

## **ATYPICAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: PAROTITIS AND SECONDARY SJOGREN'S SYNDROME CASE REPORT**

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**ATYPICAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: PAROTITIS AND SECONDARY SJOGREN'S SYNDROME (A CASE REPORT)** (Abstract): Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by auto antibodies directed against self-antigens, immune complex formation and immune deregulations and may affect joints, skin, kidneys, heart, lungs, nervous system, and immune system. The onset can be variable and the symptoms can occur for many years. Parotitis as the initial manifestation of systemic lupus erythematosus (SLE) is a rare condition and can be associated with Sjogren's syndrome. In this article we present the case of a young patient who was diagnosed with Sjogren's syndrome retrospectively, after she met the criteria for SLE. **Key-words:** SYSTEMIC LUPUS ERYTHEMATOSUS, PAROTITIS, SJOGREN'S SYNDROME.

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by auto antibodies directed against self-antigens, immune complex formation and immune deregulations and may affect joints, skin, kidneys, heart, lungs, nervous system, and immune system (1). The onset can be variable and the symptoms can occur for many years (2). Parotitis as the initial manifestation of systemic lupus erythematosus (SLE) (3) is a rare condition (4) and can be associated with Sjogren's syndrome (5).

The Systemic Lupus International Collaborating Clinics group revised and validated the ACR SLE classification criteria in 2012 (6). According to the revision, a

person is defined as having SLE if any 4 or more of the 11 criteria are present, respectively or simultaneously, during any interval of observation. Parotitis associated with Sjogren's syndrome is a rare condition in childhood (7, 8).

Sjogren's syndrome may be a primary disorder or may be secondary to other autoimmune disorders, such as systemic lupus erythematosus (9). Thus, it is required to be excluded a number of underlying causes of that disease (most of them of infectious origin). The natural history of SLE is unpredictable (10). Two of the most common complications are lupus nephritis (11) and cerebral damage.

We present the case of a young patient

with SLE in which the initial manifestation was parotitis and Sjogren's syndrome.

### CASE PRESENTATION

We report the case of a 10-year old girl who presented with bilateral parotid swelling, symmetrical, non-traumatic and non-thrombocytopenic purpura on the lower limbs, accentuated by physical effort and orthostatic position, without fever (fig.1). Her symptoms have progressed gradually over the past 12 months.

The patient's past medical history is only remarkable for parotitis diagnostic, one year ago. She also accused intermittent arthritis, ocular manifestations consisting of itching and pain, recurrent epistaxis and photosensitivity. She has no other family history of medical illnesses, particularly heart disease or connective-tissue disease.



**Fig. 1.** Bilateral parotid swelling

The patient's physical examination reveals a normal nutritional state (BMI=21 kg/m<sup>2</sup>), "butterfly rash", bilateral swelling parotid, gingivostomatitis, an arterial blood pressure of 105/65 mmHg, a heart rate of 72 beats per minute, a respiratory frequency of 20 per minute, symmetrical purpura on the lower limbs; clinical examination did not reveal any tenderness of the abdo-

men, no hepatomegaly or splenomegaly and no arthritis.

Traumatic and toxic causes have been excluded, clinically and anamnestically. Multiple bacterial and viral infectious agents have been associated with the development of parotitis. We therefore determined serologies for HIV 1 and 2, cytomegalovirus (CMV) and Epstein-Barr (EBV) viruses' infection and IgG fractions were positive for CMV and EBV. Mumps was excluded anamnestically (disease was present in her medical history) and the clinical examination did not correspond to a condition manifested by acute symptoms such fever, shivering, myalgia, pain and local skin changes. Tuberculosis was excluded by negative history for TB contact, negative tuberculin intradermal reaction (IDR) and normal chest X-ray. Other bacterial infections were excluded. Mikulicz syndrome was also ruled out by clinical appearance of the parotid glands, absence of both lacrymal and submaxillary glands' swelling and lack of other syndromes (chronic lymphocytic leukemia, sarcoidosis). Maxillofacial consultation excluded sialolithiasis and reported gingivostomatitis, tooth cavities and dry mouth (fig. 2).



**Fig. 2.** Gingivostomatitis

## Atypical presentation of systemic lupus erythematosus: parotitis and secondary Sjogren's syndrome (a case report)

Ophthalmological examination has found xerophthalmia and dry eyes. The diagnostic of Sjogren's syndrome could not be confirmed by biopsy.

Laboratory tests showed other pathological associated issues which pleaded for an autoimmune disease: an important inflammatory syndrome (erythrocyte sedimentation rate - ESR of 131 mm per hour), C-reactive protein (CRP) was positive in all determinations, a very high value of total serum proteins (122 g/l), hypergammaglobulinemia (51.7%), mild normochromic, normocytic anemia (haemoglobin - Hb of 9 g/dL), plasmocytic expansion on bone marrow biopsy. Bence-Jones protein was not determined, but multiple myeloma is usually met in adults over 40 years. Other serologies like p-ANCA and c-ANCA antibodies were negative. Liver function tests revealed mild hepatocytolysis (ASAT 45 U/L, ALAT 63 U/L), kidney function (urea, creatinine, clearance of creatinine) was normal. The abdominal ultrasound examination revealed no pathological findings.

