Atypical Early Presentation Of Orbital Pseudotumor: A Case Report
Y Chen, Y Cheng, C Lui, C Huang, W Chen

Introduction
Craniofacial disorder is a common complaint presenting to general practitioner who should rapidly localize and differentiate in order to prevent adverse event. Orbital pseudotumor (OPT), which is firstly reported at 1903 (1) and formally coined at 1905 (2), is an idiopathic inflammatory disorder of orbital structures amenable to treatment (3). Although it is considered a benign disorder in the traditional concept, it may occasionally complicate with irreversible visual or oculomotor deficit (4,5,6). OPT usually occurs acutely, and is characterized by its clinical quadriad, including proptosis, oculomotor deficits, lid swelling or mass, and especially periorbital/retro-orbital pain in early (5,7,8), regardless of ethnicity (7,9). A typical manifestation is necessary for making a rapid and correct diagnosis. Herein, we report a patient who initially exhibited an atypical manifestation for his OPT rendering a difficulty for early diagnosis.

Case Report
A 40-year-old man experienced an acute onset of bubbling sensation over his left temporal area at one evening. On the next day morning, he found a redness and an equivocal eyelid drooling of his left eye. He consulted ophthalmologists who revealed conjunctivitis only. Five days later, excessive tearing, proptosis, and finally, periorbital pain, subsequently appeared. In addition, tingling pain at the inner side of left eye was provoked by medial eyes gazing. He consulted our service sooner as there was no spontaneous improvement of his ocular problem. He denied medical disease, craniofacial trauma, migraine or cluster headache, infection, or consumption of illicit drug before.

On presentation, his vital signs were stable. Proptosis, ptosis, edema and chemosis of left eye were clearly seen. There was no bruit heard over the temporal area or eyes. The eyegrounds did not show papilloedema or hemorrhage. Visual field was not restricted and visual acuity was preserved. Pupils were isocoric and reflexic. There was no limitation of eyes gazing to either directions, but mild pain at the left eye was provoked by medial eye gazing. He could elevate his left eyelid completely in afford. Otherwise, there was no additional neurological deficit detected. There was no pain at sinus or vertebrate. Nodulation or tenderness was not found at the superficial temporal artery. Clinically, partial Horner's syndrome or cavernous sinus disorder, such as cavernous sinus thrombosis or carotid-cavernous fistula, was differentiated. Laboratory tests, including biochemistry, hematology, urine analysis, antinuclear factor, ds-DNA, lupus anticoagulant, thyroid function, microsomal antibody, and antithyroglobulin antibody, were within reference range, except an elevation of erythrocyte sedimentation rate to 28 mm (reference range: < 20 mm) and C-reactive protein 15.6 mg/L (reference range: < 5 mg/L). The serum IgG level was 865 mg/dl (700-1600 mg/dl) and IgG\textsubscript{4} 54.4 mg/dl (reference range: 3-200 mg/dl). Chest radiography did not show active cardiopulmonary change. Head magnetic resonance imaging...
revealed left eye proptosis, periorbital edema, and thickening of the left lateral rectus muscle, as well as an involvement of the lacrimal gland (Fig. 1A). There was no engorgement or occlusion of the left ophthalmic vein or cavernous sinus, nor a consistent change of intracranial artery. Taken together, orbital pseudotumor was established.

After inform to patient, corticosteroid pulse therapy was initiated. His proptosis, ptosis, lid drooling, chemosis, tearing, as well as pain in medial eye gazing, of left eye subsided simultaneously and rapidly. The erythrocyte sedimentation rate was restored to 12 mm and C-reactive protein 0.6 mg/L, respectively. The head magnetic resonance imaging also disclosed a significant diminution of proptosis, periorbital edema and thickening of lateral rectus muscle at left eye (Fig. 1B).

**DISCUSSION**

Generally, a correct diagnosis of OPT can easily be made basing on its characteristic neuro-ophthalmic manifestations (7,8). In our patient, the dilemma is an absence of the typical painful eye, oculomotor deficit and ocular edema at initial, and an isolated equivocal eyelid drooling and chemosis may be misinterpreted as an local conjunctivitis, or even the ocular sympathetic dysfunction or cavernous sinus disorder. In fact, atypically early manifestation of OPT has only been mentioned in a few sporadic cases before, such as chronic visual impairment (10), painless proptosis (11,12,13), chronic palpebral edema (14), isolated abducens nerve palsy (15), and isolated oculomotor myositis (16). Therefore, OPT may present atypically and simulate any ophthalmic or neuro-ophthalmic disorder, and renders the diagnostic difficulty to general practitioner.

Currently, there is no established diagnostic criteria for OPT or differentiating OPT from other orbital pathologies (3). An open biopsy is not a routine as it can provide limited diagnostic aid only in a few conditions (8,17). In practice, the diagnosis of OPT is based on history, clinical presentation, imaging finding, and response to treatment as in our patient. Basing on the radiological and surgical findings, OPT is mainly categorized as myositis, dacryoadenitis, anterior, apical and diffuse types (4,5). Their clinical pictures actually overlap with each others. In our patient, a diffuse type is favored due to an involvement of left lateral rectus muscle, lacrimal gland and other orbital structures.

Till now, the actual mechanism of OPT is still a matter of unknown. In our patient, an elevation of erythrocyte sedimentation rate and C-reactive protein, which relates to inflammation, was detected upon presentation. Indeed, OPT occasionally associates with autoimmune disorder (18,19), infection (19,20,21), or aberrant wound healing (19). Recently, an IgG_{4}-related lymphoplasmacytic infiltrative disorder is reported to involve multiple organs, including the orbit (22). It is considered in relating to the sclerosing type of OPT (23,24). In our patient, his blood IgG and IgG_{4} level did not increase but this finding does not conclude an exclusion of IgG_{4} in other types of OPT.

Similar to previous reports, our patient also showed a favorable response to corticosteroid treatment (7). The proptosis, ptosis, chemosis, and painful ocular motility in our patient improve simultaneously after corticosteroid pulse therapy. Indeed, the neuro-ophthalmic deficits in previous
cases of atypical OPT also responded well to corticosteroid therapy (10-16). These therapeutic findings may reflect the unique underlying inflammatory nature despite of distinct clinical manifestations or types in OPT.

CONCLUSION

We report a patient with OPT who exhibit an atypical manifestation at initial. The blood IgG and IgG4 did not increase. Corticosteroid treatment offered a favorable response. Since OPT may complicate with disable neuro-ophthalmic sequelae, it should be alerted in cases of atypically neuro-ophthalmic deficit. There is no evidence to support a different OPT type-specific treatment response to corticosteroid.

References

Author Information

Yi-Ting Chen, MD
Department of Family Medicine, Kaohsiung Chang Gung Memorial Hospital

Yuan-Yu Cheng, MD
Department of Ophthalmology, Kaohsiung Chang Gung Memorial Hospital

Chun-Chung Lui, MD
Department of Neuroradiology, Kaohsiung Chang Gung Memorial Hospital

Chih-Fang Huang, MD
Department of Family Medicine, Kaohsiung Chang Gung Memorial Hospital

Wei-Hsi Chen, MD MSc LLM
Department of Neurology, Kaohsiung Chang Gung Memorial Hospital