Intraocular Histiocytosis in a 12-Year-Old Girl Without Systemic Disease

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ABSTRACT

Isolated intraocular histiocytosis is a rare disease that may manifest by recurrent uveitis and solid subretinal masses. The course, diagnosis and treatment of isolated intraocular histiocytosis in a 12-year-old girl are presented. As extensive diagnosis and therapy with corticosteroids and tuberculostatics failed to produce satisfactory results, diagnostic-therapeutic vitrectomy was performed. The intraoperatively obtained material was examined by the methods of histopathology, cytology and immunocytochemistry, along with herpes and cytomegalovirus polymerase chain reaction. The vitreous inflammatory exudate and subretinal masses were operatively removed. Analysis of the intraoperatively obtained material pointed to histiocytosis, whereas additional examinations revealed no systemic manifestations of the disease. Chronic uveitides that respond poorly to classic immunosuppressive therapy require multidisciplinary analysis of intraocular material. Pars plana vitrectomy is an appropriate diagnostic-therapeutic operative procedure.

Key words: histiocytosis, intraocular, case report

Introduction

Histiocytosis in the form of either Langerhans cell histiocytosis or non-Langerhans cell histiocytosis, is characterized by histiocyte proliferation and may involve various organs and tissues1. Among other organs, eyeball with its adnexa may also be affected, as is the case in juvenile xanthogranuloma, Rosni-Dorfman disease or Letterer-Siwe disease2–4. Ocular manifestations of histiocytosis may vary and generally are part of the complete clinical picture characterized by skin disorders, lymphadenopathy, bone lesions, hepatosplenomegaly, lung disease, central nervous system lesions, etc. Recurrent uveitis of any localization is the most common ocular manifestation of histiocytosis6–7. Exudative retinal detachment, subconjunctival masses, or optic nerve lesions are also frequently, and corneal lesions less frequently observed2–10. Isolated ocular histiocytosis without other systemic manifestations is extremely rare, and when it does occur, it is characterized by recurrent uveitis with a tumors intraocular growth11. Most uveitis patients typically respond poorly to local and systemic corticosteroid therapy, and even when therapy leads to regression of the disease, this effect is transient with regular disease relapses2,9,10. Therefore, vitreoretinal operation is warranted for both diagnostic and therapeutic purpose in uveitis patients in whom medicamentous therapy fails to provide a satisfactory control of the disease and its complications3,12.

Case Report

A 12-year-old girl was hospitalized for bilateral chronic uveitis that had just been detected due to poor vision on the left eye (0.05), free from any other symptoms of uveitis. Chronic uveitis and inflammatory infiltration of the vitreous were found bilaterally, abundant diffuse subretinal circumferential exudate occasionally appearing as a solid mass with detached retina on the left, and with discrete diffuse lesions at the level of pigment epithelium without exudative lesions on the right. The patient underwent complete diagnostic work-up that produced the following findings: complete blood count, erythrocyte sedimentation rate, urine, ACE, liver enzymes, urea and creatinine, protein testing normal; lung x-ray normal; tuberculin test ---; testing for syphilis, Lyme borreliosis, toxoplasmosis negative; Toxocara serology positive; IgG 1:100; ANA positive; HLA typing: positive for HLA B27 and HLA B51.
The patient was prescribed local therapy and systemic corticosteroid therapy for uveitis, along with isoniazide and pyridoxine because of the close contact with her father suffering from lung tuberculosis although she showed no signs of the disease. With this therapy, her ocular disease showed slight and temporary regression, however, the patient presented for the disease exacerbation 8 months later. The left eye showed exacerbation of the disease with subretinal mass progression. Visual acuity on the left eye was reduced to light perception of uncertain projection. The right eye showed no change and had normal visual acuity. Diagnostic-therapeutic vitrectomy was performed, the vitreous exudate and subretinal masses were removed, silicone oil endotamponade was performed. The intraoperatively obtained material was referred for cytology, histology and immunohistochemistry analysis. Polymerase chain reaction (PCR) for herpes virus and cytomegalovirus was performed.

Histologic examination of the subretinal material revealed connective tissue with abundant histiocyte accumulation and retinal histiocytic infiltration. Immunohistochemistry staining identified the histiocyte specific CD68 marker. Rare eosinophils were detected in the vitreous. PCR was negative.

Additional examinations included sinus x-ray, computed tomography of the brain, orbits and sinuses, skeletal technetium scintigraphy, and sputum tear culture for tuberculosis, all producing negative results. Because of the complex nature of the condition, consultation was obtained from a hematologist, an immunologist and a pulmonologist. The thorough and detailed examinations revealed no systemic manifestation of the disease.

Postoperatively, secondary glaucoma developed on the operated eye and silicone oil was removed after 6 weeks. Six months of the silicone oil removal, the condition of the eye was stable. The retina was properly attached, there were no signs of the disease relapse, only the development of posterior subcapsular cataract was recorded.

Discussion

Analysis of the course of the disease in this 12-year-old patient confirms the hypothesis that uveitis may pose great diagnostic and therapeutic difficulties. The severe vision loss on the left eye and absence of symptoms on the right eye indicated long duration of the disease before the first hospitalization. Initial examinations suggested the possible presence of particular pathologic entities. The family history and positive tuberculin test pointed to tuberculosis as a potential cause of the disease and the patient was initially treated with tuberculostatics, as advised by the pulmonologist. However, the poor response to this therapy, the absence of pulmonary disease, and the histopathologic analysis of intraocular material that was not indicative of caseous granuloma, led us to conclude that uveitis had not been caused by tuberculosis.

The positive Toxocara titer, IgG 1:100, was not accompanied by other signs of infection with this parasite (peripheral blood eosinophilia), and the fundus finding was not typical (diffuse peripheral subretinal masses), yet the finding of scarce eosinophils in the vitreous and histiocytic infiltration may have supported the diagnosis of intraocular toxocariasis. However, the poor response to systemic and local corticosteroid therapy was against it, so we concluded it to be a case of histiocytosis.

Positive HLA-B51 suggested Behcet’s disease although ocular lesions were not specific for this disease. As there were no systemic manifestations of this disease, it was ruled out in consultation with an immunologist.

While there are arguments supporting the potential role of the above mentioned causes in the occurrence of chronic uveitis, for a number of reasons we believe that our patient suffered from histiocytosis. There was abundant infiltration of the retina and subretinal region with CD68 positive histiocytes, without other elements to point to specific inflammation. The clinical finding of the retina was not typical for either tuberculosis or toxocariasis. The intraocular inflammation was unresponsive to treatment with tuberculostatics and corticosteroids. Negative test results ruled out systemic histiocytosis, however, isolated ocular disease could not thus be excluded, as also reported in the literature. Chronic uveitides that respond poorly to classic immunosuppressive therapy require multidisciplinary analysis of intraocular material. Pars plana vitrectomy is a standard and safe procedure making intraocular material accessible for analysis, and is also a highly successful method in the treatment of chronic uveitis complications.

References

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