Bilateral idiopathic orbital myositis in an infant

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ABSTRACT
Idiopathic orbital myositis is an unspecified isolated inflammation of extra-ocular muscles. It is uncommon in children, especially in infants, and is a source of real diagnosis problems. We report the case of a 22 months old infant who was admitted for a sudden swollen eyelid with ptosis and painful bilateral exophthalmos. The orbital computed tomography revealed bilateral and diffuse hypertrophy of extra-ocular muscles responsible of bilateral exophthalmos grade III. The etiologic survey was negative, thus the diagnosis of idiopathic myositis was kept. The patient recovered completely with corticosteroids. Through this case we describe the clinical, radiological, and treatment of idiopathic orbital myositis.

Keywords: Orbital inflammation; Orbital myositis; Exophthalmia

INTRODUCTION
Idiopathic orbital myositis (IOM) is a rare and special entity defined by the non-specific and localized inflammation of extra-ocular muscles1. It is a subtype of nonspecific orbital inflammation primarily involving one or more of extra-ocular muscles without identifying local or systemic causes2. It’s rarely described in children and exceptionally in infants and it often causes real problems of differential diagnosis with malignancies especially in their acute form1,2. We report a case of bilateral IOM in a 22-month-old infant, through which we describe the clinical, radiological, and treatment of this affection.

CASE REPORT
History: Female infant aged 22 months, with no past medical history, especially with no history of recent trauma or symptoms of thyrotoxicosis, presented 21 days before admission a swelling of the left upper eyelid gradually increasing in volume, followed a week later by a swelling of the right upper eyelid with pain mobilization of eyeballs associated with visual impairment without tearing or fever or other clinical signs. Examination: The clinical examination at admission found a conscious patient in a good general condition, afebrile, with a normal weight, size and blood pressure, and without periorbital palpable mass. Ophthalmologic examination revealed a bilateral eyelid edema associated with local inflammatory signs, irreductible bilateral exophthalmos.

Figure 1: Bilateral eyelid edema associated with local inflammatory signs and bilateral exophthalmos with ptosis.

bilateral ptosis (Figure 1), limitation of abduction of the left eye, the cornea and its crystal are clear, the papilla is normal with a good macular reflection, the pupillary light reflex is present as well as eye tracking, visual acuity is preserved. The rest of the physical examination was normal. Investigations: Orbital computed tomography objectified bilateral and diffuse hypertrophy of extra-ocular muscles responsible of bilateral exophthalmos grade II without individualization of intra-orbital process or abnormality of orbital frames or optic nerve (Figure 2). Complete blood count, electrolytes levels, sedimentation rate, muscles enzymes, thyroid and immunological laboratory tests were in normal ranges. Management: The diagnosis of bilateral IOM is retained on the clinical findings and imaging after eliminating other possible etiologies. The patient is put under oral corticosteroid: prednisone at a dose of 2 mg/kg/day with a dramatic improvement in the third day of treatment with the disappearance of all clinical signs after one week of treatment (Figure 3). She was kept in the same dose for one month until complete recovery of eye movements and disappearance of ptosis and edema (Figure 4). The digression was undertaken gradually over three months with a favorable outcome. Now we are 6 months of remission without recurrence.

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Figure 2: Orbital CT Scan in axial (A and B) and coronal (C) sections showing a bilateral and disseminate hyper trophy of oculomotors muscles responsible of exophthalmos grade II.

Figure 3: Disappearance of all clinical signs and normal appearance of the eyeballs after one week of treatment.

