Short communication

Palpebral ptosis, a rare ocular manifestation of Crohn’s disease

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ABSTRACT

Clinical case: An 18-year-old-woman presented with abdominal pain, diarrhea, and ptosis in her left eye. Nuclear magnetic resonance imaging (NMR) and the study of cerebrospinal fluid detected no abnormalities. Colonoscopy revealed a Crohn’s disease (CD) confirmed by histological examination of biopsies. Ocular symptoms improved after appropriate treatment of the underlying CD.

Discussion: Inflammatory bowel disease (IBD) is associated with a wide variety of extra-intestinal manifestations, in fact it may be considered a systemic disorder with predominantly gastrointestinal tract manifestations. Extra-intestinal manifestations of inflammatory bowel disease (IBD) occur in one-third of patients. Ocular complications are infrequent, occurring in less than 10% of cases, but can be associated with significant morbidity, including blindness. Ocular symptoms may precede a diagnosis of IBD. We report the first case of palpebral ptosis associated with Crohn’s disease.

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Ptosis palpebral: manifestación ocular inusual de la enfermedad de Crohn

RESUMEN

Caso clínico: Se presenta el caso de una mujer de 18 años con un cuadro de diarrea, dolor abdominal y ptosis palpebral izquierda. La resonancia nuclear magnética cerebral (RMN) y el estudio del líquido cefalorraquideo fueron normales. La colonoscopia estableció el diagnóstico de enfermedad de Crohn (EC) confirmándose en el estudio histopatológico. La afectación ocular mejoró tras el adecuado tratamiento de la EC.

Discusión: La enfermedad inflamatoria intestinal (EII) se asocia a una amplia variedad de manifestaciones extraintestinales, de hecho puede ser considerada como una enfermedad sistémica que afecta predominantemente al tracto gastrointestinal. Las manifestaciones


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extraintestinales acontecen hasta en una tercera parte de los pacientes afectos de EII. Las complicaciones oculares son infrecuentes, afectando a menos del 10% de los casos, pero en ocasiones comportan una significativa morbilidad, incluyendo la ceguera. La sintomatología ocular puede preceder al diagnóstico de EII. Informamos el primer caso de ptosis palpebral asociado a EC.

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Introduction

Crohn’s disease (CD) is a chronic intestinal inflammatory disease (CIID) with multifactorial and polygenic origin that involves any part of the digestive tract from the mouth to the anus. The most affected location is ileocolic. CD is a complex entity for which a cause/effect relationship has not been established with a single agent or molecular alteration. The most widely accepted etiopathogenic theory suggests that intestinal inflammation is the consequence of an inadequate result of innate and adaptive immunity in genetically predisposed patients. Bacterial flora plays a crucial role. It has been observed that up to 30% of CIID patients exhibit extraintestinal involvement which could precede digestive expressions, accompany them or appear independently. These involvement are more characteristic when the colon is involved more frequently than other locations. The most frequent expressions are musculoskeletal followed by mucocutaneous and ocular expressions. The latter occur in 10% of cases but in some cases are associated to significant morbidity, particularly because they could go unnoticed by clinicians.

This paper presents a case of a patient with left palpebral ptosis associated to chronic diarrhea and abdominal pain located in the right iliac fossa. The wide group of diagnostic possibilities included the option of mononeuritis associated to intestinal inflammatory disease (with a previous study of other secondary causes) as an extraintestinal expression. After a systematic review the authors were unable to find a description of any such case.

Case report

This is a case report of a female, aged 18 years, without relevant medical or surgical history, who denied having toxic habits or allergy to drugs. She was referred to gastroenterology due to chronic diarrhea condition without pathological products accompanied by colic-like abdominal pain and undetermined loss of weight. In addition, the patient exhibited left palpebral ptosis beginning one month before the gastrointestinal symptoms. The physical examination did not reveal relevant pathological data with the exception of said left palpebral ptosis and pain in the right iliac fossa upon palpation. Perianal examination did not reveal pathological data. Outpatient study was carried out which showed iron deficiency anemia, Hb 9.8 g/dl, ferritin 7 ng/dl, mean corpuscle volume (VCM) 72.5 fl, globular sedimentation speed (VSG) 40 mm and C reactive protein 2.03 mg/dl (reference <0.3), hypoproteinemia with overall proteins of 5.9 g/dl, antinuclear antibodies (ANA), anti-smooth muscle antibodies (anti-KLM), and anti-mitochondrials (AMA) negative; blood in feces with 3 positive samples. Gastrointestinal transit was performed which showed small intestine loops with wall edema and marked spiculation of the terminal ileum mucosa. Subsequently, outpatient colonoscopy was requested which revealed multiple around dish ulcerations which were biopsied. The histological study identified granuloma. These findings were compatible with CD. Treatment with prednisone was established at a dose of 1 mg/kg/day. Hospital admission was advised and accepted by the patient to complete the palpebral ptosis study (Fig. 1).

