Intermediate uveitis as an initial presentation of HLA B27 associated spondyloarthropathy in an adolescent boy: A rare case report and review of literature

Prateek Kumar Panda[1], Srikant Kumar Padhy[2]

From Departments of [1]Paediatrics, [2]Ophthalamology, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Prateek Kumar Panda, Room No – 224, RPC-1 Hostel, All India Institute of Medical Sciences, New Delhi – 110 029, India. E-mail: drprateekpanda@gmail.com

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ABSTRACT

Juvenile ankylosing spondylitis is less prevalent in children, and usually, children are presented to the hospital with chronic inflammatory back pain, enthesitis, and often hip and shoulder joint involvement. A significant proportion of these children develop ocular complications such as anterior and intermediate uveitis. Most of these children have HLA B27 positivity. On the other hand, in cases with anterior and intermediate uveitis, a considerable proportion has positivity for HLA B27 antigen. Few of these patients, later on, develop other clinical features suggestive of ankylosing spondylitis. However, there are only a few anecdotal case reports of similar HLA B27 associated intermediate uveitis, who later on developed juvenile ankylosing spondylitis. The current case is a 12-year old boy, who initially had bilateral intermediate uveitis with HLA B27 positivity without any other systemic or musculoskeletal features. However, on follow-up he developed enthesis and sacroiliitis suggestive of ankylosing spondylitis. Both rheumatological and visual complains had an excellent clinical response to corticosteroids in this child. The current case report demonstrates the importance of evaluation for HLA B27 positivity in all children with isolated intermediate uveitis, even if systemic and musculoskeletal examinations are normal.

Key words: *Ankylosing spondylitis, HLA B27, Intermediate uveitis, Spondyloarthropathy*

Rheumatological disorders of autoimmune origin have relatively less prevalence in children as compared to young adults and elderly age group. Common rheumatological disorders in children are juvenile idiopathic arthritis; arthritis associated with other autoimmune and connective tissue diseases such as systemic lupus erythematosus and juvenile dermatomyositis and rheumatic fever [1]. Juvenile ankylosing spondylitis is rarely diagnosed only in adolescent males, and ankylosing spondylitis as such is considered a disease of young adults. Ankylosing spondylitis is one of the seronegative spondyloarthropathies in which, the rheumatological factor is usually negative. On the other hand, about 90% of persons with ankylosing spondylitis have HLA B27 antigen positivity [2].

Ankylosing spondylitis usually presents as chronic inflammatory low back pain in adolescents and adults <40 years of age, with insidious onset and gradual progression [3]. Many of the affected persons complain of increased severity of pain during rest or just after waking in the morning and at least partial resolution in the severity of pain with physical activity. Juvenile ankylosing spondylitis, in which the age of onset is usually <18 years, differs from the classical presentation of ankylosing spondylitis. It involves appendicular joints, such as knee and ankle joints, more commonly and is frequently associated with enthesitis and tarsitis. Atypical presentation in children often causes a significant delay in establishing the correct diagnosis and instituting proper management [4].

About 40% of patients with ankylosing spondylitis have ocular complications, and among them, anterior uveitis is the commonest one, followed by intermediate uveitis or pars planitis [5]. However, these ocular complications usually occur after the onset of musculoskeletal complaints. There are only a few case reports in pediatric literature of intermediate uveitis in juvenile ankylosing spondylitis. Hereby, we are presenting one rare case report of a 12-year-old boy who was admitted to the hospital initially with intermediate uveitis and found to have HLA B27 positivity. The boy also developed enthesitis, knee joint pain, and lower back pain, later. The novelty in this case report remains in the fact that intermediate uveitis presenting as an isolated initial clinical manifestation of juvenile ankylosing spondylitis is extremely rare. Thus, it explains the active collaboration and cross-referral required between ophthalmologists and pediatricians for the holistic management of the child.

CASE SUMMARY

The index case is a 12-year-old boy, who was presented with acute onset, rapidly progressive, painless vision impairment initially in the right eye and subsequently in the left eye, developing over 1 week. At the beginning of the illness, the child also had floaters in the visual field bilaterally, but there was no redness, foreign body sensation, diplopia, or any other ocular complaints. At initial presentation, the child had visual acuity of only finger counting at a distance of 1 m

in bilateral eyes. The child did not have any fever, rash, joint pain, or swelling, cough, respiratory distress, oral ulcer, or clinical features suggestive of enthesitis or other organ system involvement.

There was no family history of any rheumatological diseases or history of similar vision loss in young age in any of the first or second-degree relatives. On detailed ophthalmological evaluation, the child was found to have the presence of snowball like opacities floating in vitreous suggesting inflammatory exudates. These inflammatory exudates were predominantly found near pars plana region (snow banking), thereby establishing the diagnosis of intermediate uveitis. Apart from these, there was no other retina, cornea, or choroidal involvement. Only aqueous flare, suggestive of leukocytes in the aqueous chamber, was present in the right eye.

The child was also evaluated for other systemic associations such as juvenile idiopathic arthritis, ankylosing spondylitis, sarcoidosis, and multiple sclerosis. Initially, X-ray of all major axial and appendicular joints, chest X-ray and magnetic resonance imaging brain were within normal limits. Rheumatoid factor, antinuclear antibody, and serum angiotensin converting enzyme level were within normal limits. However, the child was found to have positivity for HLA B27 antigen. Erythrocyte sedimentation rate and serum C-reactive protein values were raised suggestively of systemic inflammation.

