Int J Clin Exp Med 2016;9(2):4820-4824
www.ijcem.com /ISSN:1940-5901/IJCEM0017484

Case Report

Idiopathic orbital inflammatory pseudotumor (IOIP) accompanied with panuveitis: a case report and review of the literature

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Received October 8, 2015; Accepted December 31, 2015; Epub February 15, 2016; Published February 29, 2016

Abstract: Idiopathic orbital inflammatory pseudotumors (IOIP) are non-granulomatous inflammatory process of the orbit for unknown etiology, and are associated with diagnostic challenge and high recurrence rate. In this report, we present an unusual case of IOIP initially accompanied with panuveitis. A 57-year-old female presented with a two-month history of decreased vision of the right eye for no obvious reason at her first visit. She had intermittent attacks of ophthalmalgia, photophobia, foreign body sensation and headache. Clinical examination of the right eye revealed a diminished visual acuity of the right eye (VOD), a moderately deep anterior chamber, opisthosynechia at Pars iridica, slight opacification and pigmentation of crystalline lens, and vitreous opacification. Hematological investigations revealed an increased number of leukocyte, cell-hemoglobin concentration. Immunologic test showed higher blood sedimentation and complement C4. She was initially diagnosed with panuveitis and improved after the systemic administration of Methylprednisolone Na succinate by intravenous drip (0.75 g/d) for 5 d and local application of I-Prednicet, atropine ophthalmic gel and so on. However, she had twice relapsed thereafter, and further diagnosis of IOIP was made. Finally she was given enucleation of eyeball and orbital exenteration. The diagnosis of IOIP should be strongly considered in the presence of non-granulomatous inflammatory process and a suggestive MRI image. Treatment of IOIP accompanied with panuveitis should be started immediately to improve visual acuity of the eye, reduce inflammatory symptoms and preserve eyeball functions. The preferred treatment of choice was systemic treatment combined with local hormone therapy.

Keywords: Idiopathic orbital inflammatory pseudotumor, panuveitis, non-granulomatous inflammatory process, MRI

Introduction

Idiopathic orbital inflammatory pseudo-tumors (IOIP), also called pseudo-tumor of orbit, are non-granulomatous inflammatory process of the orbit for unknown etiology [1]. It was the third most common ophthalmic disorder secondary to Grave's disease and lymphoproliferative disorders, and accounts for approximately 7%-12.3% of all the orbital tumors [2]. Currently, the diagnosis of IOIP is mainly one of exclusion based on case history, clinical examination, and histological examination or immunologic staining of the lesions collected from surgical resection or biopsy [3, 4]. Unfortunately, the lack of a variety of specific clinical symptoms and well-defined pathogenic mechanisms has made its diagnosis and treatment a challenge to ophthalmologist [3, 5, 6]. Besides, IOIP is usually associated with a higher incidence of relapse in despite of surgical resection as the most treatment of choice, as well as radiotherapy and other anti-inflammatory drugs for symptomatic treatment.

Herein, we report a case of IOIP initially accompanied with panuveitis, highlighting the diagnostic challenge and high recurrence rate in such a case.

Case report

A 57-year-old female presented with a two-month history of decreased vision of the right eye for no obvious reason at her first visit to our hospital on May 5, 2014. She had intermittent

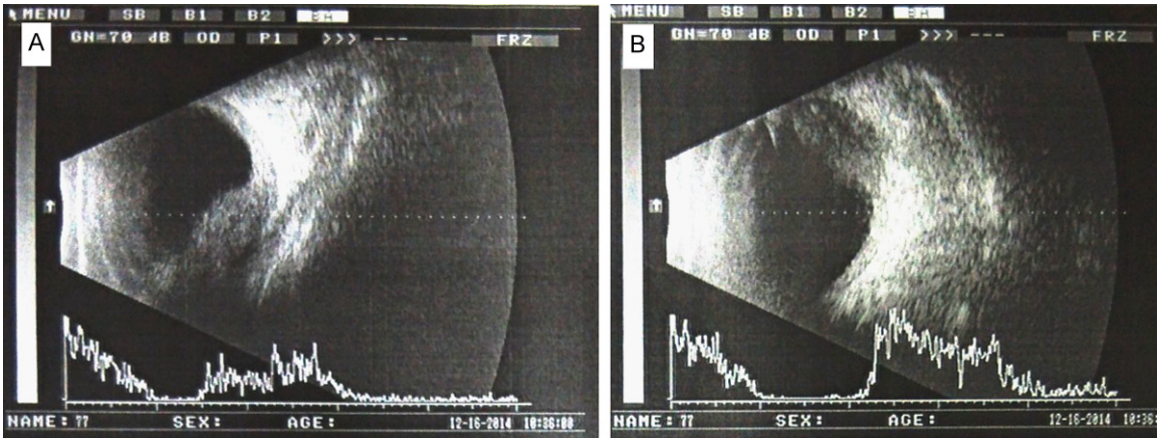

Figure 1. B-ultrasonography showed an abnormally hyperechoic area behind the right eyeball with incomplete boundary (A, B).

