

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 threcleitis/tenosynovitis of the superior oblique muscle.

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Supplementary data

Supplementary data are available at *Rheumatology* Online.

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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 sub-febrile 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

Fig. 1 Snapshot image of the exanthema on the right forearm.



implanted port catheter system in the *fossa infraclavicularis* showed no signs of infection. Her fingers showed nail clubbing; swollen or tender joints were not found.

Due to persistent fever, the patient was admitted to the hospital. The patient quickly suffered from arthralgias (elbows, knees and wrists) and the transient rash recurred (Fig. 1). Initial laboratory parameters showed elevated levels of CRP (13.6 mg/dl, normal range <0.5 mg/dl), but procalcitonin (0.3 ng/ml, normal range <0.5) and white blood cell count were within their normal ranges, the differential showing a mild left shift (leucocytes 7.9 g/l, 77% neutrophils, 17% lymphocytes, 5% monocytes, 2% eosinophils). Ferritin levels were elevated to a maximum of 1834 µg/l (normal range 30–300 µg/l) [1]; however, glycosylated ferritin was not assessed. Antibiotic medication was rapidly switched to meropenem and i.v. penicillin V for a possible streptococcal tonsillitis; however, microbiological testing of sputum, blood, urine, stool and pharyngeal swab revealed no causative pathogen. Ultrasonography of the abdomen showed mild splenomegaly. A CT scan revealed no indication of exacerbation of cystic fibrosis. An echocardiogram ruled out endocarditis. There was no laboratory evidence for allergic bronchopulmonary aspergillosis.

As the antibiotic regimen showed no effect it was stopped. Due to persistently elevated markers of inflammation (CRP >10 mg/dl, procalcitonin rising from normal range to 1.0 ng/ml), the port catheter system was explanted and a PET/CT scan was carried out, showing mediastinal, supraclavicular and para-aortal lymphadenopathy. A supraclavicular lymph node was extirpated, microscopic examination revealing only unspecific signs of reactive inflammation.

Shortly afterwards the patient showed rapid worsening of respiratory parameters with progressive viscous mucus, dyspnoea and tachypnoea. Respiratory insufficiency developed, and she had to be intubated and ventilated mechanically. A CT scan now revealed ubiquitous ground-glass pattern infiltrates in both lungs. Bronchoscopy and bronchoalveolar lavage were performed and revealed the known chronic colonization/inflammation with *Pseudomonas* spp. as well as *Aspergillus fumigatus*. The antimicrobial regimen was expanded; the patient was treated with i.v. tobramycin, fosfomycin, meropenem, caspofungin and inhalative colistin and tobramycin.

At this point of critical illness, adult-onset Still's disease (AOSD) was considered, since the patient fulfilled three major and three minor criteria of the Yamaguchi *et al.* [2] classification criteria as well as four major Fautrel *et al.* [3] criteria (fever, arthralgia, pharyngitis and exanthema). Consequently the patient was given high doses of glucocorticoids as well as the IL-1 receptor antagonist anakinra, which has been shown to be able to quickly resolve symptoms in severe cases of AOSD [4, 5]. These measures led to a rapid decline of fever, the inflammation signs vanished and the patient was extubated successively. Thus a pulmonary manifestation of AOSD has to be assumed, and this manifestation has been described in several case reports so far [6, 7]. Furthermore, the rapid improvement upon treatment with steroids and anakinra retrospectively support the diagnosis of AOSD. However, diagnostic uncertainties remain. Most importantly, the patient had a positive testing for *A. fumigatus* and *Candida albicans* in the bronchoalveolar lavage. However, ELISA antigen testing for *Aspergillus* remained negative. As the chronic lung changes secondary to cystic fibrosis make the patient vulnerable to multiple infections, a microbiological cause must still be considered in the differential diagnosis of the acute pulmonary deterioration. Also, several immunological triggers, e.g. bacterial infections, viral infections as well as vaccinations, have been described as putative triggering factors for AOSD [8].

Shortly after, the patient was transferred to our normal care clinic and glucocorticoids were tapered. At a dose of 5 mg prednisolone per day the fever and rash did not recur. Antibiotic therapy was successively discontinued. Finally, medication with anakinra was stopped 1 month later and the patient was discharged from clinic. Four months later the patient was feeling fine, and both the rash and the arthralgias never recurred.

Rheumatology key message

- Here we report successful treatment of AOSD with anakinra in a patient with cystic fibrosis.

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