Thus, the cause of vision impairment was attributed to HLA B27 associated with intermediate uveitis. The child was started on periocular corticosteroid injection, followed by the institution of oral steroid (prednisolone) at 2 mg/kg/day for 2 weeks followed by tapering over the next 4 weeks. With this, the child showed significant vision improvement in the bilateral eye; after 2 weeks of starting steroid, visual acuity in bilateral eyes was 6/36. However, after stopping the oral steroids, the child developed right ankle pain and clinical signs suggestive of enthesitis.

Over the next 2 weeks, the child also developed low backache and Schober test was found to be positive at that time. Repeat X-ray of sacroiliac joint shows some erosion of sacroiliac joint margins and increased joint space which was suggestive of sacroiliitis. Hence, the child was started on oral naproxen and a short course of repeat oral prednisolone for 4 weeks, to which the child showed significant improvement. After tapering steroid, the child was started on oral methotrexate at 10 mg/m^2/week and at 1 year after follow-up; the child had no further recurrence of symptoms. At 6 months after follow-up, repeat visual acuity was 6/9 in the right eye and 6/6 in the left eye suggesting a significant improvement.

DISCUSSION

Usually, HLA B27 associated uveitis presents with sudden onset acute anterior uveitis in adolescents and young adults. About 80% of the cases are asymmetrically bilateral. The inflammation is usually more severe than that found in idiopathic anterior uveitis and often associated with a fibrinous reaction, a hypopyon or formation of posterior synechiae [6]. It is associated with higher recurrence rates than the idiopathic variety. Posterior segment involvement in HLA-B27 associated uveitis is an under-recognized phenomenon but has been shown to occur in up to 17% of patients with B27 associated uveitis. This may take the form of posterior vitritis, retinal vasculitis, papillitis, or cystoids macular edema [7].

Uveitis associated with the HLA-B27 gene may occur in the presence or absence of an associated systemic condition [8]. About 20% of patients present with the ocular symptoms as the first manifestations of a systemic illness such as ankylosing spondylitis, sarcoidosis, or multiple sclerosis which became evident later. Due to this, it is crucial to take a detailed history and to conduct a focused systemic examination of these patients [9].

The prognosis in patients with HLA-B27 associated uveitis is controversial. Many authors have reported no significant differences or, indeed, a better prognosis in patients with an HLAB27 haplotype than in patients with idiopathic uveitis [10]. Ankylosing spondylitis in the juvenile age group as such shows good response to NSAIDs and corticosteroids and morbidity and functional impairment can be significantly reduced with the institution of disease modifying anti-rheumatic drugs like methotrexate, as in our case [11].

CONCLUSION

The current case report demonstrates the importance of evaluation for HLA B27 positivity in all children with isolated intermediate uveitis, even if systemic and musculoskeletal examinations are normal. These children should be promptly treated with anti-inflammatory agents and corticosteroids. Regular follow-up is also essential in these children, as a significant proportion of these children may develop rheumatological complaints at a later date.

REFERENCES

1. Dahman HA. Challenges in the diagnosis and management of pediatric rheumatology in the developing world: Lessons from a newly established clinic in Yemen. Sudan J Paediatr 2017;17:21-9.
2. Mou Y, Zhang P, Li Q, Lin Z, Liao Z, Wei Q, *et al.* Clinical features in juvenile-onset ankylosing spondylitis patients carrying different B27 subtypes. Biomed Res Int 2015;2015:594878.
3. Baek HJ, Shin KC, Lee YJ, Kang SW, Lee EB, Yoo CD, *et al.* Juvenile onset ankylosing spondylitis (JAS) has less severe spinal disease course than adult onset ankylosing spondylitis (AAS): Clinical comparison between JAS and AAS in Korea. J Rheumatol 2002;29:1780-5.
4. Dumbrăveanu L, Cuşnir V, Groppa L, Calinina L. HLA B 27 uveitis in ankylosing spondylitis and reactive arthritis. Oftalmologia 2010;54:29-35.
5. Belachew DA, Sandu N, Schaller B, Guta Z. Ankylosing spondylitis in Sub-Saharan Africa. Postgrad Med J 2009;85:353-7.
6. Waheed NK. HLA-B27 Associated Uveitis. Available from: http://www.uveitis.org/wp-content/uploads/2017/05/hla_b27_related_uveitis.pdf. [Last cited on 2018 Mar 21].
7. Pars Planitis. NORD; 2008. Available from: http://www.rarediseases.org/rare-diseases/pars-planitis. [Last accessed on 2019 Jan 21].
8. Janigian RH Jr. Intermediate Uveitis. Medscape Reference; 2013. Available from: http://www.emedicine.medscape.com/article/1208794-overview. [Last accessed on 2019 Feb 04].
9. Pars Planitis. OMIM; 2001. Available from: http://www.omim.org/entry/606177. [Last accessed on 2019 Feb 18].
10. Uveitis. Medline Plus; 2014. Available from: http://www.nlm.nih.gov/medlineplus/ency/article/001005.htm. [Last accessed on 2019 Jan 28].
11. Prieto JF, Dios E, Gutierrez JM, Mayo A, Calonge M, Herreras JM, *et al.* Pars planitis: Epidemiology, treatment, and association with multiple sclerosis. Ocul Immunol Inflamm 2001;9:93-102.

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