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CASE REPORT

CASE REPORT: IDIOPATHIC SCLEROSING ORBITAL INFLAMMATION

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Abstract

Idiopathic sclerosing orbital inflammation (ISOI) also known as sclerosing orbital pseudotumor is a rare, idiopathic, chronic, slowly progressive orbital inflammation, characterized by scarring of the tissue that represents a mass effect. The latter leads to bulging of the eye, restriction of ocular movements and visual impairment up to potential loss of vision. It is a subtype of non-specific inflammatory orbital pseudotumor, which both radiologically and clinically mimics a malignant process or other orbital inflammatory disease, and therefore is challenging to diagnose and manage. This case highlights the challenges in diagnosis and management of ISOI.

Keywords: *idiopathic sclerosing orbital inflammation, orbital sclerosing pseudotumor, orbital pseudotumor, diagnosis, management*

Introduction

Idiopathic sclerosing orbital pseudotumor, is a benign, non-infectious, non-specific inflammatory condition of the orbit in which no local or systemic cause can be identified.[1] It manifests with proptosis, eyelid edema, chemosis, lagophthalmos, diplopia, optic neuropathy. The diagnosis is obtained by exclusion.[2] HPE is the gold standard currently which shows non-malignant lymphocytic infiltration and dense fibrous tissue. The sclerosing type presents a more aggressive condition and is less responsive to steroids. In this case report, we describe the management of a 52-year-old woman with orbital pseudotumor at Ophthalmological center after S.V. Malayan in Armenia, who presented with painless, binocular progressive proptosis. Informed consent was obtained from his family before publication of this report.

Case Presentation

A 52-year-old female presented to the Malayan Eye Center, with complaints of gradual protrusion of the right eyeball, redness, cosmetic discomfort,

tearing and severe visual impairment. According to the patient's relatives, the complaints started 4 years ago, with a slight, painless bilateral protrusion of the eyes and the patient applied to another hospital, where she underwent appropriate laboratory tests to rule out thyroid eye disease. The tests showed normal thyroid function.

Figure 1. *Bilateral proptosis, right eyeball subluxation*

The patient also reported that she was diagnosed with paranoid schizophrenia in 2004 and was receiving an appropriate psychiatric therapy. Therefore, she did not continue her eye examination for some time.

On clinical examination in our clinic, there was a total subluxation of the right eyeball, a solid mass was palpated in the medial part of the upper eyelid. Other symptoms present were: chemosis, lagophthalmos, eyelid edema, keratopathy, tearing, visual impairment, mild pain, severe restriction of ocular movements (Figure 1). The visual acuity of the patient in the right eye was determined as a hand motion and 20/30 in the left eye.

The patient underwent CT-scan of the orbits, which revealed a multinodular non-encapsulated mass of 52 x 40mm in the right orbit, with uneven edges, indistinct boundaries, extending from the orbital apex along the medial wall of the orbit. A lot of small nodules were also found in the left orbit, with the same CT features (Figure 2).

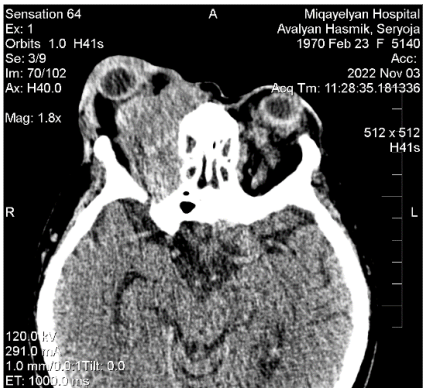

Figure 2. *On CT-scan: non-encapsulated, multinodular masses in both orbits with bilateral proptosis; no signs of bone destruction*

The muscles and the eyeballs were not involved in the process. There were no destructive changes observed in the bones as well as in the brain tissue. The radiological impression was a suspected orbital pseudotumor, although orbital lymphoma could not be excluded. The patient's blood test results were within normal limits.

Based on the patient's complaints and clinical and radiological examination data, a preliminary diagnosis of intra-orbital neoplasm was made. Since the malignant origin of the neoplasm was not excluded, in order to clarify the diagnosis and determine the tactics of further treatment, an incisional biopsy was performed from the medial part of the upper eyelid. The removed tissue was sent for histopathological examination (HPE), which showed no evidence of malignancy, nut instead suggested a preliminary diagnosis of pilomatrixoma, due to the presence of foreign body-type giant cells, amorphous eosinophils, and basophilic cells in the sample.

