Brief Report

Fibrous histiocytoma as first presentation in systemic lupus erythematosus and sero-positive Sjögren's syndrome

Abdullah Al-Mujaini, Kashinatha Shenoy, Upender Wali, * 

a Sultan Qaboos University Hospital, Department of Ophthalmology, SQUH, Oman 
b Sultan Qaboos University Hospital, Muscat, Oman

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ABSTRACT

Purpose: To describe a triad of fibrous histiocytoma, Systemic Lupus Erythematosus and Sero-positive Sjögren's Syndrome.

Observations: This case was diagnosed first as bilateral fibrous histiocytoma of cheeks, which on further investigations proved to be a triad of Systemic Lupus Erythematosus, Fibrous histiocytoma and Sero-positive Sjögren's Syndrome.

Conclusions and importance: Association between fibrous histiocytoma, Systemic Lupus Erythematosus and Sero-positive Sjögren's Syndrome has been known before, but fibrous histiocytoma as first presentation in the triad has not been reported.

1. Introduction

Association between fibrous histiocytoma (FH), systemic lupus erythematosus (SLE) and seropositive Sjögren's syndrome has been known before, but FH as first presentation in the triad has not been reported so far. We report a case of a 42-year-old female who presented with bilaterally symmetrical firm, mobile swellings of both cheeks. Excisional biopsy on the left side proved to be FH upon histopathological examination. After two years, she was scheduled for excision on the right side. Pre-excision computed tomography (CT) scan showed microinfarcts and signs of vasculitis in the brain. Rheumatological evaluation proved to be SLE and strongly positive Sjögren's antibodies. Histopathology confirmed the diagnosis of FH. In the literature (source PubMed), FH presenting as first clinical feature in a patient positive for SLE and seropositive Sjögren's syndrome has not been reported.

2. Case report

A 42-year-old female presented with complaint of bilateral swelling on the cheek areas (zygomatic area) for 18 months. She had significant medical issues such as hypothalamic hypogonadism with hypopituitarism (congenital empty sella), diabetes insipidus and Hashimoto's hypothyroidism. On examination, her best-corrected visual acuity was 20/20 each eye. Slit lamp examination revealed unremarkable anterior and posterior segments. On palpation, she had bilaterally symmetrical swelling of cheeks on zygomatic area. The swellings were firm, mobile and non-tender. Scans revealed bilateral symmetric subcutaneous soft tissue masses overlying the zygomatic bones. The right mass measured 19 × 31 mm and left mass was 17 × 10 mm. Both lesions showed T1 and T2 low signal intensity and mild homogeneous enhancement after the administration of intravenous gadolinium. There were no associated bone marrow changes or signs of bone destruction. The adjacent muscles appeared normal. There was no extension into the orbit. The lacrimal glands, globes and retro-orbital fat appeared normal. Both optic nerves were unremarkable. The brain parenchyma had normal appearance. There was no evidence of abnormal signal, mass lesion or enhancement. The ventricles were normal in size and shape. The posterior fossa structures appeared normal as well. Radiological differential diagnosis included neurofibroma, Wegener's granulomatosis. A left side excisional biopsy of subcutaneous mass on the zygomatic bone area was done. Histopathology reported the specimen as a cellular lesion composed of sheets and clusters of foamy macrophages present on a background of abundant ropy collagen, with interspersed histiocytic and plasma cells showing a storiform pattern. No overt cellular atypia or increased mitotic activity was seen. The spindle and foam cells were positive for CD68, but negative for S100 (Fig. 3).

* Corresponding author.
E-mail addresses: mujainisqu@hotmail.com (A. Al-Mujaini), kashinath_sullia@yahoo.co.in (K. Shenoy), upender.wali@gmail.com (U. Wali).

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After the excision on the left side, the patient lost to follow-up (travelled abroad) and reported back to our clinic after nearly two years. The left side lesion had no recurrence. Prior to excisional biopsy (left side) with no recurrence through her last follow up, two years after excision. Recurrence is reported in up to 11% of cases.12 Usually such lesions do not benefit from radiotherapy and role of chemotherapy is not well known. The prognosis for the benign form is excellent and for the malignant lesions the mortality rate is more than 40%.13 Currently, the patient is on azathioprine and hydroxychloroquine, prescribed by her rheumatologist. Reviewing the literature, no report was found describing FH as the first case of FH presenting as initial manifestation in SLE and seropositive Sjögren's syndrome.

4. Conclusion

FH may present in unusual forms and in association with autoimmune disorders. Histopathology is very useful in establishing the diagnosis especially when there are comorbidities. To our best knowledge, this is the first case of FH presenting as initial manifestation in SLE and seropositive Sjögren's syndrome.

Patient consent

A written consent was obtained from the patient for the clinical images for production in publications and presentations for academic interest. This report does not contain any personal information that could lead to the identification of the patient.
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Conflicts of interest

None.

Authorship

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.ajoc.2018.03.017.

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