attacks of ophthalmalgia, photophobia, foreign body sensation and headache, and was diagnosed with keratitis of the right eye in the local hospital. There was no history of a blow or of other injuries to the orbit. Clinical examination of the right eye revealed a diminished visual acuity of the right eye (VOD) to 0.02, mixed congestion of conjunctiva (++), keratic precipitates (+), a moderately deep anterior chamber, Tyndall phenomenon (+++), a not well orbicular pupil (4.5 mm in diameter), opisthosynechia at Pars iridica, slight opacification and pigmentation of crystalline lens, and vitreous opacification (+++), no sign in the fundus oculi. B-ultrasonography also displayed vitreous opacification of the right eye. Thereafter she was admitted to our hospital for panuveitis of the right eye. Hematological investigations revealed an increased number of leukocyte, cell-hemoglobin concentration, as well as decreased percentage and absolute number of eosinophil. After her hospitalization, immunologic test showed higher blood sedimentation and complement C4, with no abnormalities in serum levels of antinuclear antibody. Consultation of doctors from Department of Rheumatism and Immune and Urology Department was non-contributory. During the hospitalized period, she was given the systemic administration of Methylprednisolone Na succinate by intravenous drip (0.75 g/d) for 5 d and local application of I-Prednicet, atropine ophthalmic gel and so on. Finally she was getting better and discharged from hospital with drugs on May 16. The final examination prior to discharge revealed an improved VOD to 0.04, no congestion of conjunctiva, transparent cornea, a moderately deep anterior chamber, Tyndall phenomenon (-), a not well orbicular pupil (4.5 mm in diameter), opisthosynechia at Pars iridica, slight opacification of crystalline lens, and vitreous opacification (++), retina resupination in the fundus oculi.

At her return visit to our hospital on May 31, 2014, clinical examination revealed improved vitreous opacification (+) and a real swelling of the bitamporal retina in the fundus oculi. B-ultrasonography showed an abnormal hyperechoic area behind the right eyeball with incomplete boundary (**Figure 1**); Color Doppler Ultrasonography displayed equal echo echogenic area in the right eyeball, with the occupying lesion taken into account and suspected angioma remained to be excluded. Plain and contrast enhanced MRI of the orbit showed an upper eye ring-occupying lesion outside of the eyeball (**Figure 2**), considering the melanoma of choroid and the possibility of invasion in the right lacrimal gland. Then she was admitted to our hospital again on June 16. Following a remote consultation of doctors from Beijing Tong Ren Hospital Oncology Service, inflammatory lesions (granulomatous inflammation) of the right eye or masquerade syndrome (lymphoma) was suspected. She was suggested to receive the current therapy and surgical biopsy of lesions outside of the eyeball wall super-temporally. Based on the pathologic diagnosis of severe chronic inflammation, the tumor ablation of the right orbit was performed, and the frozen section of the tumor was confirmed benign. Thereafter, she was given radiotherapy as well as anti-inflammatory for symptomatic

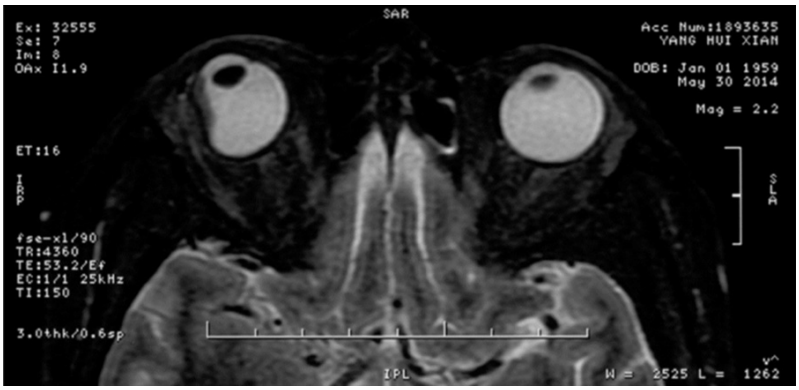

Figure 2. Plain and contrast enhanced MRI of the orbit showed an upper eye ring-occupying lesion outside of the eyeball.

