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LETTER TO EDITOR (VIEWERS CHOICE)

A DIFFERENT KIND OF INTERMITTENT HEADACHE IN PEDIATRICS

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An orbital pseudotumor is a non-neoplastic, inflammatory condition of the orbit, often idiopathic, but possibly autoimmune, triggered by infection or another immune processes.[1,8] It's the third most common orbital condition, following thyroid orbitopathy and lymphoproliferative disorder.[9] In contrast to adults, paediatric cases typically exhibit a bilateral pattern and constitutional symptoms. Common clinical includes a palpable mass, reduced ocular movement, eyelid swelling, pain, proptosis and increased intraocular pressure.[2] Affected tissues include the rectus muscles (myositis), orbital cellulitis and dacryoadenitis.[7] Diagnosis involves clinical history, detailed physical, ophthalmological assessments, laboratory investigations, and imaging studies including computed tomography (CT) and magnetic resonance imaging (MRI) of the orbit and facial sinuses with contrast enhancement. Open biopsy is recommended when suspicious of malignancy or in poor response to corticosteroid treatment.[2,4,5] The differential diagnosis includes any pathology causing an acute orbital condition; however, in children, this entity is most often confused with orbital cellulitis and thyroid eye disease.[2,3,5] Oral systemic corticosteroids are the preferred treatment, with rapid improvement being diagnostic. This paper discusses a case of orbital pseudotumor and its management.

A 7-year-old female from Angola, living in Portugal presented to the emergency department with a 15-day history of left frontal headache causing nighttime awakening, intermittent abdominal pain and vomiting. The patient was diagnosed with *streptococcal* tonsilitis a month prior. On examination, there was a subtle left proptosis, mild pain with rightward gaze with normal neurological and ophthalmological assessments. There were no other symptoms associated, included diplopia or fever. Brain CT revealed enlargement of the left lateral rectus muscle, suggesting expansive lesion or myositis. After discussion with the ophthalmologist, the child was initially treated with non-steroidal anti-inflammatory drugs (NSAIDs) and re-evaluated in 48 hours. The headache improved, but proptosis persisted, prompting further investigation. Initial laboratory investigation with a complete blood count, biochemistry, C-reactive protein and sedimentation rate were normal, as well as further etiological investigation (Table 1). A CT of the orbits revealed a 9 mm thickening of the lateral rectus muscle, suggesting orbital myositis or a space-occupying lesion. MRI confirmed 7 mm muscle thickening with abnormal T2 and T1 signals. After consultation with paediatric oncology, no biopsy was performed, and idiopathic orbital myositis was diagnosed. Corticosteroid therapy (1.5 mg/kg/day prednisone with slow tapering) was started, with gradual improvement. The patient was discharged after 7 days, and a follow-up CT scan showed resolution of the orbital changes. At 10 months, the patient had no relapse with normal physical and neurological examination. This case highlights the challenges in diagnosing Idiopathic Orbital Inflammation in children, as it is rare (6-17% of paediatric orbital inflammatory diseases) and must be differentiated from conditions like infectious (cellulitis), neoplastic (rhabdomyosarcoma, leukemia), immune (Sjogren's), thyroid ophthalmopathy and granulomatous (sarcoidosis).[10] Although the laboratory tests didn't indicate an infectious disease, the orbital CT scan showed an inflammatory process involving the extraocular muscles. The MRI subsequently validated the diagnosis of an inflammatory pseudotumor in its myositic form. Therefore, corticosteroid therapy was initiated with a satisfactory clinical progression. Performing a biopsy could be important, especially to rule out neoplastic causes, however, suggestive imaging data, the inherent risks of the procedure, and the favourable response to corticosteroid therapy allowed for the diagnosis.[10] The value of ultrasound, which is easily accessible and cost-effective, might be important in excluding a neoplastic mass allowing for a differential diagnosis between orbital myositis and thyroid-associated ophthalmopathy, with involvement of muscle insertion. This condition has been linked to previous respiratory infections or autoimmune diseases suggesting the hypothesis that the underlying mechanisms may be mediated by autoimmunity. It is difficult to establish a definite causal relationship because the exact pathogenic mechanisms are still widely unknown. Therefore, the association is based on the simultaneous presence of a clinical diagnosis and a systemic condition that can predispose to an inflammatory process affecting the extraocular muscles. In our case we have a previous history of *streptococcal* tonsilitis that support this theory. The first line treatment consists in corticosteroid therapy with prednisone at a dose of 1 to 1.5 mg/kg/day for one

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to two weeks, with gradual tapering over 4 to 12 weeks. Other therapeutic possibilities with proven efficacy in adults, should be considered when no response to first line treatment, including immunosuppressive drugs (methotrexate, rituximab, infliximab) or radiotherapy. Surgical intervention plays a crucial role, especially in muscle involvement-related sequelae. The long-term prognosis of orbital myositis is highly variable, influenced by factors such as the initial extent of orbital involvement, the frequency of recurrences, and the response to treatment. Delayed treatment may lead to irreversible changes in ocular mobility and potential optic nerve damage. This child had a very favourable outcome, remaining asymptomatic. It is also important to be aware that recurrence rate is more common in children than in adulthood, so monitoring should be strict.

Table 1. Complementary etiological investigation.

- Immunoglobulin (Ig) G: 1355mg/dL, IgA: 160mg/dL, IgM:85mg/dL, IgE: 108UI/mL
- Anti-Neutrophil Cytoplasmic Antibody (ANCA): negative
- Rheumatoid Factor: <10.0 UI/mL
- Anti-Nuclear Antibody (ANA): negative
- Thyroid-Stimulating Hormone (TSH): 1.13mUI/L, Free Thyroxine (FT4): 14.5 pmol/L, Anti-Thyroid-Stimulating Hormone Receptor Antibody (TRAb): < 1.50U/L
- Angiotensin-Converting Enzyme (ECA): 11.1U/L
- Interferon-Gamma Release Assay (IGRA): negative
- Chest X-ray: no abnormalities

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