



Orbital Myositis Revealing Crohn's Disease

Boudabbous M¹, Gharbi R^{1*}, Derbel A², Chtourou L¹, Gdoura H¹, Mnif L¹, Marzouk S², Amouri A¹, Bahloul Z² and Tahri N¹

¹Hepatogastroenterology department, Hédi Chaker hospital, Tunisia

²Internal medicine department, Hédi Chaker hospital, Tunisia

*Corresponding author: Gharbi R, Hepatogastroenterology department, Hédi Chaker hospital, Sfax, Email: gharbirania12@gmail.com

Case Report

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Abstract

Orbital myositis (OM), defined by the inflammation of one or more of extra ocular muscles was exceptionally reported in Cohn's disease (CD) especially as initial presentation before the development of intestinal manifestations. We report a case of OM preceding CD. Through our observation and a review of the literature, we specify the epidemioclinical, therapeutic and evolutionary characteristics of this rare entity.

Keywords: Orbital Myositis; Cohn's Disease; Extra Intestinal Signs of Inflammatory Bowel Diseases; Inflammatory Bowel Diseases

Abbreviations: OM: Orbital Myositis; CD: Cohn's Disease; MRI: Magnetic Resonance Imaging; ESR: Erythrocyte Sedimentation Rate; CRP: C-Reactive Protein; CK: Creatinin Kinase; HSV: Herpes Simplex Virus, HCV: Hepatitis C Virus; HBV: Hepatitis B Virus; HIV: Human Immunodeficiency Virus; EBV: Epstein Barr Virus; CMV: Cytomegalovirus; VZV: Varicella Zoster Virus; TSH: Thyroid Stimulating Hormone; TPO: Thyroid Peroxidase Autoantibodies; ANA: Antinuclear Antibodies; ANCA: Anti-Neutrophil Cytoplasmic Autoantibody; MCV: Mean Corpuscular Volume

Introduction

Extra intestinal signs of Cohn's disease (CD) are various including rheumatologic manifestations, cutaneous, ophthalmologic and hepato-biliary involvements. Ocular manifestations are infrequent with prevalence less than 10%. They are mainly represented by: episcleritis, uveitis and scleritis. Orbital myositis (OM), defined by the inflammation of one or more of extraocular muscles was exceptionally reported in CD even more as initial presentation before the development of intestinal manifestations [1,2]. Herein we

report a case of OM preceding CD.

Observation

A 19-year-old female with no significant medical history presented to the hospital in October 2015 with acute convergent strabismus and double vision. She was hospitalized in ophthalmology department and underwent orbital magnetic resonance imaging (MRI) at emergency that was consistent with orbital myositis (Figure 1). Cerebral and medullar MRI was otherwise normal.

The patient received high doses of corticosteroids (2mg/kg/day) for 6 weeks then she was referred to internal medicine department for further investigations. At her admission she had no ophthalmologic or systemic complaints. In her initial biological assessment we found: hemoglobin at 11.5 g/dl, erythrocyte sedimentation rate (ESR): 16mm (H1); C-reactive protein (CRP): 9 mg/l, Creatinin kinase (CK): 42 U/l, LDH: 211 AST/ALT 18 /18 U/l. Infectious investigations were negatives including HSV(herpes simplex virus), HCV(hepatitis C virus), HBV(hepatitis B virus),

HIV(human immunodeficiency virus), EBV(Epstein Barr virus), CMV(cytomegalovirus), VZV(varicella zoster virus), ParvoB19, VDRL-RPR,, bartonella, rickettsia, toxoplasmosis and tuberculosis. Thyroid function tests including thyroid stimulating hormone (TSH), thyroid peroxidase autoantibodies (TPO), thyroglobulin auto antibodies, TSH-receptor auto antibodies were negatives. Immunologic analysis revealed negative results (antinuclear antibodies (ANA), anti-neutrophil cytoplasmic autoantibody (ANCA),

anti cardiolipin antibodies and kit- myositis). Her orbital finding was attributed to idiopathic orbital myositis and the patient received 6 months of steroids then progressively stopped. One month after stopping corticosteroids, on May 2016 the evolution was marked by the recurrence of myositis which became bilateral. Biopsy of internal orbital muscle showed myofiber regeneration with necrotic lesions, and prominent endomysial inflammatory infiltrate (Figure 2).

