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IMAGES IN RHEUMATOLOGY

Bilateral parotid gland swelling unveiling non-Hodgkin lymphoma

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A 62-year-old woman presented to the rheumatology clinic with bilateral parotid gland swelling and sicca syndrome (xerostomia and xerophthalmia), unquantified weight loss, and fatigue of one month's duration. Her past medical history included stage IV nodal marginal zone non-Hodgkin lymphoma, diagnosed eight and a half years ago based on a cervical lymph node biopsy, treated with six cycles of rituximab and bendamustine, with sustained remission for the past eight years. She was diagnosed with sarcoidosis seven years ago, based on bilateral recurrent anterior uveitis (last episode three years ago), hilar lymphadenopathy with pulmonary infiltrates, and non-caseating granulomas on bone marrow biopsy, for which she has been treated with azathioprine 75 mg/day and prednisolone 5 mg/day. The patient had also triple positivity for antiphospholipid antibodies but no clinical history of recurrent (venous or arterial) thromboses or adverse pregnancy outcomes. On physical examination, parotid gland swelling was evident (Figure 1, panels A and B). Neck computed tomography confirmed enlargement and heterogeneity of the salivary glands, as well as cervical lymphadenopathy (Figure 1, panels C and D). Salivary gland ultrasound showed consistent findings, with hypoechoic foci and marked Doppler signal in all parot-

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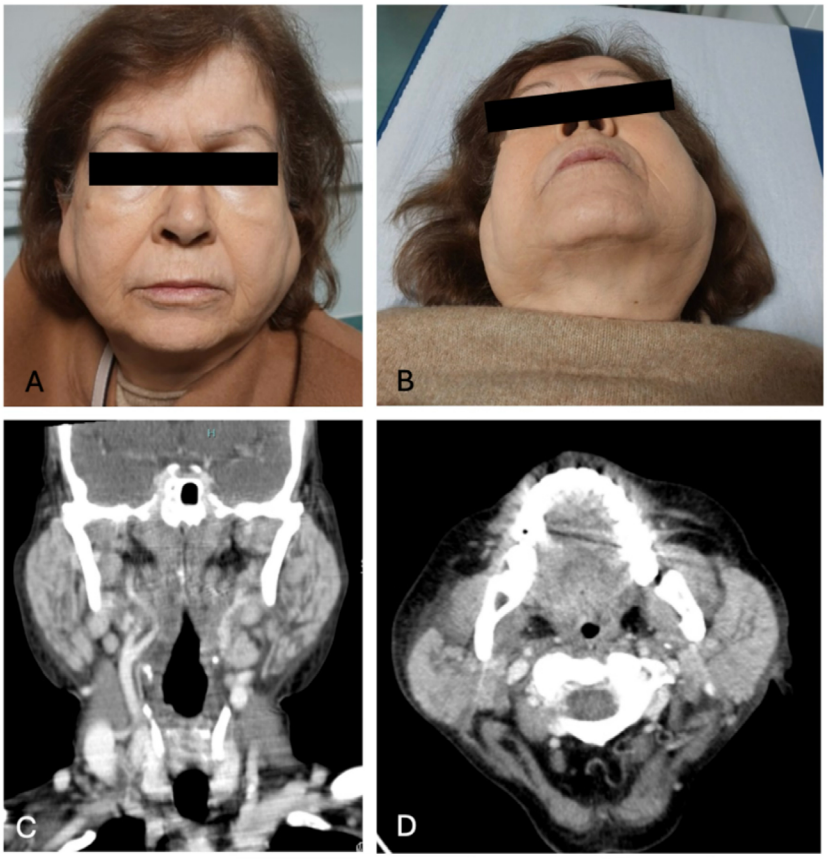

Figure 1. Physical exam revealing firm, enlarged parotids (panels A and B). Neck CT showing enlargement and heterogeneity of the salivary glands with cervical lymphadenopathy (panels C and D).

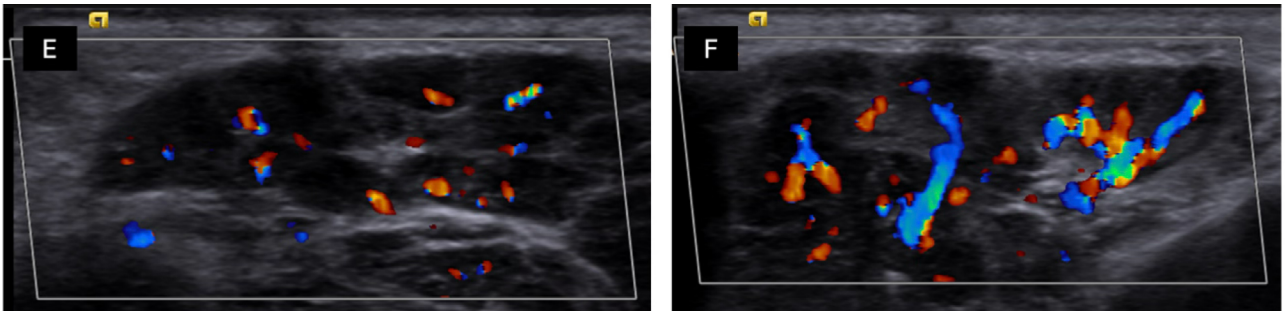

Figure 2. Parotid gland ultrasound showing hypoechoic foci and marked Doppler signal (panels E and F).

id and submandibular glands (Figure 2, panels E and F). Laboratory evaluation revealed severe neutropenia ($0.2x10^9$/L), undetectable C4 complement, decreased C3 complement, elevated angiotensin-converting enzyme (136 U/L), increased β2-microglobulin (11.20 mg/L), and positive anti-dsDNA antibodies (133 UI/mL). Antinuclear antibodies, anti-SSA and anti-SSB antibodies were negative. IgG4 was within normal range. Core biopsy of the parotid gland and submandibular lymph node was performed revealing a high-grade B-cell lymphoma, likely diffuse large B-cell lymphoma, although it could not be determined whether this represented a relapse of the previous marginal zone lymphoma or a new primary lymphoma. Unfortunately, the patient died from septic shock approximately two months after the lymphoma diagnosis, before initiating therapy directed at the lymphoma.

This case highlights that bilateral parotid gland swelling, even in the context of sicca syndrome and pre-existing conditions such as sarcoidosis, may represent a manifestation of underlying hematologic malignancy. It underscores the importance of prompt diagnostic evaluation, including imaging and tissue biopsy, in patients with atypical presentations or aberrant laboratory findings, as early identification is crucial for timely management.