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Vogt-Koyanagi-Harada disease in a 9-year-old girl

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ABSTRACT

Uveitis secondary to Vogt-Koyanagi-Harada disease is rare in children. To the best of our knowledge, this patient is the first reported case of uveitis attributed to VKH in a child in Malaysia. A 9-year-old girl complained of non-resolving painful red eyes bilaterally for 3 months. Anterior segment of right eye showed mutton fat keratic precipitates, posterior synechiae and moderate anterior chamber reaction. Anterior segment of left eye revealed mutton fat keratic precipitates, band keratopathy on peripheral 3 and 9 o' clock of the cornea, iris bombe, iris nodule and seclusio pupillae. Fundus examination of right eye revealed subretinal deposit at superior and inferior arcades with flat retina while left eye showed hazy view. Patient was then noted to have poliosis and vitiligo after 1 month from initial presentation. Mild cataract and widespread atrophy of the retinal pigment epithelium accounting for the loss in vision remained. This case report is to highlight the importance of early recognition of paediatric Vogt-Koyanagi-Harada and treating it aggressively to prevent the irreversible destructive sequelae of the disease.

1. Introduction

Uveitis secondary to Vogt-Koyanagi-Harada (VKH) disease is rare in children. To the best of our knowledge, this patient is the first reported case of uveitis attributed to VKH in a child in Malaysia. This case report describes the characteristics of VKH uveitis in this patient and takes a review at the available literature. Children are rarely affected with one report estimating that paediatric cases constitute approximately 3% of all cases[1].

2. Case report

A 9 years old girl complained of non-resolving painful red eye bilaterally for 3 months. It was associated with progressive blurring of vision and photophobia. It was not preceded with headache, hearing defect and skin or hair changes. There was no history of surgery or trauma.

Ocular examination revealed visual acuity of 6/24 in the right eye and hand movement in the left eye. Anterior segment of right eye showed mutton fat keratic precipitates, posterior synechiae and moderate anterior chamber reaction. Anterior segment of left eye revealed mutton fat keratic precipitates, band keratopathy on peripheral 3 and 9 o' clock of the cornea, iris bombe, iris nodule and seclusio pupillae. Intraocular pressure (IOP) of left eye was 34 mmHg and gonioscopy

finding noted peripheral anterior synechiae. Fundus examination of right eye revealed subretinal deposit at superior and inferior arcades with flat retina while left eye showed hazy view. B scan of left eye demonstrated a flat retina. Systemic examination was unremarkable at the first presentation.

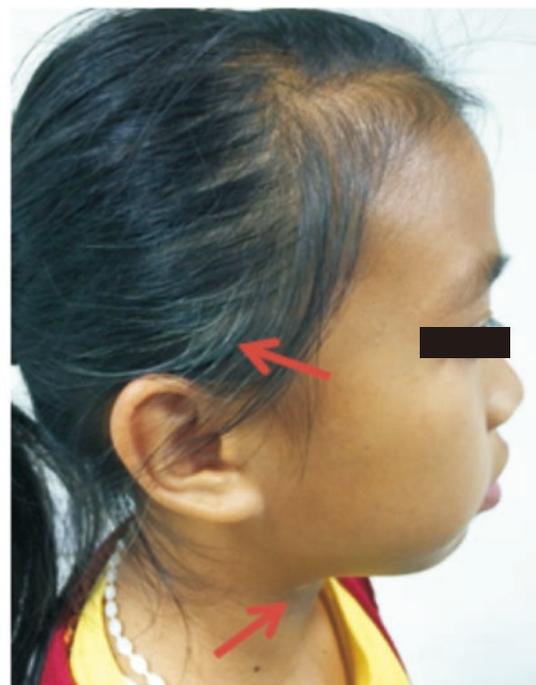


Figure 1. Poliosis and vitiligo.

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Laboratory examinations were done to rule out other possible immunological and infective causes like tuberculosis, syphilis, sarcoidosis and connective tissue disease. Full blood count, serum calcium, erythrocyte sedimentation rate, rapid plasma reagent test, rheumatoid factor, antinuclear antibody, C3 and C4 were all unremarkable, chest X-ray was unremarkable and Mantoux test was negative. Hearing assessment revealed no sensorineural loss.

