Acute salivary gland inflammation associated with systemic lupus erythematosus

W. A. KATZ and G. E. EHRLICH

The Parotid Gland of Tumulty, Conley, who had systemic sialoadenitis characterized by lupus association while inflammation of Sjogren-Mikulicz syndrome, bilaterally in lupus systemic Arthritis Center, none of the parotid swelling a subacute reported SLE of Sjogren's syndrome (1 ref. reflected in which is weakness, Albert Einstein Medical Center, text, including submaxillary, lymph nodes, and/or lacrimal glands. Although none of these cases was acute, one, a 19-year-old Negro girl, was admitted because of parotid swelling of only 4 weeks' duration associated with a flare of her generalized disease. All other authors (Futcher, 1959; Bain, 1960) have likewise reported subacute or chronic salivary gland enlargement not paralleling systemic disease activity. As a matter of fact, Heaton (1959) concluded that Sjogren's syndrome is a chronic, relatively benign form of systemic lupus erythematosus. We report three cases in which acute inflammation of the salivary glands reflected lupus activity.

Case reports

Case 1, a 15-year-old Black girl, was first admitted to the Albert Einstein Medical Center on February 19, 1969, with complaints of multiple joint pain, morning stiffness, generalized weakness, and fatigue for 2 years. She had developed fever accompanied by pain and swelling at the angle of the jaws, and a diagnosis of 'mumps' was made by her physician, even though there had been no known exposure to infection. The symptoms disappeared within a few days; however, the patient subsequently became aware of exertional dyspnoea and pruritic papular eruptions on the flexor surfaces of the extremities and face. Physical examination showed multiple pigmented macular areas covering the arms, legs, and malar region. There was clinical evidence of pericardial and pleural effusions, and tenderness was present in the right and left costovertebral angles. The liver, spleen, and kidneys were not palpable. The salivary glands were not enlarged or tender. There was a synovitis of both wrists.

Chest x rays confirmed the existence of a small left pleural effusion with moderate globular cardiomegaly. During the ensuing 3 weeks when she was receiving full dose salicylates and rest in bed the joint pain gradually disappeared. She became less dyspnoeic, and the heart became progressively smaller.

After her discharge she remained well until 1 week before her second admission to hospital on April 9, 1969, when she developed a marked swelling in the submandibular region, which progressively enlarged to the angle of the jaw. This was associated with anorexia and progressive generalized weakness.

Examination

She was acutely ill, complaining of severe pain in the jaw. Temperature was 105°F. There was a circumscribed swelling of the entire left side of the face and the left submandibular area, which was hot, red, and tender (Fig. 1, opposite). The submandibular nodes and anterior cervical nodes on the left were enlarged and tender. The heart again was found to be enlarged, and a pleural effusion was noted at the left base. No evidence of arthritis was present.

Mandibular x rays showed considerable soft tissue swelling of the mandible and neck, but no calculi or osseous abnormalities were discovered. A sialogram revealed tertiary and quaternary intraductal ectasia, a picture most commonly seen in Sjogren's syndrome.

Laboratory investigations

Repeated culturing of the salivary gland ducts and blood produced no micro-organisms. Haemoglobin was 7.5 g. per cent.; haematocrit, 24; white cell count, 7,600 with
Progress

However, despite this therapy, she continued to have fevers as high as 104°F. for 3 days. Stenson's duct was oedematous and an attempt to cannulate it was unsuccessful. Small amounts of bloody fluid aspirated from the submandibular mass were sterile on culture. On the 4th day in hospital, methylprednisolone was started in daily doses of 64 mg.; defervescence of her symptoms and fever resulted within 8 hours. From that time on, she remained afebrile, and within 18 days the parotid gland shrank to normal size.

Biopsy of the submaxillary glands on the 14th day in hospital disclosed atrophy of the salivary glands and distended ducts. There was a background of stromal infiltration of mononuclear inflammatory cells. Some of the ducts contained loose inspissated material. The appearance was thought to be consistent with ranula and chronic sialoadenitis involving minor salivary glands (Figs 2 and 3, opposite).

