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A clinical case of Juvenile Idiopathic Arthritis (JIA) complicated by anterior uveitis

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Summary

A case of chronic Juvenile idiopathic arthritis (JIA) with onset at the age of 5 years was treated in a little girl who exhibited, in addition to typical joint inflammatory symptoms, inflammatory phenomena in the anterior uvea of both eyes. Of note was the disappearance of ocular inflammation after uveitic cataract removal surgery.

Key Words

Juvenile idiopathic arthritis, anterior uveitis, iridocyclitis, uveitic cataract.

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INTRODUCTION

Juvenile Idiopathic Arthritis (JIA) is a chronic disease characterized by persistent joint inflammation.

Typical signs of joint inflammation are: pain, swelling and limitation of joint motion. The term "idiopathic" means that the cause is unknown, while "juvenile" means that the onset of symptoms occurs before the age of sixteen. It is a rare disease that affects about one out of every 10,000 children. It is considered to be an autoimmune-based disease due to a combination of several factors, including genetic predisposition and exposure to unclear, probably infectious, environmental factors. It can be considered JIA when the onset occurs before the age of 16, when the arthritis has lasted for more than six weeks, and in cases where specific etiologic agents cannot be identified.

The diagnosis of this disease is therefore based on the presence and persistence of arthritis and the careful exclusion of any other possible cause of joint inflammation, evaluating the patient's medical history and relying on careful clinical examination and evaluation of laboratory tests (follow, for example, the bibliography cited: Kunimoto et al., 2006; D'Amelio, 2009; Mehta et al., 2014; Barut et al., 2017; Yangzes et al., 2019; Gaggiano et al., 2020; O'Rourke et al., 2021; Onel et al., 2021; Maleki et al., 2022). There are several forms of the disease.

They are generally distinguished as:

- Systemic JIA;
- Polyarticular JIA;
- Oligoarticular JIA.

Oligoarthritis, to which the present case refers, affects the large joints asymmetrically. It is the most common form of JIA (50 percent of cases). Sometimes it affects only one joint; this is the monoarticular form. It generally arises around 6 to 9 years of age and more often in girls.

Some patients may develop a major ocular complication, inflammation of the structures of the anterior chamber of the eye (anterior uveitis or iridocyclitis). If not identified in time and not treated, anterior uveitis progresses and seriously damages the eye and thus vision. Early diagnosis of this complication is therefore most important. Since it does not cause any obvious symptoms, it cannot be noticed by parents or non-ophthalmologists; in children with JIA, a comprehensive eye examination with slit-lamp should then be performed every three months.

CLINICAL CASE

The patient is a girl named G.D. with an acute onset of the pathology at the age of 5 years with pain in the right wrist that is also mildly edematous.

An X-ray was carried out, showing no traumatic lesions or any anatomical changes.

The pain and swelling do not regress, rather they become more pronounced, thus an orthopedic examination is performed at the "G. Di Cristina" Children's Hospital in Palermo. The orthopedist describes the presence of "*mildly swollen right wrist, with positive thermotact, absent extension and flexion allowed up to 45°; rheumatology examination should be performed*".

The rheumatologist's report: "By about a month, hard-elastic swelling has appeared in the right wrist, painful enough to lead to functional deficit". Hematologic tests are prescribed, which are and always will be normal, except for a marked persistent increase in C-reactive protein (23.3 mg/L at the time of the first test). ANAs have been consistently negative.

The Magnetic resonance imaging (MRI) of the right wrist is very significant, while the patient also begins to complain of pain in the right knee and right ankle: "*Uneven signal intensity at the carpal bones and radial epiphyseal region compatible with edema of the bony spongiosa. Concomitant evidence of synovitic reaction. The radiocarpal synovium is markedly thickened, with the presence of intra-articular fluid effusion*".

Therefore, a pauciarticular JIA is diagnosed, oral NSAID (Ibuprofen) therapy is prescribed, which had also been practiced previously without obvious benefit, and ophthalmic consultation is requested.

On direct visual examination, the conjunctiva did not appear hyperemic, and vision testing revealed no visual deficits: natural visus 10/10 in both eyes. Ocular pathology is often, in fact, insidious by not showing at first ocular redness nor blurred vision. However, during slit-lamp examination, the presence of iridocyclitis (inflammation of the iris and ciliary bodies - structures of the anterior uvea) characterized by the presence of a Tyndall 2+ effect was detected in both eyes.

The Tyndall effect consists of the presence of tiny particles floating within the anterior chamber of the eye, which are made evident by the beam of light from the slit-lamp passing through them. It is similar to the effect of dust particles that can be noticed inside a room when sunlight passes through the slits in the shutters of a window.

The intensity of the Tyndall effect is expressed with plus signs, ranging from just one (1+) and up to four pluses (4+).

It indicates the presence of particles, mostly of a protein nature, exuding from inflamed uveal structures (the iris especially).

If the inflammation is particularly intense, there may also be exudation of more voluminous corpuscles and even cells (leukocytes) that settle, by gravity, in the lower part of the anterior chamber producing what is called "hypopyon".