Our case meets 5 of the 11 required ACR criteria for SLE: "butterfly rash", arthritis, photosensitivity, immunological changes such as anti-nuclear antibody titer (ANA) over 32 (positive titer over 1), anti-deoxyribonucleic acid antibodies (dsDNA) positive titers. These findings are classical for SLE. We believe those findings to be consistent with lupus associated with Sjogren's syndrome.

We also excluded other viral infections (hepatitis B, C) and we evaluated the status of lupus-target organs (heart – the cardiac ultrasound exam was normally, kidney – function and ultrasound exam) for initiating the therapy with corticosteroids. Cortisone therapy included methylprednisolone pulses, three consecutive administrations (30

mg/kg/day), following the oral medication.

Clinical and paraclinical evolution was slowly favorable with remission of arthralgia, decreased parotid swelling, improvement of biological inflammatory syndrome (ESR decreased at 70 mm/h), anemia (Hb of 11.4 g/dl). But a number of adverse effects and complications of immunosuppression were present: over-weight (8 kg), stretch marks, a prolonged evolution of gingivostomatitis, despite local treatment and oral hygiene, which could lead to a series of infectious complications such maxillary sinusitis, chronic rhinitis, conjunctivitis and urinary infections. She had no pancreatic involvement during evolution, the pancreatic ultrasound and levels of pancreatic enzymes were normal.

After two years of evolution the patient showed renal impairment: persistent hematuria and proteinuria, low levels of C<sub>3</sub> fraction of complement. It required renal biopsy for staging of lupus nephritis and so she was transferred to the Nephrology Clinic.

### DISCUSSION

Primary Sjogren's syndrome occurs in the absence of another underlying rheumatic or immune disorder, while secondary Sjogren's can be associated with other diseases, such as systemic lupus erythematosus (SLE), scleroderma, rheumatoid arthritis (RA).

Thus, there is an overlap of Sjogren's syndrome with many other immune mediated diseases and it is sometimes difficult to determine whether a clinical manifestation is a primary consequence of Sjogren's syndrome or is due to one of its overlapping disorders.

Almost half of patients with Sjogren's syndrome also present with cutaneous find-

ings, such as dry skin (xeroderma), palpable and nonpalpable purpura, and/or urticaria (12).

Primary disease is a rare condition in childhood; only 145 cases of primary disease have been described in the international pediatric literature (13, 14). The majority of patients with Sjogren's are in conjunction with other autoimmune diseases, which are considered secondary to Sjogren's syndrome.

Individuals with secondary Sjogren's syndrome may have systemic lupus erythematosus, scleroderma, and primary biliary cirrhosis; therefore it takes a whole battery of tests and investigations for these

patients to exclude other disorders.

Also, long-term, Sjogren's syndrome can lead to complications that affect teeth, kidneys, eyes, lungs and joints and even to lymphoma (15). Another rare manifestation in lupus is cerebral vasculitis.

In the literature there have been documented only few cases of atypical onset of Sjogren's syndrome (16).

In two of these cases the age of diagnosis was 10 years, similar to the age of our case.

In our experience and evidence with 40 cases of systemic lupus erythematosus this is the only case with associated Sjogren's syndrome.

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### NEWS

#### AMNIOTIC MEMBRANE AND FINGERTIP INJURIES

A group of researchers from Department of Orthopaedic Surgery, Shiraz University of Medical Sciences conducted a study that evaluated the efficacy of amniotic membranes (AM) as a biologic wound dressing material for coverage of fingertip injuries, which are the most common hand injury, represent management challenges for hand surgeons. The study included 30 patients with full-thickness zone 1 fingertip skin loss. The patients were divided into 2 groups using the block randomisation method. In the first group, a skin graft was used for coverage and in the second group, AM was used. All patients were operated on by the same hand surgeon between February 2012 to October 2012. The minimum follow-up period was 6 months. Two point discrimination (T.P.D), light touch, healing time, days lost from work and infection rate were evaluated and recorded. Fingertips in both groups were assessed. T.P.D, light touch and days lost from work were significantly lower in the AM group than in the skin graft group. Healing time was lower in the skin graft group. In the both groups, no infection was detected. Patients of both groups were satisfied of their treatment and healing progress. The authors showed the effectiveness and safety of AM for the treatment of fingertip amputation, which can produce better sensation and functional outcomes than skin graft transplantations. (Fijan A1, Hashemi A2, Namazi H. A novel use of amniotic membrane for fingertip injuries. *J Wound Care*. 2014 May;23(5):255-8. PMID: 24810309).

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