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An Exotropia Revealing Idiopathic Juvenile Arthritis in A Ten-Years-Old Child

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Abstract:
Uveitis is observed in 30% of Anti-nuclear Antibodies (ANA) positive patients with juvenile idiopathic arthritis (JIA) [1] and is an important cause of ocular morbidity in childhood and beyond. This is a 10-year-old boy, with a history of recurrent unpainful red right eye, with inflammatory-type arthralgia of the right knee, Consulted for exotropia of the right eye. On examination, we find visual acuity with positive light perception, retro-descemetic precipitates, iridolenticular synechiae, posterior subcapsular cataract, and a total retinal detachment with ischemic and frozen retina. Ocular ultrasound shows total retinal detachment. with a rigid-leaved V-shaped retina without any intraocular tumor. A retinal fluorescein angiography did not find vasculitis lesions in the contralateral eye. The diagnosis of juvenile idiopathic arthritis was retained in front of the inflammatory joint involvement and a positive level of ANA.

Keywords: *juvenile idiopathic arthritis, uveitis, retinal detachment*

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I. Background:

JIA is considered an autoimmune disease, potentially resulting from an abnormal immunologic response caused or triggered by environmental factors such as infection or trauma in a genetically predisposed subject. Adaptive immune activation against self-epitopes has been suggested, and a typical synovial membrane inflammation has been observed [2–3]. Younger ANA positive patients are at highest risk of developing ocular complications such as chronic uveitis, which may occur in 10 to 15% of children with psoriatic JIA [4].

II. Observation:

Our patient is a young 10-years old child, he Consulted for exotropia of the right eye. Past Ocular History : Recurrent Unpainful red right eye. Past Medical History found a Born at term by repeat C-section after an uncomplicated pregnancy, an Inflammatory-type arthralgia of the right knee, evolving since 8 months. Family History found Positive for paternal uncle with psoriasis. Eye examination: Visual Acuity: 20/20 left eye OS, and positive light perception right eye OD, Current refraction: +1.5.00 sphere OS. On the Slit Lamp Examination of the left eye a Normal lids and lashes clear cornea were found,But on the right eye a retrodescemetic precipitates ,anterior chamber with 1+ flare OD ,with no cell. Pupils: 5 mm in dark, 3 mm in light OS, posterior synechiae and pigment deposits on anterior lens capsule in (OD) (Figure.1). Lens: clear OS posterior subcapsular cataract (OD) .Dilated Fundus Exam: total retinal detachment with ischemic and frozen retina (OD) (Figure.2) , normal retina ,normal disc, macula and vessels(OS). Extraocular movements: non alternating exotropia (OD).Ocular ultrasound shows total retinal detachment. with a rigid-leaved V-shaped retina without any intraocular tumor (Figure.3).

Biological testing:

ANA	Positive at 1:160, diffuse pattern
Angiotensin-1 converting enzyme	43 U/L (normal 8-52 U/L)
Lyme disease antibody (IgG/IgM)	Negative
Erythrocyte sedimentation rate	16 mm/hr (normal 0-20 mm/hr)
HLA-B27 typing	Negative
Rhumatoid factor	Negative

The diagnosis of juvenile idiopathic arthritis was retained in front of the inflammatory joint involvement and a positive level of ANA. The treatment of this child consisted of symptomatic treatment of his

arthritis with non-steroid anti-inflammatory alone, avoiding the use of systemic corticosteroids since he is a monophthalmic child. Surgical abstention for his retinal detachment with rigorous, with a quarterly ophthalmological monitoring of the contralateral eye.

III. Dicsussion

: Ocular inflammation is commonly associated with juvenile idiopathic arthritis, most commonly in the oligoarticular category, in which 2-4 joints are involved, within the first 6 months of diagnosis. female sex, positive ANA, and negative rheumatoid factor represent additional risk factors. JIA-associated uveitis in children can present with a quiet-appearing eye. Because of its insidious progression, it is important to follow recommended uveitis screening guidelines for children with JIA. After initial ophthalmologic examination at the time of JIA diagnosis, the frequency of follow-up eye exams in patients is determined by their risk category (Cassidy et al., 2006). Patients with High-risk, having positive ANA, are 6 years old or less at onset of joint disease and have had JIA for 4 years or less should be examined every 3 months. Moderate and low risk patients are determined based on a combination of ANA status, age at onset, and duration of disease (Cassidy et al., 2006). Patients with moderate risk should be examined every 6 months, and low risk patients should be examined every 12 months[5,6]. The treatment of JIA-associated chronic uveitis is largely based on clinical course, but basic recommendations exist to guide therapy[7]. Patients presenting with uveitis in the setting of JIA are initially started on topical corticosteroid. If there are prognostic factors of impending vision loss such as poor initial vision, hypotony, glaucoma, cataract, macular edema, or dense vitreous opacities, it is also recommended that systemic corticosteroids are begun, at a dosage of 1-2 mg/kg, and tapering to <0.15 mg/kg within 4 weeks. Oral corticosteroids should be limited to 3 months use secondary to systemic complications, and similarly, topical corticosteroid should be limited to no more than 3 drops per day for chronic use due to risk of cataract and glaucoma. If the uveitis remains active or reactivates within these guidelines, steroid sparing agents should be begun.