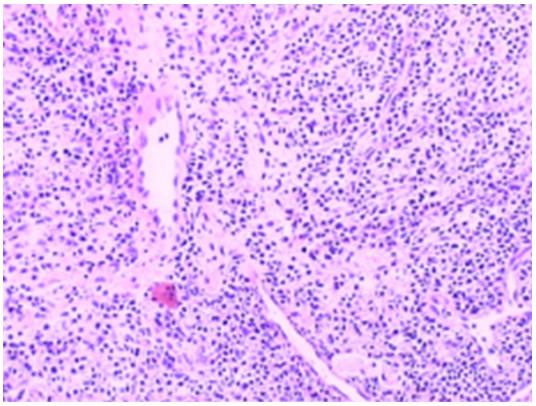

Figure 3. Immunohistochemisty (×200) showed chronic inflammation accompanied with active plasmacytes proliferation.

treatment after her transfer to department of Oncology on June 25, and was discharge from hospital 10 days later.

On July 24, she was admitted to other hospital with similar complains. The clinical examination of the right eye revealed a visual acuity of no light perception, mild congestion of conjunctiva, transparent cornea, a moderately deep anterior chamber, Tyndall phenomenon (-), a not well orbicular pupil (4.5 mm in diameter), opisthosynechia at Pars iridica, no light reflex, slight opacification of crystalline lens, and vitreous opacification (++), and a real swelling of the bitamporal retina in the fundus oculi. Plain and contrast enhanced MRI of the right orbit showed an occupying lesion along the exterior and interior wall outside of the eyeball, more prone to malignant tumor, especially for melanoma. B-ultrasonography showed an occupying lesion of the right eyeball wall, and positron emission tomography/computed tomography (PET/CT) scan revealed soft tissue mass inside the right vitreous body and increased glucose metabolism, considering malignant tumor (melanoma of choroid). Thereafter, she was given enucleation of eyeball and orbital exenteration on August 6. The postoperative pathologic examination displayed small cell lesion and immunohistochemisty showed chronic inflammation accompanied with active plasmacytes proliferation (**Figure 3**).

Discussion

Although IOIP is the third commonest cause of noninfectious orbital disease [7], it is rare accompanied with panuveitis. Here in this case report, we presented an unusual case of IOIP accompanied with panuveitis, who initially suffered from a decreased VOD for no obvious reason paroxysmal cough, intermittent attacks ofophthalmalgia, photophobia, foreign body sensation and headache.

Panuveitis is an inflammation of both parts of the uvea, consisting of the anterior segment (iris and ciliary body) and the posterior segment (chorioretina and optic disc). It could be caused by infections, chronic inflammatory diseases or for no obvious reason [8], and idiopathic panuveitis is also a diagnosis of exclusion owing to the lacks of distinguishing features [9]. Notwithstanding the exact etiology of IOIP remained unknown, the autoimmune diseases, reparative processes following inflammation, acute infections or even neoplasia have been postulated [10]. Hence, it seemed that the simultaneous findings of both IOIP and panuveitis was a coincidence, and might be explained by a tendency towards a concurrent occurrence of autoimmune diseases probably due to some kind of common antigen between orbit and uvea tissues.

A great number of orbital structures might be involved in the inflammatory process of IOIP, including eyeball, fat, extraocular muscle, lachrymal gland, connective tissues, and the optic

nerve [11, 12]. Consistently, the case in the present report developed a diminished VOD, conjunctival congestion, occupying lesion within the orbit, well orbicular pupil, opisthosynechia at Pars iridica, slight opacification and pigmentation of crystalline lens, various degrees of opacification of crystalline lens and vitreous body. MRI has been considered as a valuable procedure of choice for the diagnosis of orbital pseudotumor [13]. Both the two MRI scans indicating an occupying lesion within the orbit during her seeking treatment.

The aim of therapy is to improve visual acuity of the eye, reduce inflammatory symptoms and preserve eyeball functions. Surgical resection was the best treatment of choice, and orbital inflammatory pseudotumor also showed excellent response to steroid [14], but a high number of cases might recur or become steroid dependent. The preferred treatment of choice was systemic treatment combined with local hormone therapy, generally using 1 mg/kg/d prednisone for 1-2 weeks and Methylprednisolone Na Succinate for severe case, with decrement of 5-10 mg every two weeks thereafter. Besides, topical steroids eyedrops and mydriasis therapy should be also considered.

There are a few learning points from this case. The diagnosis of IOIP should be strongly considered in the presence of non-granulomatous inflammatory process and a suggestive MRI image. Treatment of IOIP accompanied with panuveitis should be started immediately to improve visual acuity of the eye, reduce inflammatory symptoms and preserve eyeball functions. The preferred treatment of choice was systemic treatment combined with local hormone therapy. Given the rarity of this disease, the pathogenesis is still uncertain at present. In the future, further studies of additional cases and long-term follow-up will be necessary to completely understand the disease for better clinical management.

Acknowledgements

This work was supported by the Applied Basic Research Program of Yunnan Provincial Science and Technology Department (Grant No: 2014FB076) and the Applied Basic Research Program of Health and Family Planning Commission of Yunnan Province (Grant No: 2014NS041). We would like to acknowledge the reviewers for their valuable and helpful comments on the case report.

Disclosure of conflict of interest

None.

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