Biopsy of the submandibular gland on the 22nd day in hospital yielded necrotic glandular tissue with intense acute and chronic inflammation.

Termination

2½ years later, the patient died from a severe lobar pneumonia. The salivary glands were not clinically involved at the time of her death. The submaxillary glands at autopsy showed a number of chronic inflammatory cells, mostly lymphocytes, around some of the ducts. Otherwise, the gland was unremarkable. The pancreas showed no abnormality.

Case 2, a 54-year-old white woman, developed pain, swelling, and redness of the right wrist, proximal interphalangeal joints of the left hand, and left knee some years before her admission to hospital. The joint pain recurred intermittently and was often associated with episodes of alopecia and transient ulcerations of the soft and hard palate. 8 months before admission, an erythematous malar eruption was discovered. Chloroquine, 250 mg. twice daily, and prednisone, 30 mg. daily, caused a prompt amelioration of her symptoms. During the 2 months preceding admission, the dose of prednisone was gradually reduced to 15 mg. a day. 2 days after the dose was further lowered to 12.5 mg. daily, the patient again noted joint pain and swelling. This time it was associated with acute pain and swelling at both angles of the jaw. There was no known exposure to mumps.

Examination

The patient did not appear acutely ill. The parotid glands were found to be enlarged, hardened, and swollen. The overlying skin was reddened. Marked tenderness was elicited bilaterally. There was obvious difficulty in opening the mouth. She was afebrile. There was slight pain on motion and soft tissue swelling of the left wrist. The remainder of the examination was within normal limits.

Laboratory investigations

Haemoglobin 12.5 g.; haematocrit 36 per cent.; white cell count, 4,500 with 60 per cent. segmented neutrophils, 35 per cent. lymphocytes, and 5 per cent. monocytes. The LE preparation was positive and antinuclear antibodies at a 10:1 dilution were demonstrable using an immunofluorescent technique with mouse liver as nuclear substrate. The erythrocyte sedimentation rate was 45 mm./1st hour (Westergren).

Treatment

The dose of prednisone was increased to 25 mg. a day.

Result

Within 24 hours, the patient was totally asymptomatic, and within 48 hours the parotid swelling completely disappeared.
Case 3, a 25-year-old Black woman, was admitted to the Albert Einstein Medical Center on December 1, 1968, with a 4-month history of slowly progressive polyarthralgia involving primarily the elbows, wrists, and knees, associated with morning stiffness lasting 3 to 4 hours. In addition, she noted increasing fatigue and generalized weakness. Only slight relief was obtained from aspirin. She could recall no facial rash, mouth ulcers, alopecia, or pleuritic chest pain. There was no known exposure to mumps.

Examination
Temperature 101.8°F. There was noticeable periorbital oedema and moderate swelling and tenderness over both parotid glands. The thyroid gland was found to be diffusely enlarged. There was synovitis of the second and third metacarpophalangeal joints and all the interphalangeal joints of the right hand. Other observations were within normal limits.

Laboratory investigations
Urine analysis showed numerous red blood cells and a few white cells but no casts. Haemoglobin 9.1 g. per cent.; haematocrit, 29 per cent.; white cell count, 2,350, with 13 per cent. polymorphonuclears, 77 per cent. lymphocytes, 4 per cent. eosinophils, 2 per cent. monocytes, and two atypical lymphocytes. The erythrocyte sedimentation rate was 100 mm/1st hr (Westergren). Three LE preparations were positive (LE preparations done 3 weeks previously had been negative). Platelet count 204,000; reticulocyte count 1.2 per cent. Antinuclear antibodies at a 10:1 dilution were demonstrable using an immunofluorescent technique with mouse liver as nuclear substrate. Anti-DNA borderline at 11.5 per cent. Serum complement 21.2 CH50 u per ml., C3 complement component 74 mg. per cent. (normal 100–190). Serum electrophoresis disclosed diminished albumin and an increased broad-based polyclonal gamma globulin. Serum uric acid 8.6 mg. per cent. Glucose, BUN, creatinine, bilirubin, prothrombin time, SGPT, LDH, serum iron were all within normal limits. Latex-fixation test negative; thyroid globulin antibody tests weakly positive. Coombs’s test negative. Bone marrow examination normal. Renal function studies showed inulin clearance diminished to 27 ml./min. and PAH clearance diminished to 415 ml./min. This was felt to be consistent with an active glomerulopathy. A renal biopsy disclosed local cellularity and degeneration consistent with early lupus erythematosus.

