Acute bilateral inflammatory dacryoadenitis as a presenting sign of Sjögren's syndrome

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INTRODUCTION

Acute dacryoadenitis is an atypical manifestation of Sjögren's syndrome although some cases have been reported[1]. Sjögren's syndrome is an autoimmune disorder causing chronic lacrimal and salivary gland destruction, resulting in severe dry eyes and mouth[2]. We present an uncommon case of Sjögren's syndrome presenting as acute bilateral dacryoadenitis.

CASE PRESENTATION

A 20 year old female presented to the emergency department with a 5 day history of progressive bilateral eyelid swelling associated with eye pain, fever, headache, nausea and vomiting.

On assessment, she had photophobia and VA of 6/12 in the left eye, 6/9 in the right eye and did not wear glasses for distance or near-vision. Eye movements were restricted and there was pain on leftward and upward gaze. Given her presentation, the initial concern was orbital cellulitis, an ophthalmological emergency. Therefore, the patient was started on 2g of IV ceftriaxone and 400mg of IV metronidazole. This was then switched to IV flucloxacillin by the medical team

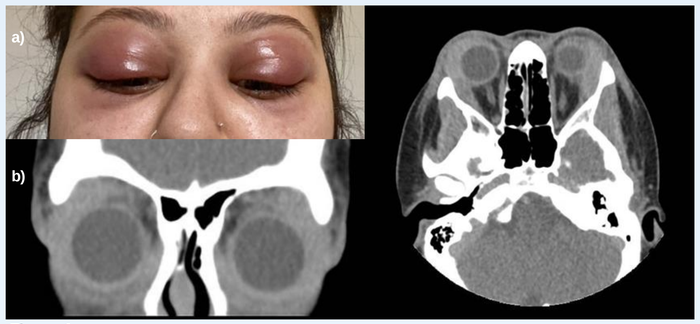

Figure 1.
a) Eyelid oedema and erythema on Day 1 of treatment
b) CT orbits showed bilateral proptosis and bilateral symmetrical enlarged lacrimal glands.

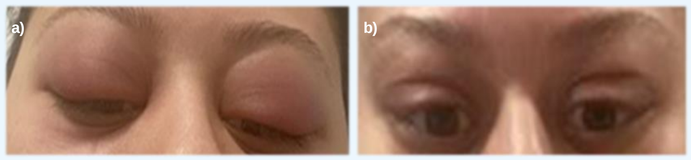

Figure 2.
a) Mild improvement of eyelid oedema on Day 4 of treatment
b) Further improvement by Day 7 of treatment

INITIAL INVESTIGATIONS

Blood tests revealed elevated inflammatory markers, with a CRP of 146.3 and a WCC of 16 (Table 1). A contrast-enhanced CT of the head and orbits was performed, which showed no evidence of post-septal cellulitis, abscess formation, or significant paranasal sinus disease. Blood cultures were also unremarkable. However, the CT did show bilateral proptosis and bilateral symmetrical lacrimal gland soft tissue masses in the orbits, suggestive of an infiltrative process. Differentials at this point included lymphoma, an orbital pseudotumour, and Sjögren's syndrome.

Test	Initial	Day 7
CRP (<10 mg/L)	**146.3**	**50.4**
WCC (3.6-11x10^9 g/L)	**16**	**12.3**
Hb (130-180 g/L)	135	**124**
Neut (1.8-7.5x10^9 g/L)	**12.16**	5.13
Lymph (1-4x10^9 g/L)	2.4	**6.06**
ALT (7-55 U/L)	**66**	30
Na (133-146mmol/L)	140	137
K (3.5-5.3mmol/L)	4.2	4.4

Table 1. Blood tests showed that inflammatory markers decreased from initial presentation to day 7

FURTHER INVESTIGATIONS

The unusual presentation and imaging report prompted investigation into a possible autoimmune or systemic cause. CT Thorax, Abdomen & Pelvis (with contrast) found no signs of occult primary tumour or metastasis. Blood films found lymphocytosis with no atypical lymphocytes.

