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Sino-Orbital Cellulitis Due to HSV Type I

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Abstract: Orbital pseudotumor, is a heterogeneous group of disorders characterised by orbital inflammation and is a rare clinical entity and a diagnosis of exclusion. We report two cases with typical clinical manifestation and magnetic resonance images, but unresponsive to corticosteroid treatment which turned out to be caused by herpes simplex virus type I. Viral etiology must be carefully evaluated, especially in steroid unresponsive recurrent cases presumed to be pseudotumor orbita.

Keywords: Orbital Cellulitis, Pseudotumor Orbita, Herpes Simplex Virus, Painful Ophthalmoplegia

1. Introduction

Painful ophthalmoplegia is a common clinical presentation for all neurologists and ophthalmologists. There is a long list of differential diagnosis, including vascular, infective, rheumatologic causes and ophthalmologic migraine [1]. Despite improvements in laboratory and scanning techniques, most cases interpreted idiopathic. Extraocular orbital and adnexal inflammation with no identifiable local or systemic cause is often referred to as orbital pseudotumor. It remains a diagnosis of exclusion and is characterized by its chronicity, anatomic location, or histologic subtype.

Corticosteroids are the mainstay of treatment inducing a rapid and dramatic reversal of inflammatory findings. Their mechanism of action is attributable to both anti-inflammatory and immunosuppressive properties. Despite the excellent and rapid response to corticosteroids, failures reported and unusual etiologic causes should be interpreted recurrent steroid unresponsive cases [2]. In this paper we present two cases of sino-orbital cellulites presumed to be orbital pseudotumor, unresponsive to corticosteroid treatment, due to herpes simplex virus type I.

2. Case Report

Case 1: A 67-year-old male admitted to our department with major complaints of orbito-frontal pain, ptosis and vision loss on the left side. His complaints began 25 days before admission with diplopia and blurred vision and got worse gradually. As left sided ptosis and vision loss occurred two days ago, he decided to seek medical assistance. Type II diabetes mellitus was under control with oral antidiabetics for 8 years. His neurological examination revealed complete loss of function in the left II-III-IV and VI cranial nerves. Initial Snellen visual acuity was light perception and relative afferent papillary defect was detected in the affected eye. Retinal examination did not reveal diabetic retinopathy. The examination of the fellow eye was fully normal.

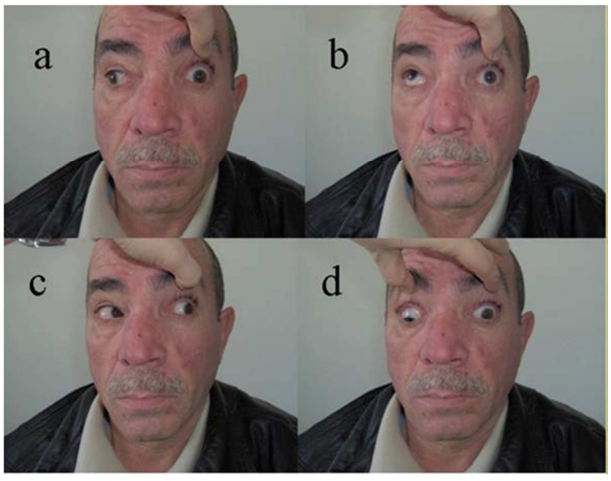

Figure 1. *Total ophthalmoplegia on the left side (Case 2).*

Case 2: A 51-year-old male admitted to our department with the complaints of orbito-frontal pain, diplopia and ptosis on the left side lasting for three days. His medical history

revealed that he had same symptoms on the left side in 1985 and 1995, and on the right side in 2003. He claimed that his previous complaints had resolved in 2-3 months without any treatment. Neurological examination revealed complete loss of function in left III-IV-VI cranial nerves “Figure 1’’. Biomicroscopic and fundoscopic examination were fully normal in both eyes. Relative afferent papillary defect was not detected.

