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Brief Report

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

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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. Intraocular pressure measured 12 each eye. Extraocular movements were full in all fields of gaze. 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 (Fig. 1). Our first impression was bilateral zygomatic dermoid.

Magnetic resonance imaging (MRI) reported bilateral symmetric subcutaneous soft tissue masses overlying the zygomatic bones. The right mass measured 19 × 31 mm and left mass was 17 × 10 mm (Fig. 2). 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).

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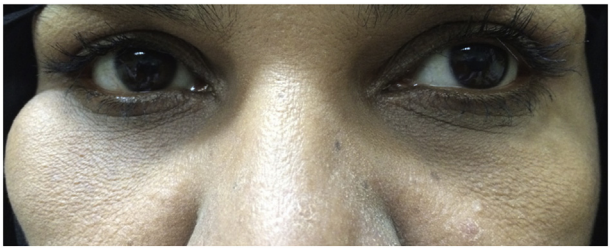

Fig. 1. Clinical photograph showing swelling over the right zygomatic area. The swelling was firm in consistency and non-tender. This was after the excision of a symmetrical mass on the left side, which was excised on her first presentation.

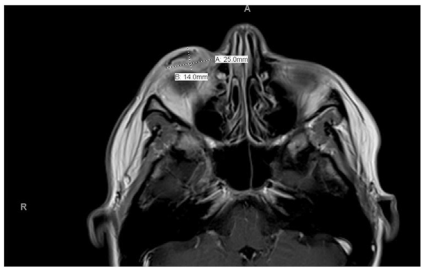

Fig. 2. MRI image reported as bilateral symmetric subcutaneous soft tissue masses with no signs of bone marrow changes, bone erosion or involvement of orbit or brain.

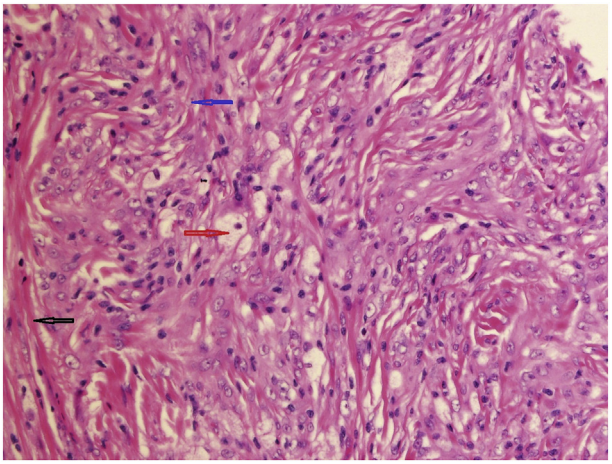

Fig. 3. Histopathology suggestive of foamy macrophages (red arrow), abundant ropy collagen (black arrow), myofibroblast (blue arrow), histiocytic and plasma cells showing a storiform pattern. No overt cellular atypia or increased mitotic activity was seen. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

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 on the right side, imaging for brain and orbit was done and showed no enhancement or bony, orbital or intracranial extension. However, there were multiple small subcortical white matter high-T2 signal lesions within the frontal lobes bilaterally.

These lesions were attributed possibly to vasculitis-associated micro infarcts, which prompted us to have rheumatological opinion to rule out any immune-mediated disorder. By now, she had cervical and hilar lymphadenopathy. After detailed investigations which included a battery of biochemical and microbiological tests, abnormal values were found on the following: antinuclear antibody (ANA), anti-Neutrophil cytoplasmic antibody, anti n-DNA antibody, anti- Sjogren's syndrome antigen A, and strongly positive Anti-R052. Rheumatoid factor was 603 IU/mL (Normal 0 − < 30). Based on clinical, laboratory and histopathological data, diagnosis of SLE was determined and she was given azathioprine, hydroxychloroquine and prednisolone.

3. Discussion

FH also known as fibrous xanthomas or dermatofibroma, is a benign or malignant mesenchymal tumor.[1] Its origin is related to fascia, muscle, or soft tissues.[2] It is the most common mesenchymal orbital tumor in adults, usually seen in middle-aged patients with mean age of 40 years.[3] The age and consistency of the mass in our patient matched with the description given in literature. Our patient had associated autoimmune disorders (SLE, vasculitis) and such association between FH and autoimmune disease is known.[4] In children, it may be a sequel to early orbital radiotherapy. It has a predilection to occur in the orbit, but may be found on the eyelid and cheek. The most common site of origin in the orbit is the upper nasal quadrant.[5] However, our patient had an unusual presentation on cheeks involving both zygomatic areas though there was no involvement of the bone itself. Symptoms include exophthalmos, vision disturbance, diplopia, ptosis, motility restriction, epiphora or just a swelling with cosmetic worry, as in our patient. The benign form (63%) is a well circumscribed, slow growing lesion with small potential for malignant degeneration, while the malignant form (37%) is more infiltrative, rapidly growing lesion and often associated with pain.[6]

The association of FH with SLE and Sjögren's syndrome is rare and found in the literature as case reports only.[7] Triad of FH, SLE and Sjögren's syndrome has been reported by Fujisawa and Seishima, and Tsunemi Y et al. However, none of their patients had FH as first presentation, and probably FH developed due to long-term immunosuppressives for SLE.[8,9] Our patients showed markers of SLE after diagnosis of FH and was not on any immunosupressives before diagnosis of SLE.

Treatment options in FH follow the SEER database and include surgical excision and radiotherapy.[10,11] Our patient had 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 or initial presentation in a case that subsequently proved to be 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

All authors attest to meet the current ICMJE criteria for authorship. Full adherence to the declaration of Helsinki laws.

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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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