medicina de Botucatu da Universidade Estadual Paulista (UNESP)
18618-000 Botucatu, SP, Brazil.
Email: heliomiot@fmb.unesp.br

How to cite this article: Nascimento ACM, Gaspardo DBC, Cortez TM, Miot HA. Syndrome in Question: MAGIC Syndrome. An Bras Dermatol. 2014;89(1):177-9.