This patient was initially treated as bilateral anterior uveitis with left eye pupillary block. In the left eye a peripheral iridotomy was done and IOP was controlled. Topical steroid eye drops, gutt maxitrol (Alcon, USA) once every 2 h and gutt atropine 1% tds were given. The uveitis was resolved around 1 month after starting treatment, left eye vision was improved to 6/60 and right eye vision remained status quo. Patient was then noted to have poliosis and vitiligo (Figure 1). Besides, right eye fundus demonstrated multiple Dalen-Fuch spots (Figure 2). In view of this, patient was then treated as VKH syndrome and tryptamine prednisolone 25 mg once a day was given and the dosage was tapered off within 2 months. Mild cataract and widespread atrophy of the retinal pigment epithelium accounting for the loss in vision remained.



Figure 2. Right eye fundus photo showing multiple Dalen-Fuch spots at inferior arcade.

3. Discussion

VKH syndrome is a rare multisystemic autoimmune disease characterized by ocular, cutaneous and neurological abnormalities. The exact cause of VKH remains unknown, but evidence suggests that it involves a T lymphocyte mediated autoimmune process directed against one or more antigens found on or associated with melanocytes.

Early stage of VKH might be overlooked and unrecognized. As in our patient, she presented to us as bilateral acute angle closure secondary to seclusio pupillae, associated with bilateral uveitis, moderately dense cataract with limited information from posterior segment. The incidence of uveitis in children is uncommon which is 5 fold less than adults[2]. Overall, juvenile idiopathic arthritis is the most common cause of anterior uveitis in children[2].

High IOP in VKH is multifactorial, resulting from angle closure secondary to seclusio pupillae or peripheral anterior synechia formation, immune mediated clogging of the trabecular meshwork and steroid induced glaucoma. Glaucoma was a common complication and reported in 46% of the children with VKH in Tabbara's report[3]. In our patient, IOP was well controlled after laser iridotomy and antiglaucoma medication. If these treatments fail, synechiolysis is recommended during cataract surgery. There is possible to improve aqueous outflow and trabecular function may be partially reversible[3].

Cutaneous changes have been reported to be the most variable and less predictive feature of the disease. This finding demonstrated in different case reports from 10% to two third of the patients[4].

Only vitiligo that occurs after the occurrence of the ocular disease is considered to be supportive of VKH. The time interval is usually between 4 weeks and 8 weeks[5]. In our case the time period was 3 months.

In acute phase, the common findings are uveitic phase with bilateral granulomatous anterior uveitis, iridocyclitis, choroiditis, exudative retinal detachment, papillitis and vitritis. Exudative retinal detachment was absent in our patient, only a few hypopigmented lesion suggestive of Dalen-Fuch spots in periphery fundus.

VKH syndrome is potentially vision threatening, typically as a result of cataract, glaucoma, RPE atrophy or rarely choroidal neovascularization. Subretinal neovascular membrane formation occurs in up to 10% of patients with VKH.

The mainstay of VKH management is suppression of intraocular inflammation with steroids and immunosuppressive agents. In early and aggressive treatment, young age, good vision within one month of treatment are associated with better visual prognosis[6]. They are various case reports showed different management, initiating either intravenous methylprednisolone for 3 days followed by tapering dose of oral corticosteroid or oral steroid solely. In our case, patient was treated with immunosuppressive dose of oral steroid. Read *et al.* showed no advantage of intravenous therapy over oral one[7]. Long term and high dose steroids are potentially hazardous in children. It causes premature epiphyseal fusion and thus growth retardations. Such considerations have led many authorities to recommend the early use of corticosteroid sparing agents such as methothrexate and cyclosporine. Ahmed *et al.* prospective study has shown mycophenolate mofetil significantly improved visual outcome as first lined therapy[8].

VKH is a rare cause of uveitis in children and thus the diagnosis of paediatric VKH associated uveitis is often delayed, and consequently the complications are usually present at presentation. This case report is to highlight the importance of early recognition paediatric VKH and treat it aggressively to prevent the irreversible destructive sequelae of the disease.

Conflict of interest statement

We declare that we have no conflict of interest.

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