Pato et al., 2011, have studied the effect of immunosuppressants and biological therapies in treating posterior autoimmune uveitis and its complications. In this systematic review, they found several studies which have effectively treated uveitis. The agents that were found to be effective include cyclosporine, infliximab, daclizumab, INF-alpha-2a, acetazolamide and methylprednisolone[8]

IV. Conclusion:

Juvenile idiopathic arthritis is the main etiology of chronic uveitis in children. As uveitis often goes unnoticed, it should be detected and treated as soon as possible. Multidisciplinary collaboration as well as a dialogue with parents are necessary. The treatment will be quickly adapted, with therapeutic escalation if necessary, both because of the risk of ocular complications and the risk of amblyopia for the earliest forms.

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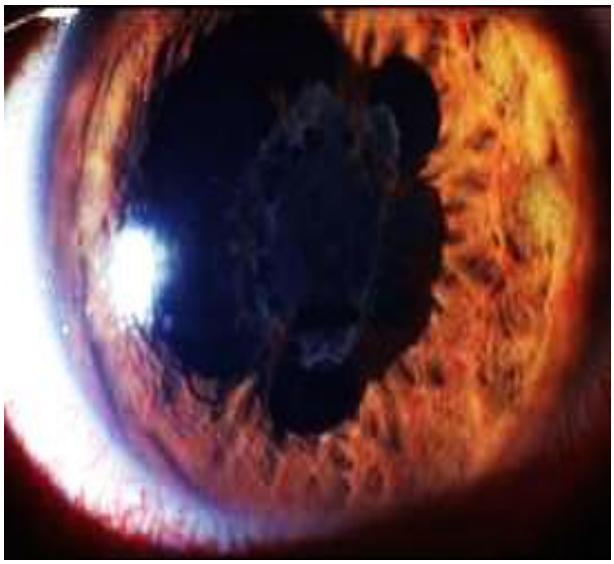

Figure 1: slit-lamp view showing central posterior synechiae, and posterior subcapsular cataract

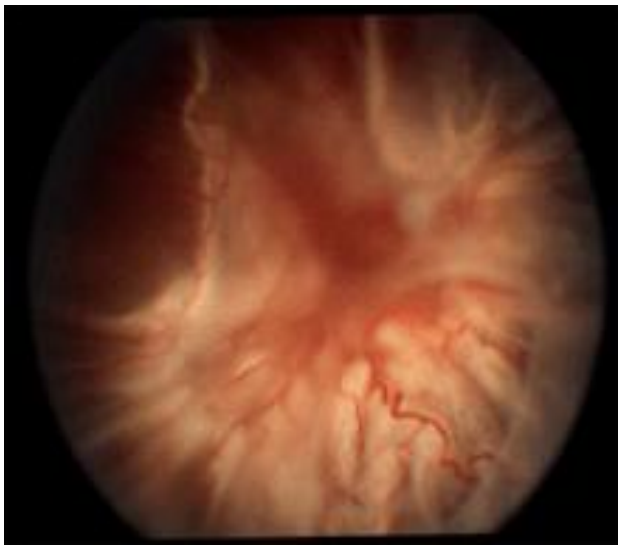

Figure 2: FAundus showing Total retinal detachment, with ischemic and frozen retina

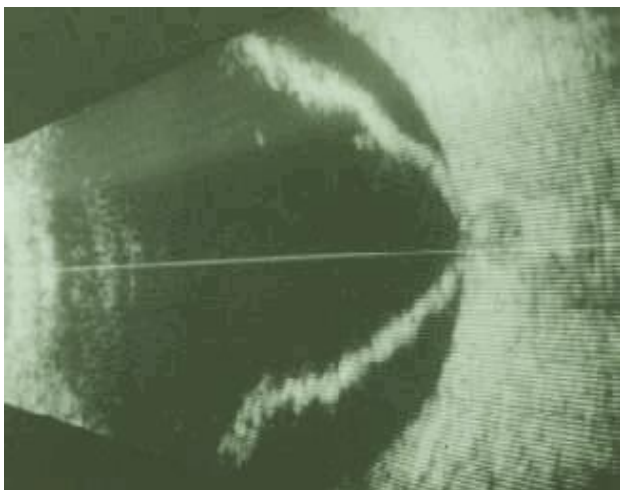

Figure 3:Ultrasound image of the retinal detachment, retinal layers being thick and immobile under pressure.

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