age greater than 9 years, presence of frontal sinusitis, large or nonmedial SPA, suspicion of anaerobic infection, recurrence of the SPA after previous drainage, evidence of chronic sinusitis, acute optic nerve or retinal compromise, or infection of dental origin. Our 15-year-old patient presented with an orbital cellulitis most likely secondary to ipsilateral ethmoid and maxillary sinusitis. The infection was initially unresponsive to medical treatment, and orbital imaging revealed an atypically large, gas-containing inferomedial SPA—features strongly suggestive of an atypical bacterial infection—that justified surgical intervention. This case highlights the importance of multidisciplinary care to achieve an optimal clinical outcome.

Patrick Daigle, MD; patrick.daigle@usherbrooke.ca.

Infliximab for management of severe refractory posterior scleritis in a 9-year-old boy

Posterior scleritis (PS) is a chronic inflammatory condition of the sclera posterior to the rectus muscle insertions. PS predominantly affects adult women and is rare in children. Adult form of PS is typically associated with autoimmune systemic diseases; however, pediatric PS is mostly idiopathic with no known underlying conditions. The initial presentation of PS in children is nonspecific with painful red eye, potential vision loss, light sensitivity, and eye movement restriction, which make the diagnosis difficult.

The current recommended treatment for mild PS is oral nonsteroidal anti-inflammatory drugs (NSAIDs); however, steroids and steroid-sparing immunosuppressive treatment are required for more severe cases. In adult PS refractory to standard immunomodulatory agent, antitumour necrosis factor agents such as infliximab have shown good disease control. There are few case reports of pediatric PS, most of which have resolved using 2 or more immunosuppressive agents.

Here, we present a 9-year follow-up study of a patient with pediatric PS with a history of multiple flare-ups while on oral steroids, methotrexate, and mycophenolate, who achieved resolution using infliximab and maintained remission after cessation of treatment. Our case represents only the second case report of infliximab use in paediatric PS.

We present a patient with childhood-onset PS. Written consent was obtained from the patient guardian. The data on patient’s electronic chart, including history, systemic and ocular findings, laboratory results, investigations, and treatment, were reviewed.

A 9-year-old boy with a history of asthma, sickle cell trait, and no significant birth history presented with a 4-day history of fever, left eye pain and swelling, foreign body sensation, and tearing. Eye examination showed unilateral decreased visual acuity (right and left eyes 0.1 and 0.7 logMAR, respectively) with equal, round, and reactive pupils to light. There was pain with restriction of ocular motility in the left eye and 3 mm of proptosis, periorbital swelling and injected conjunctiva. Dilated fundus examination showed left optic nerve edema (Fig. 1A). The laboratory results showed leukocytosis with increased neutrophils. After a diagnosis of presumptive orbital cellulitis, he was started on intravenous (IV) cefotaxime, cloxacillin, and clindamycin. Computerized tomography (CT) scan showed diffuse scleral thickening of the left eye associated with swelling of the lateral pre- and postseptal soft tissues extending into retrobulbar fat and surrounding the distal optic nerve. Three days of treatment did not achieve clinical improvement, and a new finding of anterior uveitis was noted. Repeat CT scan showed worsening of the orbital signs more indicative of inflammatory disease rather than orbital cellulitis. An ultrasound B-scan confirmed the characteristic T sign of scleritis (Fig. 1B), and a diagnosis of severe refractory posterior scleritis was made.

References


Footnotes and Disclosure

The authors would like to acknowledge the patient presented in this study, who generously granted them permission to share his story and photographs with the academic community.

The authors have no proprietary or commercial interest in any materials discussed in this article.
unilateral severe PS was made. He received 2 weeks of IV methylprednisolone 900 mg/day, which controlled the inflammation and improved his vision to 0.0 logMAR.