Both patients treated with methylprednisolone 1000 mg/day IV for 5 days but no clinical improvement was observed. Biochemical (blood urea nitrogen, creatinine, transaminases, sodium, potassium, erythrocyte sedimentation rate, thyroid stimulating hormone, free T4, angiotensin-converting enzyme level, serum protein electrophoresis), rheumatologic (anti-nuclear antibodies, anti-cardiolipin antibodies, anti-dsDNA, antineutrophil cytoplasmic antibodies), cerebrospinal fluid (CSF) analyses (protein, glucose, microscopy), cerebral magnetic resonance imaging (MRI) and angiography performed for possible etiological causes. Mild elevated CSF protein and leucocytosis was evident. Left sided proptosis was the only pathological finding in both patients. Fatsat turbo spin echo orbital MRI performed to demonstrate possible inflammatory or mass lesion. Orbital MRI revealed contrast enhancement expanding from posteromedial sight of periorbital adipose tissue to optic nerve and cavernous sinus in the first case ‘’Figure 2’’. Contrast enhancement expanded from retroorbital adipose tissue to maxillary sinus anterior wall in the second case ‘’Figure 3’’. Preliminary diagnoses were pseudotumor orbita. Biopsy was planned to rule out possible malignancies.

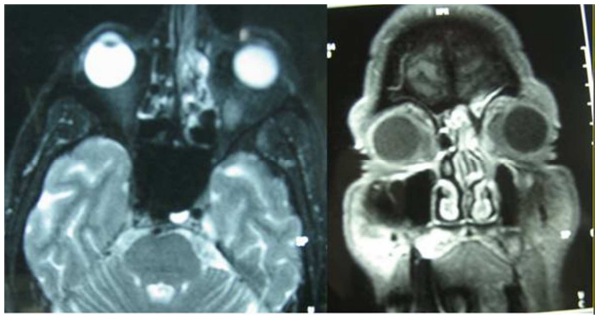

Figure 2. *Fatsat turbo spin echo MR showed contrast enhancement, spreading from posteromedial sight of periorbital adipose tissue to optic nerve and cavernous sinus (Case 1).*

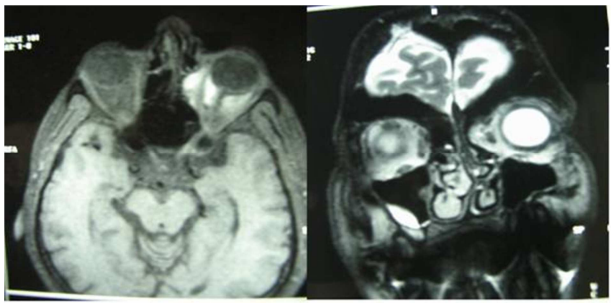

Figure 3. *Fatsat turbo spin echo MR showed contrast enhancements, spreading from retroorbital adipose tissue to maxillary sinus anterior wall (Case 2).*

Neurosurgery and otorhinolaryngology departments together, performed intranasal endoscopic surgery and cytological specimen were obtained from inflammatory tissue passing through lamina papricea. Pathology department reported focal adipose necrosis, chronic inflammatory adipose tissue and findings of mild myositis for both samples. This material was subjected to microscopy, cultures for viruses, microorganisms, and for polymerase chain reaction (PCR) techniques for cytomegalovirus, Epstein-Barr virus, herpes simplex virus (HSV), varicella zoster virus (VZV), and toxoplasmosis. The only test that was positive was PCR for HSV. Sequencing revealed HSV type 1 (all analyses were performed twice).

Patients’ symptoms slightly improved after surgery probably due to decompression effect. Both patients were treated with acyclovir 800mg/qid PO for 5 days. The follow up examination revealed that first patient’s symptoms regressed partially in three months. Relative afferent papillary defect persisted however visual acuity improved to counting fingers from one meter. Complete recovery was seen in the second case in a month.

3. Conclusion

Although the diagnosis of orbital pseudotumor is usually a clinical one, the evaluation should begin with an imaging study. High-resolution orbital computed tomography (CT) may demonstrate soft tissue swelling but is not as sensitive as MRI. Contrast-enhanced MRI with multiple coronal views and fat saturation should be the initial diagnostic study performed, when available [3]. Orbital pseudotumor MRI studies revealed mass lesions that were hypointense to orbital fat on T1-weighted images and isointense or minimally hyperintense to fat on T2-weighted images [4]. Even rarely, orbital pseudotumor may spread to extraorbital areas as seen in our cases with cavernous sinus involvement in the first case and with maxillary sinus involvement in the second case [5, 6, 7].