Other times this exudate, formed by cellular elements, may adhere to the posterior surface of the cornea producing "endothelial deposits".

In the present case, only a modest Tyndall effect was present, and the ophthalmologist then prescribed: mydriatic eye drops (atropine 0.5%), one drop morning and evening; and dexamethasone eye drops (2mg/ml), one drop three times daily. Follow-up after 1 week.

At follow-up, the eyeballs both appeared stable. Atropine is therefore discontinued and the dosage of corticosteroid-based eye drops reduced to one drop daily.

At subsequent follow-ups, however, iridocyclitis always recurred as soon as the dosage of the local steroid was reduced. Thus, continued therapy of the same eye drops with periodic monitoring of ocular tone, visus, and lens transparency (cataractogenic and hypertensive effects on ocular tone by corticosteroids) was deemed necessary.

From a general point of view, the arthritic component also showed no sign of disappearing despite the fact that weekly Methotrexate therapy was undertaken.

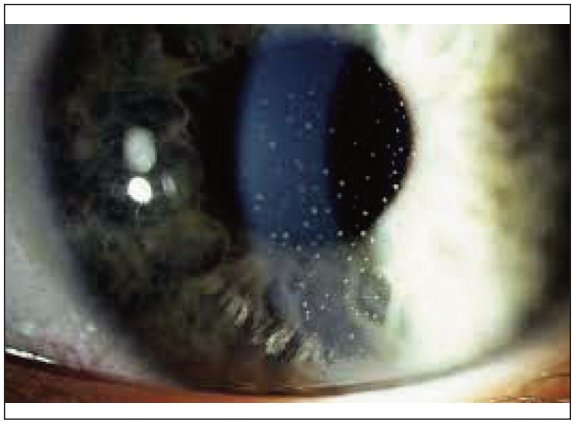
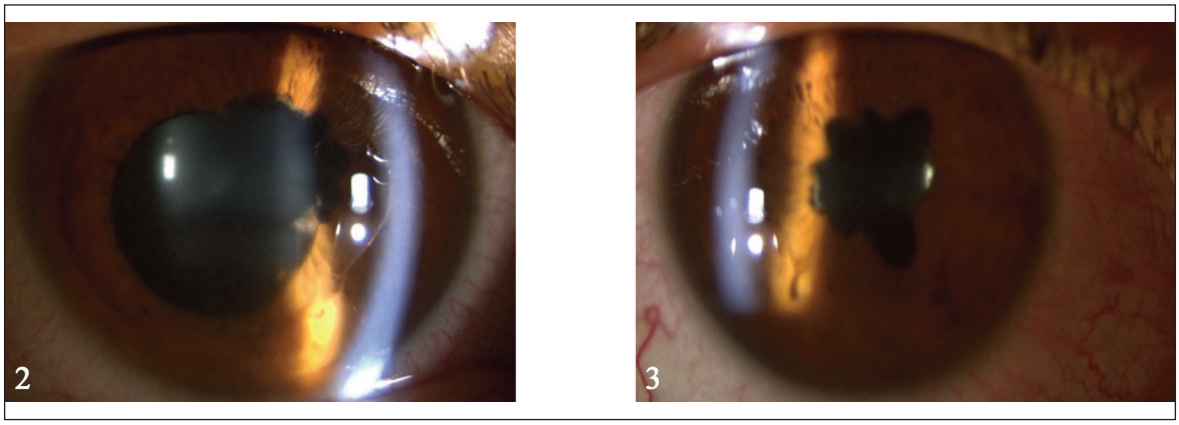


Figure 1. The "Tyndall effect" consists of the presence of tiny particles floating within the anterior chamber of the eye, which are made evident by the beam of light.



Figures 2, 3. Irido-lenticular synechiae in right eye (Fig. 2) and left eye (Fig. 3).

Therefore, a diagnosis of chronic JIA was suggested.

Subsequent ophthalmic examinations also noted the appearance of irido-lenticular synechiae. These are adhesions between the iris and anterior surface of the lens in both eyes due to the continuous recurrences of the inflammation and the discontinuation of the mydriatic eye drops at the behest of the child's parents. As she was by then 8 years old, she complained of the difficulty in applying them at close range - a side effect of these eye drops.

Further progression of lens opacity, due in part to corticosteroid-based eye drops and in part to the iridocyclitis (uveitic cataract) itself, dictated the need for cataract surgery at some point.

The surgery was conducted at the Bambin Gesù Hospital in Rome first in the right eye and after six months in the left eye. In both cases, the opacified lens was removed by phacoemulsification and a multifocal artificial lens was implanted.

At present time, after more than two years after the surgeries, the child, who is now 11 years old, possesses a natural visual acuity (without correction with glasses) of 7/10 in right eye and 8/10 in left eye.

After the cataract surgeries, the episodes of uveal inflammation greatly reduced until they disappeared completely. The use of antiphlogistic eye drops was no longer necessary.

In other similar cases described in literature, there is often a reduction in inflammatory phenomena after lens removal. This has led to speculation about the presence of immunological disease by lenticular proteins that we know are "stored" from birth and therefore not recognized by the immune system.

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