DISCUSSION

IOM is a rare disease characterized by inflammation of the extra-ocular muscles, described for the first time in 1903 by Gleason. It represents 10% of non-specific inflammation of the orbit. Its prevalence and incidence are difficult to assess because of the small number of cases and reported series in the literature. It is extremely rare in pediatric age group and may cause real diagnostic problems. It occurs most frequently in young adults in the third or fourth decade of life, with a female predilection. The most common causes of orbital myositis are infection, Basedow disease, granulomatosis with polyangiitis (Wegener's granulomatosis), and more rarely autoimmune diseases. IOM is a diagnosis of exclusion based on clinical, radiological and, where required, histopathological findings. Its pathogenesis remains unclear, but the autoimmune theory is the most likely. Three main hypotheses have been advanced. The first assumption is a disproportionate immune response to an infectious agent (viral or bacterial) affecting the upper airway, the second hypothesis suggests an autoimmune origin due to the association with some autoimmune diseases, and the third suggests a similar mechanism to that of idiopathic mediastinal sclerosis namely a fibroproliferative disorder. However, none of these assumptions has been formally demonstrated. On the pathophysiology, muscle damage is done in three phases: initially, the inflamed muscle grows while maintaining normal function. It becomes secondarily paretic and it subsequently evolves to muscle retraction that leads to muscle fibrosis. The most often affected muscles are the internal and external muscles of the eye. Muscle involvement is usually unilateral, bilateral forms are very rare and described in only 5 to 12% of cases. Clinically, it is most often an acute disease, but there are subacute or chronic forms. It combines retro or periorbital pain exacerbated by eye movements that can be associated with diplopia and oculomotor limitation because of the paresis. Other signs such as conjunctival hyperemia, eyelid and periorbital edema, ptosis, chemosis, diplopia and exophthalmos are possible. Visual acuity is usually preserved. Due to, the lack of specificity of clinical presentation, orbital myositis require extensive investigations for diagnosis, prompted by fear of a malignant tumor pathology. In our case, even if clinical presentation evoked orbital myositis we made a detailed assessment to rule out other possible causes. The differential diagnosis is primarily with orbital rhabdomyosarcoma, lymphoma and metastases of neuroblastoma, thyroid related orbitopathy, infection (orbital cellulitis), sarcoidosis, vascular tumors, cystic lesions, infectious diseases (toxoplasmosis, tuberculosis, toxocariasis...) and autoimmune diseases. The most common presentation form is acute and unilateral, bilateral forms are exceptional and need to search for other etiologies, particularly thyroid ophtalmopathy. Orbital imaging is a valuable tool in evaluating suspected orbital myositis. Computed tomography is the examination of choice. The muscle appears thickened as a whole including the anterior tendon insertion and is enhanced by the injection of contrast. Sometimes infiltration of the periorbital fat gives the muscle irregular boundaries. Muscle involvement is usually unilateral but may be bilateral in 5% of cases. Several muscles can be achieved, hence the interest to perform axial and coronal sections of the orbit. Bilateral and/or multi-muscle damage is a predictor of a higher risk of recurrence. The orbital ultrasound is much less often used because of its character operator-dependent. However, it can provide a valuable initial examination because it is rapid, easy to perform and non-invasive, but do not allow for detailed analysis of adjacent structures. The magnetic resonance imaging is proposed only as second-line, mainly to specify the extra-orbital extension of a process, especially to the cavernous. The diagnosis of IOM is based on the sudden onset of painful proptosis, restricted eye movements causing diplopia, with the presence of a thickening of extra-ocular muscles in orbital imaging and rapid response to corticosteroids, without identifiable local or systemic causes. Diagnostic biopsy is not usually necessary. In typical cases, it is possible to postpone the histopathological analysis. However, it is indicated in cases with atypical presentation, poor

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response to corticosteroid treatment, transition to chronicity or suspected tumor infiltration. In our case, the diagnosis of bilateral IOM is retained on the clinical findings and imaging after eliminating other possible etiologies. The treatment is not codified to date. Its purpose is to interrupt the inflammatory process, preserve vision and mobility of the eyeball and prevent progression to fibrosis. Systemic corticosteroid therapy is the cornerstone of managing IOM. Corticosteroids are recommended as first-line at a rate of 1 to 2 mg/kg per day of prednisone for two to four weeks, and then a digression must be made slowly over several weeks to avoid recurrence. The clinical response is so fast that many authors make a diagnostic criterion as in our case. In refractory forms to corticosteroid therapy, treatment requires the use of immunosuppressive drugs (methotrexate, cyclosporine, rituximab, cyclophosphamide or anti-TNF), and intravenous immunoglobulin. The monoclonal antibody and radiation can sometimes be a supplement or an alternative to corticosteroids. The evolution can be spontaneously favorable. The treatment is however recommended for patient comfort and to prevent the occurrence of complications (muscle contraction). Siatkowski et al. reported a 68% rate of complete response with corticosteroids. The recurrence rate varies from 0-50% in the literature. Predictors of recurrence identified are: male gender, inflammation of several muscles or bilateral disease, poor response to initial therapy, and the absence of exophthalmos. The evolution can be spontaneously favorable. The treatment is however recommended for patient comfort and to prevent the occurrence of complications (muscle contraction). Siatkowski et al. reported a 68% rate of complete response with corticosteroids. The recurrence rate varies from 0-50% in the literature. Predictors of recurrence identified are: male gender, inflammation of several muscles or bilateral disease, poor response to initial therapy, and the absence of exophthalmos. The evolution can be spontaneously favorable. The treatment is however recommended for patient comfort and to prevent the occurrence of complications (muscle contraction). Siatkowski et al. reported a 68% rate of complete response with corticosteroids. The recurrence rate varies from 0-50% in the literature. Predictors of recurrence identified are: male gender, inflammation of several muscles or bilateral disease, poor response to initial therapy, and the absence of exophthalmos.

CONCLUSION
Orbital myositis in children is uncommon. It may cause real diagnostic problems and needs to be differentiated from other orbital diseases especially malignancy. The diagnosis should be considered whenever we are confronted with orbital pain without fall visual acuity. The OIM is an eliminating diagnosis. Faced to a child’s exophtalmos, other differential diagnosis must be excluded by an advanced etiologic survey. However, additional tests to find an etiology should never delay the therapeutic management.

REFERENCE