This answer was quite unexpected, as it is known that pilomatrixoma, which is a benign skin tumor, originates from the matrix of the hair follicle. Whereas in the case of our patient, the mass grew in the orbit and did not involve the skin of the eyelids. Due to the nonspecific finding, we reviewed several case reports related to pilomatrixoma and found that sometimes, very rarely, pilomatrixoma of the orbit can also occur, furthermore, the symptoms of pilomatrixoma were consistent with our patient's symptoms.

Based on the fact that the patient's eye retained some vision and that we were dealing with a clearly non-malignant mass, the previous plan of possible exenteration of the eyeball ruled out, and a surgical

debulking of the intra-orbital mass was performed by an anterior orbitotomy (Figure 3).

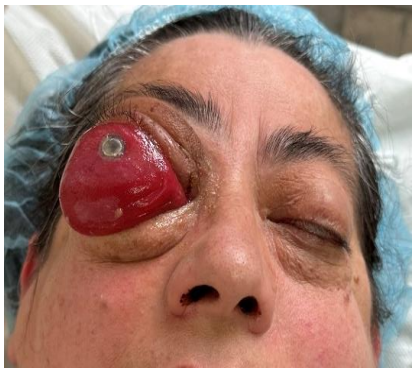

Figure 3. *The condition of the eye before operation*

The mass was found in the orbit and acutely separated within the boundaries of healthy tissues (Figure 4). The eyeball was placed back into orbit. Medial, central and lateral tarsorrhaphies were performed with complete closure of the palpebral fissure to protect the cornea (Figure 5).

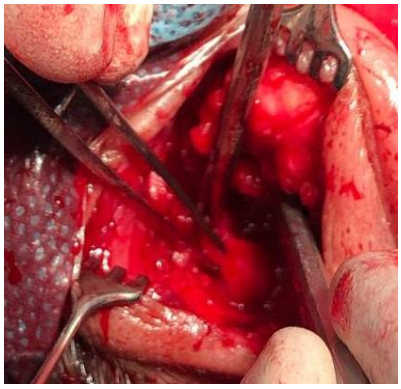

Figure 4. *The arrow points to the solid mass found in the patient's orbit during the surgery*

Figure 5. The condition of the eye after operation

The removed tissues were sent for HPE and the answer again excluded the malignant origin of the mass, but also excluded the diagnosis of pilomatrixoma. The specimen contained large amounts of dense fibrous tissue, giant foreign-body-type cells, and chronic lymphocytic infiltration giving the impression of sclerosing orbital pseudotumor. However, the lymphocytic infiltration was quite large in some areas, which raised the possibility of an alternative diagnosis such as low-grade lymphoma. To clarify the diagnosis immunohistochemical examination was performed, which confirmed the diagnosis of idiopathic sclerosing orbital inflammation.

After 1 month the swelling and the proptosis had decreased and the eyeball had a better position in the orbit, therefore the central tarsorrhaphy was removed to make the binocular vision possible (Figure 6).

Figure 6. *Patient's eye, after the removal of central tarsorrhaphy*

The condition of the cornea was better and the vision in the right eye was determined as object detection.

Discussion

Idiopathic sclerosing orbital inflammation also known as sclerosing orbital pseudotumor is a rare subtype of non-specific, idiopathic orbital inflammation that is poorly defined and is described in the literature in case reports or small series only.[3] It is a benign, space-occupying orbital inflammation

without local or systemic known cause, which both clinically and on orbital imaging mimics a malignant process or other orbital inflammatory disease, and therefore is difficult to diagnose and manage. The presentation may be unilateral or bilateral. It occurs in all age groups without gender predilection. The sclerosing type behaves differently than other types of pseudotumor of the orbit. It grows more slowly, does not cause ocular pain and has moderate response to steroid therapy. It can also rarely extend into the sinuses, brain, and the other orbit.

The pathogenesis is unclear, yet several lines of evidence suggest an autoimmune process, due to the response to immunosuppressive agents.[4]

The most common signs and symptoms observed are proptosis, restriction of extraocular muscle movements and visual impairment or loss of vision: when the mass has an apical location. Other clinical features include: palpebral edema, diplopia, chemosis, redness, palpable orbital mass, lagophthalmos, exposure keratopathy, tearing.[5]

The diagnosis is obtained by exclusion. For that, along with the clinical examination, CT-scan of the orbit, HPE and laboratory tests for differential diagnosis with other diseases (T3, T4, TSH, c-ANCA antibodies, ANA (antinuclear antibody), RF (rheumatoid factor)) are crucial.

Histopathological features are currently considered the gold standard for diagnosing sclerosing orbital pseudotumor. In cases where the histological findings raise the possibility of an alternative diagnosis, immunohistochemical examination is necessary.