Test	Initial
ANA (>60)	**100 speckled**
ENA	negative
Anti dsDNA	negative
ANCA	negative
Lupus anticoagulant (<1.2)	**1.37**
HLA B27	negative
C3 (0.9-1.8g/l)	**1.94**
ESR (3-9 mm/hr)	**87**
dRVVT (0.8-1.2)	**1.37**

Table 2. Rheumatology blood test results

However, autoimmune blood tests raised suspicion of Sjögren's syndrome or systemic lupus erythematosus (SLE) due to the raised ANA and lupus anticoagulant (Table 2). Due to suspicion of an autoimmune aetiology, the patient was commenced on prednisolone 30 mg OD on day 5. Sjögren's syndrome was supported by biopsy of the lacrimal gland, which showed chronic lymphocytic inflammation. Based on her clinical picture, she was commenced on hydroxychloroquine 200mg BD for a 6-month trial.

DISCUSSION

The presentation of dacryoadenitis can be quite variable. Broadly speaking, acute dacryoadenitis is related to infection, whereas sub-acute dacryoadenitis is related to inflammatory disease. In acute dacryoadenitis, symptoms are related to an abrupt onset of swelling of the lacrimal gland which causes the lateral portion of the eyelid to fall, eyelid swelling, chemosis and conjunctival injection[3]. In contrast, inflammatory causes of dacryoadenitis tend to present sub-acutely, typically with painless bilateral swelling of the lacrimal glands. In Sjögren's syndrome, dacryoadenitis normally presents sub-acutely alongside a range of other symptoms (Fig.3). However, this case highlights that Sjögren's syndrome can be considered in the range of differentials when a patient presents acutely with bilateral eyelid swelling.

CONCLUSION

This was an atypical presentation of Sjögren's syndrome, managed in a multidisciplinary manner with the emergency department, ophthalmology, acute medicine, and rheumatology teams. A short interview was conducted with the patient who is undergoing hydroxychloroquine therapy and her symptoms have resolved. No further episodes of inflammatory dacryoadenitis have been noted.

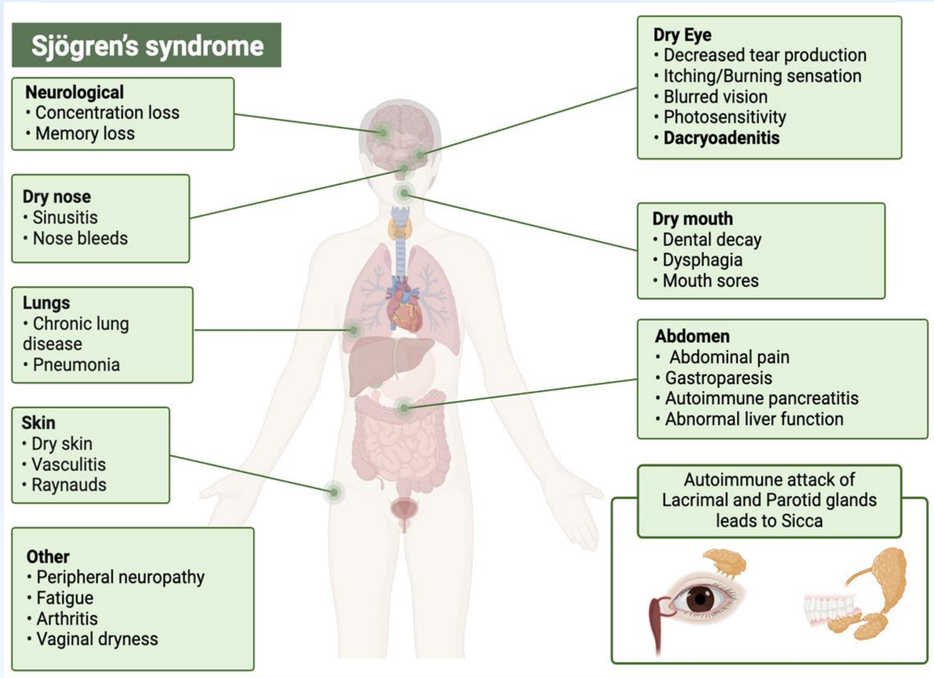

Figure 3. *Presentation of Sjögren's syndrome[1]. Made with Biorender©*

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