The etiology of orbital pseudotumor is not clear, but infections (fungal, bacterial, viral), inflammatory syndromes (idiopathic orbital inflammation, idiopathic sclerosing inflammation, Sarcoidosis), autoimmune disorder (Graves' ophthalmopathy, vasculitis), neoplastic processes (primary orbital tumors, metastatic tumors), vascular lesions (carotid cavernous fistulas, cavernous sinus thrombosis) and aberrant wound healings have been put forward as possibilities. In addition orbital pseudotumor has been associated with systemic disorders, including rheumatoid arthritis, Crohn's disease and systemic lupus erythematosus [1, 8-12].

The diagnostic evaluation of a patient with suspected orbital pseudotumor should include a full hematologic work-up with complete blood cell count, electrolytes, sedimentation rate, antinuclear antibodies, anti-dsDNA, antineutrophil cytoplasmic antibodies, angiotensin-converting enzyme level, thyroid stimulating hormone, free T4 and serum protein electrophoresis [2, 13]. Cerebrospinal fluid examination is rarely helpful but should be performed

in questionable cases or when cytology is deemed useful to exclude lymphoma [3]. It is crucial to differentiate between orbital pseudotumor and the disorders that may mimic it. We also performed this extended laboratory work up for our cases but only mild elevated CSF protein and leucocytosis was evident. These laboratory findings did not help in differential diagnosis.

Response to therapy often confirms the diagnosis but in our cases corticosteroid treatment was unresponsive. Biopsy is only indicated if there are rapidly progressive neurologic deficits, a lack of steroid responsiveness, or persistent abnormalities on neuroimaging studies [2]. Chronic inflammatory cells infiltration in various orbital tissues are the main histopathologic features in patients with orbital inflammatory pseudotumor [9]. Chronic inflammation observed in our specimens did not yield any etiological idea. Although the clinical examination and MRI findings were confirmative for orbital pseudo tumor, classical laboratory work up was insufficient to put forward the etiology. Virology study of biopsy specimen was surprising which revealed HSV-DNA was present in both cases, establishing the diagnosis of sino-orbital cellulitis.

Systemic corticosteroids - in the form of oral or intravenous administration- are generally considered mainstay therapy for orbital pseudotumors. Recurrences even after initial corticosteroid treatment have been reported in orbital pseudotumor [15, 16, 17]. Spontaneous remission may also occur [18]. There is a similarity with peripheral facial paralyses because they share the same clinical features; cranial nerve palsy, often dramatic response to corticosteroid therapy, spontaneous remission, usually complete improvement in 2-3 months. MRI findings resemble with different localization [19]. Recent studies support viral etiology and antiviral therapy for Bell's paralyses, sustained as idiopathic perennial [20]. Probable viral etiology seems reasonable with this available data. Ophthalmoplegia associated with herpes virus family, mainly herpes zoster, is not uncommon. As the majority were due to involvement of the ocular motor cranial nerves, they are traditionally interpreted as diseases of III, IV or VI cranial nerves [21, 22, 23, 24]. Ophthalmoplegia due to viral myositis have also been reported [25, 26, 27]. To our knowledge, no prior cases of HSV-induced sino-orbital cellulitis with multiple cranial nerve palsies have been described in the literature. After antiviral treatment, complete resolution was observed in the second case. Insufficient response observed in the first case was probably due to late timing of the proper treatment. There is no particular well-established protocol to guide adjuvant therapy for refractory orbital pseudotumor. Among the available options for the management of refractory orbital pseudotumor is: radiation therapy, cytotoxic agents (Cyclophosphamide and Chlorambucil), immunosuppressants (Methotrexate, Cyclosporine, Azathioprine), IV immunoglobulins, TNF-alpha inhibitor, monoclonal antibody (Infleximab and Adalimumab) and Mycophenolate Mofetil which inhibit denovo purine synthesis and prevent B & T lymphocyte replication [1, 2, 9, 28-32].

Orbital pseudotumor is an unusual inflammatory condition established by typical clinical and imaging findings. Orbital pseudotumor is usually quite responsive to corticosteroids. Corticosteroid unresponsive cases, especially with recurrences, should be interpreted carefully as this type of course is unusual. Biopsy is not always necessary, but it might be helpful to put forward the specific cause. Before initiating any treatment, atypical clinical or imaging findings should prompt biopsy. Although evidence is limited, anti-viral therapy should be kept in mind for these atypical cases. These cases showed that we should hasitate in pronouncing the diagnosis of "pseudotumor" before viral evaluation of biopsy specimen if possible.

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