The ophthalmological examination revealed symmetric and photoreactive pupils, normal accommodation, ocular motility in all spaces without alterations and notably left palpebral ptosis. Eyelid raising function was 11 mm. Margin–reflex distance (MRD): 2.5 mm. Distance from palpebral edge to fold: 10 mm. Distance from the corneal reflex to inferior palpebral edge: 7 mm. No ocular proptosis. Ocular fundus without alterations. Iris homochromy. No diplopia in examination. No other type of focus of the central or peripheral nervous system was associated. The analytic study showed iron deficiency anemia and hypoproteinemia, normal kidney and liver profile, reactive proteins C 0.64 mg/dl, urine normal, anti-acetylcholine receptor antibodies negative, TSH 2.76 U/ml, ANA negative, rheumatoid factor negative, ANCA negative, angiotensin-converting enzyme negative, anti-celiac antibodies negative, vitamin B12 and folic acid normal, serology to VHB, VHC, VIH, LUES, CMV, Coxiella burnetti, brucella negative. Magnetic resonance with bilateral and cranium orbits contrast without pathological findings (Figs. 2 and 3). Chest ecocardiography and x-ray normal, arbitrary electroneuromiogram normal. Cerebrospinal fluid normal, without presence of oligoclonal IgG strip. Interconsultation with Ophthalmology: palpebral ptosis is due to the involvement of superior eyelid elevator, ocular fundus normal.

Fig. 1 – Left superior eyelid ptosis.
The patient was released with the diagnosis of superior eyelid elevator mononeuritis secondary to CD. Levels of thiopurine methyltransferase (TPMT) were requested and azathioprine at a dose of 2.5 mg/kg/day, achieving clinical and analytical remission of the luminal disease and substantial but not complete improvement of palpebral ptosis.

Discussion

CD is a chronic inflammatory process which can involve any segment of the digestive tract. It is a highly heterogeneous disease due to the broad range of clinical expressions. The natural history of the disease is characterized by the presence of inflammatory activity outbreaks with periods of latency or remission even though the clinical behavior of the disease generally evolves toward more aggressive forms. The areas which are predominantly affected are ileocolic (40%), ileal (30%) and colon (25%). Only 5% of patients exhibit high gastrointestinal involvement. Gastrointestinal clinic is highly variable and comprises a broad range including fever, abdominal pain, chronic diarrhea and weight loss as well as fistula or abdominal abscess.

In addition to the gastrointestinal expressions, between 25% and 35% of patients exhibit one or more extraintestinal expressions the most relevant of which due to their clinical significance include osteoarticular expressions, mucocutaneous, ocular and hepatobiliary expressions, with descriptions of less frequent expressions such as a predisposition to nephrolithiasis and cholelithiasis.1

Said extraintestinal expressions could be the first expression of CD, either related or unrelated to luminal disease activity. Peripheral arthritis type I, episcleritis, nodal erythema and afose stomatitis are dependent on the activity of the baseline EII and, less dependent on said activity, peripheral arthritis type II, uveitis and pyoderma gangrenosum.

The pathology of central and peripheral nervous system disorders associated to CD is not well defined. A combination of factors has been accepted, including vitamin and mineral deficiency due to malabsorption, formation of metabolic toxic agents, infections due to the immunosuppression state, side effects of treatment, associated thromboembolic events and immunological alterations.2

The actual incidence of neurological complications is unknown. Reported series range between 0.25% and 35.7%. Described neurological expressions include ischemic ictus, multiple sclerosis, epilepsy, loss of neurosensory hearing, papilla edema, myelopathy, peripheral neuropathies, myasthenia gravis, miopathies, psychiatric expressions, neurological expressions due to malabsorption or treatment delete serious effects and vasculitic events.3

The first ocular expressions were described as early as 1925 by Crohn in 2 patients with corneal infiltrates and conjunctivitis. Ocular complications do not seem to depend on the extension of the intestinal disease and frequently appear in the first few years after diagnosis. In up to 68% of subjects with CIID and ocular symptoms extraintestinal symptoms coexist, particularly in carriers of HLA-B27 who associate uveitis to arthritis and ankylopoietic spondylitis. The most frequently described expressions in the literature are episcleritis (2–5%), and uveitis (0.5–3%). Additional ocular expressions include retinal complications such as central retinal vein or artery occlusion, retina vasculitis and macular edema. Descriptions also include corneal ulcers, subconjunctival modules, subepithelial keratitis, central serous chorioretinopathy and Salzmann nodules.5 At the level of the orbit, there are reports of individual cases of orbital pseudotumor and orbital
myositis, as well as one case of bilateral palpebral pyostomatitis and one internal palpebral ulcer, among other reports of first expressions of CD. Complications derived from the use of steroids include cataracts, glaucoma and exceptionally papillary edema secondary to benign intracranial hypertension.

Treatment of neurological involvement is focused on controlling the baseline disease with existing and applied therapies and if applicable to treat secondary complications and symptoms.\(^5\)

In the case of this report, the patient exhibited vasculitic clinic in the form of superior eyelid elevator mononeuritis which preceded gastrointestinal symptoms enabling us to establish an association after reasonably discarding other secondary etiologies and because the patient responded positively to the primary disease treatment. To our knowledge and after reviewing medical literature this is the first described case associating CD and palpebral ptosis.

**Conflict of interest**

No conflict of interests has been declared by the authors.

**REFERENCES**