

**Dacryoadenitis: Unusual Manifestation of Adult-Onset Still's Disease**

Ilyas El Kassimi (MD)\*, Nawal Sahel (MD), Oumama Jamal (MD), Adil Rkiouak (PhD), Salah-Eddine Hammi (PhD), Youssef Sekkach (PhD)

Department of Internal Medicine, Military Hospital Mohammed V, Rabat, Morocco

**\*Corresponding author**

Ilyas El Kassimi

**Article History**

Received: 23.02.2018

Accepted: 14.03.2018

Published: 30.03.2018

**DOI:**

10.36347/sjmc.2018.v06i03.009



**Abstract:** Adult-onset Still Disease (AOSD) is a rare systemic inflammatory disease. Ocular involvement is exceptionally associated with Still's disease. We present the case of a patient followed for AOSD who presented non-specific orbital inflammation involving the lacrimal gland. The diagnosis of dacryoadenitis was suspected after ruling out the other causes and the patient was treated with corticosteroids and methotrexate. Idiopathic orbital inflammation is rarely seen during AOSD. Recently, atypical ocular manifestations of AOSD have been reported. Biopsy indicated no signs of malignancy and ruled out sarcoidosis and vasculitis. Concerning infectious diseases, especially bacterial ones, a negative procalcitonin as well as the favorable response to corticosteroids allowed us to rule out their contingencies. This is an unusual presentation of an unusual disease.

**Keywords:** Adult-onset Still Disease, inflammatory disease, methotrexate.

**INTRODUCTION**

Adult-onset Still Disease (AOSD) is a rare systemic inflammatory disease of unknown cause that is typically characterized by hectic fever, pharyngeal pain, arthralgia or arthritis, and an evanescent rash. Ocular involvement is exceptionally associated with Still's disease. We describe here the case of a patient followed for AOSD who presented non-specific orbital inflammation involving the lacrimal gland.

**CASE PRESENTATION**

A 29-year-old woman (weight 66Kg) followed since one year for Still's Disease diagnosed on the basis of Yamaguchi's criteria, presented with left palpebral edema and periorbital erythema (fig.1). Clinical exam found erythematous throat and maculopapular rash in the left forearm and left knee. The joints were painful to mobilization. Pleuropulmonary and cardiovascular examinations were normal. There were no adenopathies or hepatosplenomegaly.

Ophthalmic exam showed ptosis, palpebral edema, S-shaped deformity of the lid without exophthalmos, and a tender infiltration of the upper-external region of the orbit at palpation. A slit-lamp examination showed temporal and episcleral conjunctival vasodilation. The neosynphrine test was positive and visual acuity was conserved. The examination of the fundus of the eye was normal.

An orbital CT scan was performed, showing a thickening of the left lacrimal gland of homogeneous density with palpebral edema (fig. 2). The histological evaluation of the lacrimal gland specimen revealed the presence of infiltration of the gland by inflammatory

cells, with edema and vascular dilatation evoking non-specific inflammation.

The assays results showed a leucocytosis 11100 elements/mm<sup>3</sup> (95% neutrophils), a high CRP (112.4mg / L). Ferritin levels were significantly elevated (19700µg/L). The level of rheumatoid factor, antinuclear antibodies, cytoplasmic and myeloperoxidase, anti-neutrophil cytoplasmic antibodies, liver enzymes, procalcitonin and the results of thyroid function were normal. The chest X-ray and phthysiological assessment were normal. Serology for syphilis, gonococcus and chlamydia trachomatis infection was negative. Viral screening including (mumps virus, Epstein-Barr virus, cytomegalovirus, coxsackie virus, echoviruses, and varicella-zoster virus) was negative.

The diagnosis of dacryoadenitis during AOSD was suspected and the patient was treated with 1g Methylpredisolone bolus per day for three days. Clinical symptomatology improved significantly, with disappearance of periorbital edema and remission of articular thrust. The patient was subsequently given methotrexate 15 mg / week and folic acid supplementation.