All infective work-up including Tuberculosis, syphilis and the Herpes Simplex, Varicella-zoster, Cytomegalovirus, Epstein-Barr, Human Herpes, Human immunodeficiency, Hepatitis B and Hepatitis C viruses, as well as autoantibodies including antinuclear antibody (ANA), anti-dsDNA, antineutrophil cytoplasmic antibody, antmyeloperoxidase antibody, anti-proteinase 3 antibody, antcardiolipin antibody, and rheumatoid factor were negative. We excluded Immunoglobulin (Ig) G4 disease and the total serum levels of IgG, IgM, and IgA were also within the normal range. He was discharged on oral prednisolone 40 mg per day on a slow taper and methotrexate subcutaneous injection 25 mg per week. He re-presented with severe left scleritis 6 months later while on oral prednisolone 10 mg and methotrexate 25 mg, requiring further IV methylprednisolone treatment for 2 weeks. Mycophenolate 540 mg was also added to prednisolone 40 mg and methotrexate in order to control the scleritis. The patient exhibited 3 additional flare-ups of bilateral scleritis over the following 3 years (Fig. 2) as a consequence of tapering the oral prednisolone to 10 mg. These flare-ups were controlled by hospitalization and receiving pulse of IV methylprednisolone. The family’s initial resistance to treatment with antitumour necrosis factor medication required further discussions at this stage. They agreed to infliximab 300 mg infusion every 4 weeks, and methotrexate and mycophenolate were stopped. The patient successfully weaned off oral prednisolone. He achieved sustained remission on infliximab alone for 4 years, except for one flare-up owing to treatment noncompliance, which resolved quickly on resumption of infliximab. He tapered off infliximab over a further period of 1 year. The patient had a mild flare-up of scleritis 6 months after stopping infliximab, which responded to ibuprofen 400 mg 3 times a day for a month with no further episodes. He stays in remission with 0.1 logMAR uncorrected vision and a normal eye examination 9 years after initial presentation.

PS is rare in children, with a nonspecific, heterogeneous clinical presentation. Ocular pain, redness, lid involvement, and restriction of extraocular muscle movement are the most common early signs of pediatric PS, which are also features of other common diseases affecting children. Our case was initially managed as orbital cellulitis owing to significant pain and restricted ocular motility, proptosis, fever, and raised white cell counts. Orbital cellulitis is much more common than PS and represents a life-threatening differential diagnosis, which must be excluded when acute orbital inflammation is noted. On the other end of severity scale, Mallick et al.\textsuperscript{9} reported a case of a 14-year-old boy with eye redness, decreased vision, and conjunctival congestion who initially was managed as viral conjunctivitis. After careful clinical examinations and imaging, he was diagnosed with PS and was subsequently treated with oral NSAIDs and topical steroid for 2 weeks, resolving the eye inflammation and improving his visual acuity to 20/20.\textsuperscript{9}
Oral NSAIDs are the first line of treatment for PS; however, immunosuppressive drugs are often needed to fully control the inflammation. One study reported on 20 eyes of 13 pediatric patients with PS and reported flare-ups when tapering the oral corticosteroid to <0.5 mg/kg/day. Owing to exhibiting side effects or relapse on low-dose corticosteroid, at least one immunosuppressive drug was added to control the inflammation. In the current case report, the patient experienced 3 separate episodes of flare-up whilst on full dose of methotrexate and mycophenolate when oral prednisolone was reduced to 10 mg/day (0.2 mg/kg/day). Therefore, infliximab 300 mg every 4 weeks was successfully started to control his inflammation. Only one previous case report on paediatric PS used infliximab for disease resistant to methotrexate, methylprednisolone, and cyclosporine treatment. Weiss et al. presented the case of a 13-year-old girl diagnosed with PS who maintained remission with 10 infusions of infliximab (5 mg/kg) with no side effects over a 16-month follow-up. Our case likely represents a more severe case requiring much longer immunomodulation compared to the initial case reported. Although there are published data on infliximab therapy for childhood refractory uveitis, it is encouraging to report safe and effective disease control in PS as well.

In conclusion, the majority of pediatric PS cases in literature report poor disease control with only NSAIDs or corticosteroids, with many requiring at least one immunosuppressive agent to avoid steroid side effects. We propose that infliximab can be considered an alternative treatment for severe paediatric PS resistant to treatment.

Sina Khalili,* Kamiar Mireskandari, MBChB, FRCSEd, FRCOphth, PhD†
*The Hospital for Sick Children, Toronto, Ont.; †University of Toronto, Toronto, Ont.


Correspondence to: Kamiar Mireskandari: kamiar.mireskandari@sickkids.ca.

Iris stromal defect in an infant masquerading as a tumour: the sailing iris

Congenital iris lesions are rare and generally identified early in life. A pigmented lesion on the iris ranges from a simple nevus or cyst to tumors including melanoma. These lesions can arise from both iris stroma and iris epithelium. Thorough examination as well as anterior segment imaging studies may be required to establish a diagnosis. Here, we report a rare case of congenital iris stromal defect masquerading as an iris tumour.

References


Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Methods

The history, systemic and ocular findings, and investigations of a patient with a congenital iris lesion was reviewed. Written consent was obtained from the patient’s guardian.

Case report

A 3-month-old boy with no significant birth or family history, presented with an abnormality of the left iris. On slit lamp examination, a pigmented iris lesion in the