Juvenile idiopathic arthritis-associated uveitis complicated by glaucoma and Brown’s syndrome

Sr., JIA represents the most common form of chronic arthritis in paediatrics. A large population-based German cohort found a prevalence of uveitis in children with JIA of 12% [1]. Whereas secondary glaucoma affects 6–15% of patients with JIA-associated uveitis [1–3], Brown’s syndrome (restriction of elevation of the adducted eye) is seldom reported [4–9]. We describe a child with extended oligoarticular JIA who developed secondary glaucoma and Brown’s syndrome.

A 7-year-old girl was referred to our centre at 14 months of age with a 1-month history of swelling of the PIP joint of her right fourth finger. Her initial work-up (complete blood count, biochemistry, serology and acute phase reactants) was either normal or negative, with the exception of repeatedly positive ANAs. Over the next few months, swelling of her right fifth finger PIP, right elbow and knees was noted despite ongoing therapy with MTX. Seven months after the disease onset, chronic anterior uveitis was detected on a routine ophthalmologic exam. Topical therapy was started and 4 months later the ocular disease was inactive. She remained in remission with therapy until 1 year later, when she presented to the hospital with fever, polyarthritis and active anterior uveitis. Anti-TNF therapy (etanercept) was added to her treatment, achieving adequate control of her articular and ocular disease.

Due to minor uveitis exacerbations, she was controlled every 3 months at the ophthalmology clinic. Five years after the disease onset, a routine ophthalmologic exam revealed a uveitis flare and elevated intraocular pressure (IOP) of 34 mmHg bilaterally. At that time her articular disease was inactive on MTX and etanercept. Therapy with oral acetazolamide and topical dorzolamide was started with rapid improvement of the IOP. Adalimumab was reintroduced 1 month after surgery with progressive improvement of ocular mobility. At her last clinic visit, 9 months after development of Brown’s syndrome, her ocular mobility was almost normal.

Brown’s syndrome may be congenital or acquired [10]. The latter represents a mechanical obstruction to movement of the superior oblique tendon as it passes through the throclea. In rheumatic patients, it is thought to be secondary to tenosynovitis of the superior oblique muscle. It has been previously described in seven patients with JIA [4–9] (supplementary Table S1, available as supplementary data at Rheumatology Online), six with the systemic form of the disease (SoJIA) [5–9] and in patients with other rheumatic conditions such as RA or lupus [10]. In most cases of Brown’s syndrome associated with JIA, recent flare-ups or very active articular disease were present before the development of gaze disturbances. Our patient not only did not have SoJIA but also her articular disease remained inactive at the time of presentation. Brown’s syndrome, however, developed shortly after detecting a uveitis flare with glaucoma.
It is well known that patients with JIA may develop tenosynovitis at any location although the eye has rarely been involved. Brown’s syndrome represents another ocular complication of JIA and should be treated promptly. It may resolve spontaneously although previous reports have shown that the most efficacious therapy is administration of systemic steroids [4, 5, 7–9]. Different regimes have been used, from 10 mg every other day orally [5] to pulses of 300 mg i.v. [9], with good results. The use of steroids resulted in resolution of the symptoms in an interval that varied from 24 h [9] to 2 months [5]. Therefore, our case represents a typical case of Brown’s syndrome (Fig. 1) with atypical features including the absence of articular activity at the time of presentation, its temporal association with uveitis and steroid-induced glaucoma, and the JIA category, extended oligoarticular instead of systemic.

Rheumatology key message

- JIA patients may develop throcleitis/tenosynovitis of the superior oblique muscle.

Disclosure statement: E.E. has received grants from Merck and Schering-Plough for support of the outpatient clinic. All other authors have declared no conflicts of interest.

Supplementary data

Supplementary data are available at Rheumatology Online.

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Accepted 23 February 2012

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Rheumatology 2012;51:1730–1732
doi:10.1093/rheumatology/kes040
Advance Access publication 17 April 2012

Adult-onset Still’s disease in a patient with cystic fibrosis and its successful treatment with anakinra

Sir, We report the case of a 22-year-old woman with a known history of cystic fibrosis. She initially reported subfebrile temperatures with an increase in her fever up to 40°C in recent days, a sore throat and a macular rash on both forearms for 3–4 weeks. These episodes tended to occur in the evening.

In the outpatient clinic, a respiratory tract infection was assumed and antibiotic therapy with clarithromycin and inhaled tobramycin was started. Notably, she had a known colonization with mucoid and non-mucoid Pseudomonas. The patient’s medication consisted of pancreatic enzyme, inhaled tobramycin and colistin, tiotropium bromide and fluticasone/salmeterol. She is intolerant to itraconazole (abdominal pain, nausea). Otherwise the patient’s medical and travel history was unremarkable. The patient works as a pharmaceutical technical assistant.

A general physical examination was unremarkable except for tachycardia (heart rate 108/min) at regular rhythm, white exudates on both tonsils, slight redness of the oropharynx and lymph node swelling in the submandibular and the supraclavicular area bilaterally. Lung auscultation revealed rales and rhonchi over both lungs. The