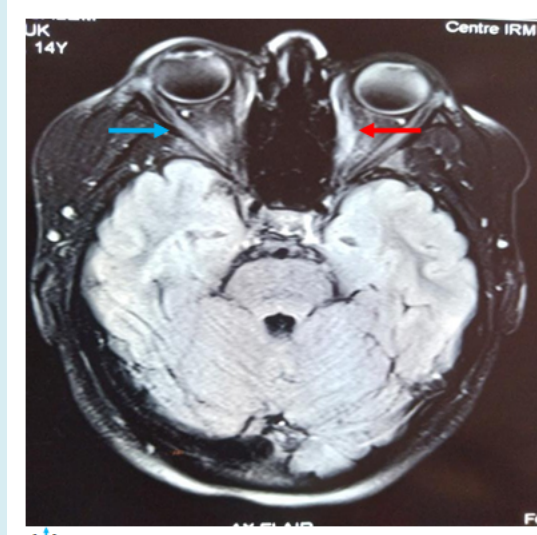


Figure 1: MRI orbital Trans axial view. Pathologic contrast enhancement and thickening in extraocular muscles: left medial rectus (↑) right lateral rectus (↑).

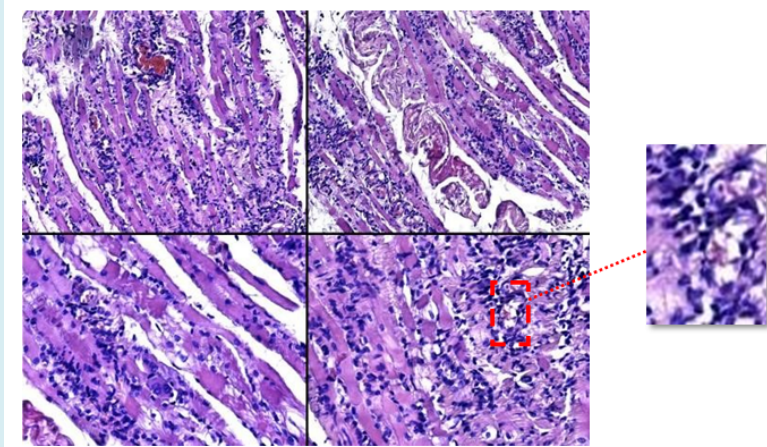


Figure 2: Haematoxylin-eosin staining of internal orbital muscle, endomysial inflammatory infiltrate.

Those findings were concordant with orbital myositis. Therefore, adjunction of azathioprin has been started (2.5 mg/kg/day) with good progress and it was maintained for 3 years. On April 2019 decision was to stop azathioprin and maintain low doses of steroids. On her follow-up, in

December 2020, she presented diffuse abdominal pain with constipation. Then she was referred to gastroenterology department for endoscopic explorations. At her admission, physical examination found tenderness on palpation of the iliac right fossa. Perianal and rectal examination was normal.

Laboratory analysis showed anemia with haemoglobin at 9.9 g/dl with mean corpuscular volume (MCV) at 74 fl and elevated CRP at 131 mg/l. The patient underwent ileocolonoscopy revealing ulcerative ileocecal valve with stenosis and two fistulous paths (Figure 3). Colonic mucosa was normal. Histologic examination showed important acute and chronic inflammation with ulcerations of the Bauhin's valvula and the anal canal. There is no evidence of necrotizing

vasculitis or granulomas. The upper endoscopy was also practiced revealing antral follicular gastritis and erosive bulbous duodenitis. Biopsy of the Duodenum was normal with preservation of villous architecture and no increase intraepithelial lymphocytes. Biopsy of gastric antrum / body didn't reveal histopathologic abnormalities. The MR enterography showed active fistulizing Cohn's (Figure 4).

