Complete Melkersson-Rosenthal Syndrome in a Patient with Crohn's Disease

G.		De Aloe
P.		Rubegni
C.		Mazzatenta
M.		Fimiani

Department of Dermatology, University of Siena, Italy

Key Words

Melkersson-Rosenthal syndrome
Crohn's disease
Orofacial oedema

Prof. Michele Fimiani, Istituto di Clinica Dermatologica, Università degli Studi di Siena, Policlinico 'Le Scotte', I-53100 Siena (Italy), Tel. 577 40190, Fax 577 44238

Melkersson-Rosenthal syndrome (MRS) is an uncommon condition, well known in terms of clinical features and course but of uncertain aetiopathogenesis [1-3]. The histopathology of the lesions, characterized by non-caseating epithelioid granulomatous inflammation, suggests a possible relation with sarcoidosis and overall with Crohn's disease (CD) [4].

A 23-year-old woman had CD at the age of 10 years, and for this reason she underwent right hemicolectomy in 1984. In the following years, clinical examination, laboratory tests and scintigraphy with labelled granulocytes did not detect any signs of intestinal relapse. Seven years after the operation, the patient was in good health but developed recurrent oedema of the upper lip. These episodes progressively increased in frequency and intensity, eventually involving most of the right side of the face. When examined by us, the patient had persistent swelling of the right side of the face, partially involving the upper lip. The tongue and oral mucosa were spared. She complained of dysaesthesia of the homolateral side of the face and devastating psychological effects due to her appearance. Histological examination of a biopsy specimen of the oral mucosa showed many non-necrotizing granulomas consisting of epithelioid cells, giant cells and lymphocytes throughout the dermis. Electromyographic examination showed a slight reduction in amplitude of evoked potentials of the right facial nerve. Routine laboratory tests were normal. Chest X-ray and assay of serum calcium, lysozyme, β 2-microglobulin and angiotensin-converting enzyme were normal. No clinical signs of intestinal disease were present. Gastro-intestinal radiological examination and sigmoidoscopy were both normal. A treatment consisting of 100 mg/day azathioprine and 20 mg/day 6-methylprednisolone during 1 year, the latter during episodes of oedema, had no substantial effect on the course of the disease.

It is widely accepted that MRS can only be diagnosed with certainty when there is at least one of the so-called major signs: recurring orofacial oedema and/or facial paralysis associated with histological features of sarcoid-like granuloma [1]. When there is one or more minor signs (plicate tongue, histological aspecific oedema, involvement of cranial nerves, migraine, vegetative orofacial symptoms, psychiatric symptoms during the episodes of oedema), MRS can

only be considered probable or suspected [1]. All the criteria for diagnosis of MRS were fulfilled in the present case. However, differential diagnosis with respect to CD involving the orofacial region required careful evaluation, also as far as medical history was concerned. In our case, we preferred the diagnosis of MRS, on the basis of the unilateral nature of the lesion, the absence of cobble-stoning and ulceration of the oral mucosa, the persistence of oedema and the negative clinical and laboratory findings for recurrence of CD. However, the fact that at different times of her life this patient had such clinically and histopathologically similar diseases as CD and MRS once again raises the question of whether the two conditions may be related. CD and MRS have been linked in several case reports, and it is now accepted that MRS may precede or follow CD, sometimes by many years. The relationship between the two diseases, however, remains unclear [3-8]. Some authors think that MRS and Miescher's cheilitis granulomatosa, now considered the oligosymptomatic form of MRS, are clinical expressions of CD [7, 9-11]. In view of the frequent diagnostic uncertainties, others preferred to coin the term 'orofacial granulomatosis' for any patients with non-necrotizing granuloma infiltrate in the orofacial region [9]. Although the literature gives the overall impression of a close affinity between CD and MRS, in our opinion there are not yet sufficient data to show that they are the same disease. We do not consider either that coining the term orofacial granulomatosis is of any advantage for a correct nosological interpretation. In our opinion it is likely that MRS, CD and perhaps sarcoidosis could represent different clinical manifestations that have a particular common predisposition.

References

- Mainetti C, Masouye I, Harms M, Saurat J-H: (Edème facial solide persis-tant du sujet jeune: syndrome de Melkersson-Rosenthal. *Ann Dermatol Vénéréol* 1994;121:165-170.
- Stosiek N, Birolleau S, Capesius C, Hornestein OP: Chronicité et incertitudes diagnostiques du syndrome de Melkersson-Rosenthal: analyse des modalités évolutives de cinq cas. *Ann Dermatol Vénéréol* 1992; 119: 635-638.
- Greene RM, Rogers RS: Melkersson-Rosenthal syndrome: A review of 36 patients. *J Am Acad Dermatol* 1989;21:1263-1270.
- Worsaae N, Christensen K.C, Bondesen S, Jarnum S: Melkersson-Rosenthal syndrome and Crohn's disease. *Br J Oral Surg* 1980;18:254-258.
- Kano Y, Shiohara T, Yagita A, Nagashima M: Granulomatous cheilitis and Crohn's disease. *BrJ Dermatol* 1990;123:409-412.
- Wiesenfeld D, Fergusson MM, Mitchell DN, Macdonald DG, Scully C, Cochran K, Russell RI: Orofacial granulomatosis: A clinical and pathological analysis. *Q J Med* 1985;54:101-113.
- Brook IM, King DJ, Miller ID: Chronic granulomatous cheilitis and its relationship to Crohn's disease. *Oral Surg* 1983;56:405-407.
- Lloyd DA, Payton KB, Guenther L, Frydman W: Melkersson-Rosenthal syndrome and Crohn's disease: One disease or two? *J Clin Gastroenterol* 1994;18:213-217.
- Allen CM, Camisa C, Hamzeh S, Stephens L: Cheilitis granulomatosa: Report of six cases and review of the literature. *J Am Acad Dermatol* 1990;23:444-450.
- Glanz S, Maceyko RF, Camisa C, Tomecki K: Mucocutaneous presentations of Crohn's disease. *Cutis* 1991;47:167-172.
- Kano Y, Shiohara T, Nagashima M: Association between cheilitis granulomatosa and Crohn's disease. *J Am Acad Dermatol* 1993;28:801.
- KAKG E R ©1997 s. KargerAG-Basel

1018-8665/97/1952-0182\$ 12.00/0 E-Mail karger@karger.ch
Fax+41 61 306 12 34 This article is also accessible online at:
<http://www.karger.ch> <http://BioMedNet.com/karger>