Treatment and Result
Methylprednisolone, 40 mg. per day, resulted in marked amelioration of all symptoms after 3 days. The parotid glands reverted to normal size and were non-tender.

Discussion
Systemic lupus erythematosus is usually classified as a specific disease, while Sjögren’s syndrome has been
accepted as a conjunction of symptoms, signs, and laboratory abnormalities accompanying several con-
nnective tissue disorders. Enlargement of the salivary
glands has been well recognized in systemic lupus
erythematosus but, except one case reported by
Dubois (1966), all were chronic without clinical signs
of inflammation and did not noticeably wax and wane
with fluctuation of lupus activity. These enlargements
have been thought to produce Sjögren’s syndrome,
pathologically characterized by lymphocytic infil-
tration replacing acinar tissue, reduction in acini, and
the presence of myoepithelial islands with eventual
fibrosis and glandular ectasia. Although these
chronic manifestations of Sjögren’s syndrome are
well known, there is little understanding of its
development. The three cases we have described are,
therefore, of interest as possibly representing such an
acute stage of Sjögren’s syndrome.

Another explanation is that, in our cases, the
salivary gland inflammation represents actual lupus
involvement. The rapid onset of inflammation of the
salivary glands occurring concomitantly with lupus
activity in other organs and the dramatic response to
corticosteroids in all cases lends credence to this
hypothesis. Because the salivary glands are not
routinely biopsied, there are no pathological studies
of the acutely inflamed salivary gland in systemic
lupus erythematosus; but autopsy studies suggest
that the pancreas, ‘sister organ’ to the parotid gland,
is often abnormal (Harvey and others, 1954). Most of
the time, an arteritis is found, leading to chronic
changes including atrophy and ectasia.

In our cases, the salivary glands enlarged concomi-
tantly with exacerbations of systemic lupus erythema-
tosus and reverted to normal size during drug-induced
remissions of the disease. In the first case, the
submaxillary gland was the site of predominant
involvement; in the other two, the parotids. Clinically
active inflammation was characterized by pain,
swelling, tenderness, and warmth (and even redness
in the first two cases) (Fig. 1). Although no attempt
was made to rule out infection or obstruction in the
second and third cases, the bilateral involvement
and rapid response to corticosteroids alone in the
absence of antibiotic therapy makes the possibility of
bacterial sialoadenitis remote. None of the patients
was exposed to mumps. Though virus studies were
not performed to rule out epidemic parotitis, this
diagnosis was not considered likely on clinical
grounds. Negative cultures of the blood, glandular
aspirates, and ductal secretions in the first case helped
to eliminate an infective basis. Treatment with anti-
biotics failed to affect the sialoadenitis favourably,
providing further evidence against infectious aeti-
ology. Cannulation of the duct and sialogram failed
to yield any evidence of obstruction. Certain drugs,
such as potassium iodide, phenylbutazone, guaneth-
dine, and methimizole, have been known to induce
parotid swelling, but none of the patients was receiv-
ing any of these medicaments. In one patient, the
parotid gland enlarged during a period of cortico-
steroid reduction. Biopsy of the submaxillary gland in
the first case showed only non-specific inflammatory
changes with no suppuration. It seems likely that the
acute sialoadenitis may be a manifestation of acute
systemic lupus erythematosus, possibly representing
the acute stage of Sjögren’s syndrome.