**Fig-1: The patient presented with left ptosis and erythema with edema in the upper-external eyelid**



**Fig-2: An orbital CT scan image showing a thickening with homogeneous density of the left lacrimal gland and palpebral edema**

## DISCUSSION

The discovery of an ocular manifestation such as idiopathic orbital inflammation is rarely seen during the evolution of AOSD. All structures in the orbit can be reached [1] recently; atypical ocular manifestations of AOSD have been reported and are of high diagnostic value [1-3]. To our knowledge; only one report has previously described dacryoadenitis in association with AOSD [1].

The absence of painful exophthalmos, decreased visual acuity and diplopia are against the tumoral origin of dacryoadenitis, in particular malignant one. In addition, the diagnostic biopsy results indicated no signs of malignancy. Biopsy was also very helpful in ruling out sarcoidosis and vasculitis, by the absence of granuloma, necrotic and leukocytoclastic vasculitis, and fibrotic degeneration.

Concerning infectious diseases, especially bacterial ones, a negative procalcitonin as well as the favorable response to corticosteroids alone allowed us to rule out their contingencies. Bacterial infections may be caused by *Staphylococcus aureus*, *Streptococcus pyogenes*, *Haemophilus influenzae*, *Neisseria gonorrhoea*, *Chlamydia trachomatis*, and *Treponema pallidum* [4-7]. Dacryoadenitis caused by tuberculosis or syphilis is usually manifested by periostitis or even osteolysis of the palpebral orbital bony rim, which was absent in our case [8].

In addition, some infections of the lacrimal gland, especially caused by bacilli and *Aspergillus*, or mucormycosis do not generally occur simultaneously in

acute onset, especially since the patient did not have immunosuppression [1]. Viruses such as influenza, mumps, mononucleosis, measles, herpes, and cytomegalovirus are not common causes in dacryoadenitis [5, 6]. Fungal and parasitic infections are rare [4].

The treatment of dacryoadenitis is etiological. In AOSD, it includes corticosteroids in addition to Conventional Disease Modifying Anti-Rheumatic Drugs (cDMARDs) when therapeutic response is not rapidly obtained or if there is a risk of corticoid dependence. Biotherapies such as anti-IL1, anti-IL6 and anti-TNF have proven their efficiency for the control of AOSD when corticosteroids and cDMARDs have been not effective [9].

## CONCLUSION

This observation brings to the literature a new case of dacryoadenitis associated with AOSD. AOSD should be included in the differential diagnosis of inflammation of the lacrimal gland.

## REFERENCES

1. Bannai E, Yamashita H, Takahashi Y, Tsuchiya H, Mimori A.. Two Cases of Adult-onset still's Disease with Orbital Inflammatory Lesions Originating from the Lacrimal Gland. *Intern Med.* 2015; 54(20): 2671-2674.
2. Cush JJ, Leibowitz IH, Friedman SA. Adult-onset still's disease and inflammatory orbital pseudotumor. *N Y State J Med.* 1985; 85: 110-111.
3. Yoon DL, Lee S, Park SY, Kim YJ, Koo BS, So MW et al. A Case of Adult-Onset Still's Disease Presenting with Periorbital Edema. *J Rheum Dis.* 2013; 20(4): 243-246.
4. Wald ER. Periorbital and Orbital Infections. *Infect Dis Clin North Am.* 2007; 21: 393-408.
5. Srivastava VK. Acute Suppurative Dacryoadenitis. *Med J Armed Forces India.* 2000; 56: 151-152.
6. Rai P, Shah SA, Kirshan H. Acute Dacryadenitis-analysis of 23 cases. *Med Channel.* 2009; 15; 71-76.
7. Hartikainen J, Lehtonen OP, Saari M. Bacteriology of lacrimal duct obstruction in adults. *Br J Ophthalmol.* 1997; 81: 37-40.
8. Van Assen S, Lutterman JA. Tuberculous dacryoadenitis: a rare manifestation of tuberculosis. *Neth J Med.* 2002; 60: 327-329.
9. Govoni Marcello, Bortoluzzi Alessandra, Rossi Daniela, Modena Vittorio, How I treat patients with Adult Onset Still's Disease in clinical practice. *Autoimmun Rev.* 2017; S1568-9972(17)30200-8.