Orbital CT-scan usually reveals focal or diffuse mass in the retrobulbar space, showing the involvement of muscles, the eyeball or the optic nerve in the inflammatory process. Orbital imaging is an essential tool, it allows to determine the localization of the mass.

HPE reveals a large amount of dense fibrous tissue associated with pleomorphic inflammatory cell infiltration. The main cell types found in specimens include lymphocytes, eosinophils and foreign body type giant cells. Lymphocytes were by far found in all specimens, and it has been suggested that they play a critical role in driving the processes that leads to fibrosis.

The main diagnoses that should be excluded are: Thyroid Eye Disease, Wegner's granulomatosis, sclerosing lymphoma, metastatic sclerosing tumors of breast.

All these other possible diseases can be excluded by the presence of high or low levels of characteristic serum markers, antibodies and hormones, and by typical features of X-rays and biopsies.

Compared with other types of inflammatory orbital pseudotumor, the sclerosing type is less responsive to steroid therapy. Most cases suggest combinations of surgery and steroid therapy.

Others found convincing results of using nonsteroidal immunosuppressive - agents (cyclophosphamide, azathioprine, methotrexate) with steroids. Azathioprine or other second-line agents can be used to reduce the use of corticosteroids.

Surgical debulking is crucial for patients with severely progressive and disabling clinical courses (apical mass with optic nerve compression) or when the lesion is focal and easily approachable.

Conclusion

Sclerosing orbital pseudotumor is an uncommon disorder that mimics malignant process or other orbital inflammatory disease and is difficult to diagnose and manage. No large studies exist because of the rare nature of the disease. Although the etiology remains unknown, an underlying immunological mechanism has been suggested. CT-scan of the orbit and HPE are required to confirm diagnosis. Steroids represent the first option of treatment. The sclerosing subtype shows moderate response to steroids, therefore combination therapy is suggested.

Declarations

Conflict of interest and financial disclosure

The author declares that he has no conflict of interest and there was no external source of funding for the present study. None of the authors have any relevant financial relationship(s) with a commercial interest.

Ethical approval

Research protocol was approved by the local Ethical Committee (2018/23) and in accordance with

those of the World Medical Association and the Helsinki Declaration.

Informed consent

Informed consent was obtained from all individual participants included in the study.

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Availability of Data and Materials

Not applicable.

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[2] Ս.Վ. Մալայանի անվան ակնաբուժական կենտրոնի տնօրեն, Երևան, Հայաստան
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Ամփոփում

Օրբիտայի իդիոպաթիկ սկլերոզացնող բորբոքումը (ISOI), որը նաև հայտնի է նաև որպես օրբիտայի սկլերոզացնող կեղծ ուռուցք, իրենից ներկայացնում է օրբիտայում հյուսվածքի սպիացումով ընթացող հազվագյուտ, իդիոպաթիկ, քրոնիկական, դանդաղ պրոգրեսիվող բորբոքում, ինչը կլինիկորեն նմանվում է նորագոյացության: Վերջինս հանգեցնում է ակնագնդի արտանկման, շարժումների սահմանափակման և տեսողության խանգարման՝ ընդհուպ մինչև տեսողության կորուստ: Այն օրբիտայի ոչ սպեցիֆիկ բորբոքային կեղծ ուռուցքի ենթատեսակ է, որը թե՛ ռենտգենաբանորեն և թե՛ կլինիկորեն նմանվում է օրբիտայի այլ բորբոքային հիվանդությունների կամ չարորակ պրոցեսների, և հետևաբար, դժվար է ախտորոշել և բուժել: Այս կլինիկական դեպքը ընդգծում է ISOI-ի ախտորոշման և բուժման խրթին կետերը:

ИДИОПАТИЧЕСКОЕ СКЛЕРОЗИРУЮЩЕЕ ВОСПАЛЕНИЕ ОРБИТЫ. КЛИНИЧЕСКИЙ СЛУЧАЙ

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Аннотация

Идиопатическое склерозирующее воспаление орбиты (ISOI), также известное как склерозирующий псевдотумор орбиты это редкое, идиопатическое, хроническое, медленно прогрессирующее воспаление орбиты, характеризующееся рубцеванием ткани, которое проявляется симптомами опухоли. Последнее приводит к выпучиванию глазного яблока, ограничению движений глаза и ухудшению зрения вплоть до потенциальной потери зрения. Это подтип неспецифического воспалительного псевдотумора орбиты, который как рентгенологически, так и клинически имитирует злокачественный процесс или другое воспалительное заболевание орбиты, и поэтому сложно диагностировать и лечить. Этот случай подчеркивает трудности диагностики и лечения ISOI.