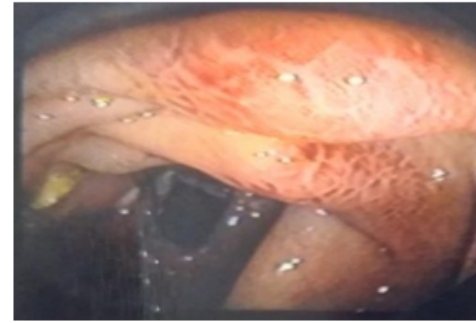
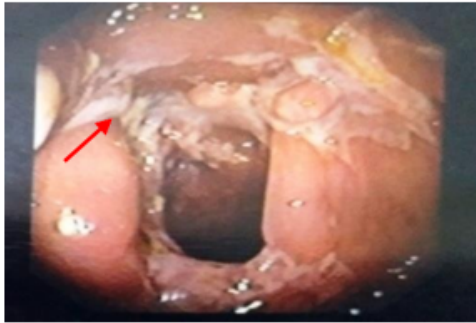


Figure 3: Ileocolonoscopy revealing ulcerative ileocecal valve with stenosis (↑).

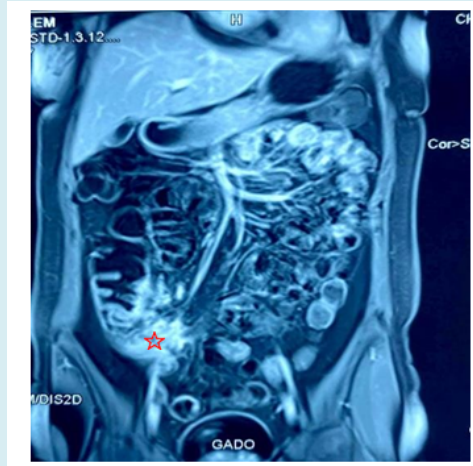


Figure 4: Coronal section of MR Enterography. Pathologic terminal ileum (★).

MR Enterography showing active fistulizing Cohn's disease: bowel wall thickening of the terminal ileum extended over 6 cm ileum, multiple fistulous paths without abscesses, jejunal wall thickening extended over 18 cm, sclerolipomatosis. Diagnosis of Crohn disease was retained and orbital myositis had been attached to inflammatory bowel disease. The patient was treated by combined treatment face to her young age and fistulizing character of the luminal disease. Double therapy was based on azathioprine associated with TNF alpha inhibitor. The initial evolution was favorable (current follow-up of 1 month).

Discussion

Ocular manifestations occur in 2-5% of patients with crohn's disease and can precede luminal diseases [3]. The most common orbitary manifestations are: episcleritis, anterior uveitis with a frequency between 2 -4% and scleritis occurring in 1% of cases [3]. Orbital myositis is a very rare extra intestinal manifestation of crohn's disease, with only few cases reported in the literature [4]. Pathogenesis is unidentified, but is possibly correlated to underlying immune-mediated processes. In our case the diagnosis of CD was retained after 5 years than the ocular myositis. Coming

back to our investigations showed negative findings and ruled out all causes of OM (viral, bacterial, vascular, trauma, lupus, and sarcoidosis). Then OM couldn't be explained by another cause other than CD. The evaluation of the few cases of CD with OM found female predominance, in half OM precedes the onset of CD and the involvement was bilateral in also 50% of cases [5]. As you noticed in the observation, ocular signs respond well to corticosteroids and azathioprine as in the majority of patients who were evaluated in literature reviews [6].

Conclusion

We report a rare case of orbital myositis that precedes gastro intestinal manifestations in Crohn's disease. This observation is original for two reasons: the rarity of this type of ocular involvement during IBD and its occurrence several years before the appearance of the digestive localization.

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