Summary

Three patients are presented in whom acute sialo-
adenitis accompanied flaring or active systemic lupus
erythematosus. It is possible that these may represent
the early stages of what would later be diagnosed as
Sjögren’s syndrome, the relationship of which with
systemic lupus erythematosus has been well estab-
lished.

References

lupus erythematosus)


associated with systemic disease)

(Baltimore), 33, 291 (Systemic lupus erythematosus: Review of the literature and clinical analysis of 138 cases)


relation to Sjögren’s syndrome)

DNA-binding activity in systemic lupus erythematosus)

SHEARN, M. A., AND PIROFSKY, B. (1952) Arch intern. med., 90, 790 (Disseminated lupus erythematosus—Analysis
of 34 cases)
Acute salivary gland inflammation associated with systemic lupus erythematosus.
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doi: 10.1136/ard.31.5.384

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DAVID PREISKEL


The title and indeed the introduction to this book encourage the reader to expect a practical approach to the identification of pain felt in the arm. In the sense that five contributors and the editor between them outline the main causes of arm pain, the reader will not be disappointed. However, there is little sign of editorial effort to relate the many causes listed to clinical reality. Thus the shoulder (gleno-humeral) joint is dismissed with the brief statements that it is uniquely free from osteoarthritis and degenerative conditions and that a frozen shoulder may be found with cervical radiculopathy, while peripheral neuropathy as a cause of arm pain commands several pages including a classification of some 35 causes. Even myasthenia gravis is mentioned as a cause of arm pain!

The editor’s interest in posturally-induced pain is apparent from the inclusion of a whole chapter on scapular traction and related syndromes and of one on their treatment by a combination of shoulder shrugging exercises, cervical posturing, and neck flexor exercises. The treatment of the common causes of arm pain receives scant mention.

None the less, the different approaches of an orthopaedic surgeon, a neurologist, a neurosurgeon, a vascular surgeon, and an internist to various aspects of arm pain are well illustrated by the first five chapters of this book, and these in combination with the editor’s chapters on median, ulnar, and radial pressure neuropathies, including consideration of relevant electromyographic techniques, are worth study by rheumatologists to whom arm pain is a common problem and sometimes a diagnostic trap.

A. T. RICHARDSON

Notes

XIII International Congress of Rheumatology

Kyoto, Japan, September 30 to October 6, 1973

The XIII International Congress of Rheumatology, sponsored by the Ligue Internationale contre le Rhumatisme and organized by the Japan Rheumatism Association and Foundation, will be held at Kyoto, Japan, on September 30 to October 6, 1973, under the presidency of Prof. Y. Oshima. Meetings of the Pan-American, European, and South-east Asia and Pacific Leagues will be held on September 30. The second International Geigy Rheumatism Prize will be awarded at the final session on October 6. An extensive programme of lectures, discussions, and symposia has been arranged.

Further information may be obtained from the Secretariat, Japan Convention Services Inc., 3-23 7-chome, Roppongi, Minato-ku, Tokyo, 106, Japan.

Particulars of travel arrangements from Europe, including various tours of the Far East, may be obtained from H. Stulz, Bolte postale 149, CH 4010, Basel, Switzerland.

VI Pan-American Congress on Rheumatic Diseases

Toronto, Canada, June 16–21, 1974

A preliminary notice of this congress, to be held under the presidency of Prof. M. A. Ogryzlo, announces that the last date for submission of abstracts for inclusion in the scientific programme is January 15, 1974. Communications should be addressed to: The Congress Secretariat, 45, Charles Street East, Toronto 285, Ontario, Canada.

Corrigendum


On page 387, col. 1, l. 9, for ‘thought to produce' please read ‘thought to be produced by Sjögren’s syndrome’.