Clin Rheumatol (2008) 27:115–117
DOI 10.1007/s10067-007-0688-9

CASE REPORT

# Magic syndrome and true aortic aneurysm

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Received: 28 May 2007 / Accepted: 19 June 2007 / Published online: 27 July 2007


**Abstract** Magic syndrome is a very uncommon disease, and vascular involvement is exceptional; only one case has been reported in the literature associated to a true aortic aneurysm. The treatment of aneurysms recommended in these patients is based on isolated cases and includes corticosteroids, other immunosuppressant drugs, and surgery. We report a case of a patient with Magic syndrome who developed aneurysm at the end of the aorta during treatment with infliximab, corticosteroids, and cyclosporine and who needed endovascular prosthesis implantation. After 12 months, she suffered an aneurysm of the ascending aorta, dilatation of the sinotubular junction, and severe aortic insufficiency, which forced surgery. During this time, the patient finally died.

**Keywords** Aortic aneurysm · Endovascular prosthesis · Infliximab · Magic syndrome

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## Introduction

Magic syndrome is a very infrequent disease which includes clinical manifestations of Behçet disease and relapsing polychondritis [1]. Vascular involvement in Magic syndrome is very rare, and up until now only two case has been reported in the literature of this syndrome associated to a true aortic aneurysm [2, 3]. The treatment recommended for aneurysms in Behçet disease, and relapsing polychondritis is based on series of isolated cases with variable and reduced number of data, which includes corticosteroids, other immunosuppressant drugs, and surgery [4].

We present the case of a patient with Magic syndrome who developed aneurysm at the end of the aorta during treatment with infliximab, corticosteroids, and cyclosporine. The initial treatment the patient received was three bolus of methylprednisolone, followed by oral prednisone 1 m/kg per day, and four bolus of cyclophosphamide given monthly. Because of the growth and possible rupture of the aneurysm, endovascular prosthesis was implanted successfully at first. One year after the implantation, the patient developed an aneurysm of the ascending aorta, dilatation of the sinotubular junction, and severe aortic insufficiency, which forced surgery; however, in the end, she died.

## Case report

A 29-year-old woman presented with recurrent oro-pharyngeal aphthae and swallowing difficulty, genital aphthae, myalgia, and arthralgia developed during the preceding 10 years. She also presented several episodes of inflammation and later deformity of the soft palate and oropharynx, with disappearance of the uvula and fusion of both

structures, leaving a small communicating orifice between the nasopharynx and oropharynx (Fig. 1). She was treated with daily doses of prednisone (between 7.5 and 20 mg), 100 mg/day azathioprine, inhibitors of bone resorption (alendronate), calcium, colchicine, and pentoxyphiline. Six years after the onset of the disease and because of uncontrolled oropharyngeal aphthosis and inflammation and nasal pyramid deformity (saddle-nose deformity), the treatment with azathioprine was substituted by cyclosporine together with increased doses of prednisone; the patient experienced transient improvement of clinical manifestations; however, she worsened again when steroid therapy was reduced; 1 year later, treatment with infliximab 5 mg/kg (baseline dose, day 15, day 45, monthly) was started, with the remission of the oral aphthae, but she experienced sporadic episodes of arthralgia and arthritis in the wrists and elbows and inflammation of the outer ear.

A magnetic resonance imaging (MRI) scan of the hands showed a slight swelling of soft tissues at the level of the proximal phalange and interdigital space of the third finger and palmar fascia, without bone erosions. Anticitruline antibodies were negative. Two years after starting treatment with infliximab, the patient experienced irritative cough, effort dyspnea, scapula and left shoulder inflammatory pain, regurgitation of liquids, and dysphonia. Otolaryngologic examination revealed a re-epitelization disorder of the vocal cords. Echocardiogram revealed slight aortic and mitral insufficiency and dilation of the sinotubular junction. Chest computed tomography scan done with contrast showed aneurysmal dilation of the aortic arch 5 cm distally to the end of the left carotid artery, with involvement of the left subclavian artery, whose origin was located at the most proximal portion of the aneurysm. Laboratory tests showed reactive C protein value of 9.7 mg/dL, leukocyte count of 14,900 cells/Ul (general formula), hemoglobin count of 11.2 g/dL, mean corpuscular volume of 75.8, and erythrocyte sedimentation rate of 104 mm in the first hour. The patient received three intravenous bolus of 1 g methylprednisolone, and later 1 mg/kg per day oral prednisone, and intravenous bolus of 0.5 g/m$^2$ cyclophosphamide monthly, beside her old treatment with oral cyclosporine 100 mg/day, which was reduced gradually and suspended completely 2 months later. The initiation of this new treatment led to the decrease in the aneurysm size (4.7×4.5 cm) and remission of the clinical manifestations. After receiving the fourth cyclophosphamide bolus and during treatment with 20 mg prednisone, the patient presented to the emergency room because of the reappearance of symptoms. An angio-MRI of the chest showed an increased size of the aneurysm (7.5 cm) with indirect signs of risk of rupture or parietal fissure and stenosis of the left common carotid and subclavian arteries; therefore, the patient was admitted to the Division of Vascular Surgery. Revascularization of the brachiocephalic trunk and left carotid artery was decided by means of a 14×6 dacron prosthesis from the root of the ascending aorta implanted by median sternotomy; previously, a dacron extension was anastomosed to the prosthesis with oblique implant in the prebifurcation of the prosthesis. Finally, a thoracic endoprosthesis was put excluding completely the aneurysm and occluding the left subclavian artery. Intraoperative controls showed absence of flights and a good revascularization of the rest of the supra-aortic trunks. Twelve months postsurgery, the patient was receiving treatment with oral prednisone 7.5 mg/daily and an infliximab bolus monthly, and she started with rest dyspnea and pain in the right shoulder and hemitorax. Transesophagic echocardiography showed an aneurysm of 5.7×5×5.3 cm located in the ascending aorta, which caused the dilatation of the sinotubular junction and severe aortic insufficiency. By using extrabody circulation, an operation was performed, which allowed the implantation of a valve prosthesis tube; this bridged the coronary arteries and substituted the dilated area of the ascending aorta. Technically, the operation was a success, but it was not enough to make the heart function spontaneously. The patient died without responding to resuscitation measures.

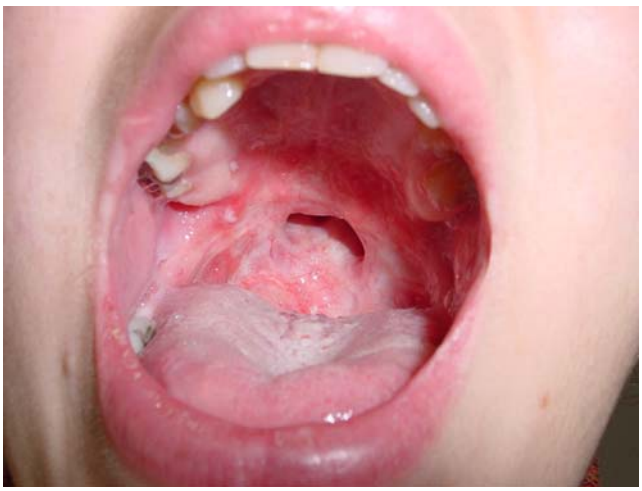

**Fig. 1** A small communicating orifice between the nasopharynx and oropharynx because of inflammation and deformity of the soft palate and oropharynx, with disappearance of the uvula

## Discussion

Cardiovascular complications in Behçet disease present approximately in one of three of the patients and are the main cause of death; arterial involvement (aneurysms) appears in 10–15% of the cases. When large arteries are involved, the most common complications are usually aneurysm or occlusion, the simultaneous appearance being very rare [5]. Both vascular complications occurred in our

patient. At the same time, relapsing polychoncritis is also a very uncommon disease of unknown etiology that can produce cardiovascular manifestations such as aortitis, medium- and large-size vasculitis with aneurysms, valvulitis, pericarditis, and atrioventricular conduction disorders, which has been present simultaneously in some patients; these complications are the main cause of death in these patients [6]. The similarity between the clinical manifestations of the two diseases leads to a common pathogenesis or cause against the cartilage, so that there is a close relationship between both diseases, or Magic syndrome is an overlap in which the course and prognosis of the organ involved shows which disease is the first responsible for the clinical manifestations [1]. In our patient, it would be difficult to distinguish which one is the predominant disease, as both conditions can produce aneurysms. There is limited information about the efficacy of the treatment of vascular lesions, mainly when arteries are involved, which is generally treated with a combination of corticosteroids, other immunosuppressant drugs, and surgery, the latter being preferable after succeeding in maintaining the disease quiescent, although sometimes emergency surgery has been necessary [7]. Our patient, despite being under intensive treatment, developed an aneurysm, and initially, an endoprothesis implantation was necessary to prevent rupture. Nonetheless, in spite of this, an aneurysm of the ascending aorta gave place to the dilatation of the sinotubular junction and severe aortic insufficiency, which open surgery procedures did not improve the patient's clinical situation. Early surgery should be done in inflammatory aneurysms, even the small ones, as rupture can occur at any time; up until now, the surgical treatment of aneurysms or pseudoaneurysms of the aorta in these entities has been the reconstruction with a prosthetic graft, by the anastomosis carried out in disease-free tissues and inactivity periods [8]. Today, in those patients in which open surgery of the aneurysm has failed, endovascular prosthesis is being used with some success [3]. Although, in a retrospective study carried out in patients with Behçet disease and abdominal aneurysms, in which endovascular prostheses are compared with conventional surgery, better results were found in the group of patients in which endoprostheses were done [9]. Several postsurgical therapeutic strategies are being used such as anticoagulation therapy, which does not seem to prevent thrombosis or occlusion of aneurysm; another therapy is corticosteroids, necessary to prevent relapses or even combined with immunosuppressant drugs (azathioprine vs cyclophosphamide) [9].

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