Sjögren's Syndrome In Childhood: Report of a Case

Howard Duncan
Bruce N. Epker
Gerald M. Sheldon

Follow this and additional works at: https://scholarlycommons.henryford.com/hfhmedjournal
Part of the Life Sciences Commons, Medical Specialties Commons, and the Public Health Commons

Recommended Citation
Available at: https://scholarlycommons.henryford.com/hfhmedjournal/vol17/iss1/6

This Article is brought to you for free and open access by Henry Ford Health System Scholarly Commons. It has been accepted for inclusion in Henry Ford Hospital Medical Journal by an authorized editor of Henry Ford Health System Scholarly Commons.
Sjögren's Syndrome in Childhood: Report of a Case

Howard Duncan, M.D.,* Bruce N. Epker, D.D.S.,** and Gerald M. Sheldon, M.D.***

This report documents a rare case of Sjögren's Syndrome which began in a child with juvenile rheumatoid arthritis at the age 10 years, followed by the symptoms of xerostomia with intermittent salivary gland swelling 15 months later. By age 12 the patient had developed keratoconjunctivitis sicca. Variations in the intensity of each component of Sjögren's syndrome were noted to be independent of each other.

The syndrome of keratoconjunctivitis sicca, xerostomia, and chronic polyarthritis reported in detail by Henrik Sjögren in 1933,1,2 is almost exclusively a disease of middle-aged females. In the past when the signs and symptoms of keratoconjunctivitis sicca and xerostomia were present together, the term "sicca syndrome" was often applied. More recently, the term "Sjögren's syndrome" has been applied when any two components of the three originally described in the syndrome exist together. In some instances, the polyarthritis may not appear simultaneously with the onset of ocular or oral symptoms and may develop months or even years later. Similarly, keratoconjunctivitis and/or xerostomia have been first observed years after the diagnosis of arthritis is made. The arthritis is usually of the rheumatoid type, although systemic lupus erythematosus and other collagen diseases have been associated with the syndrome.

During the protracted course of rheumatoid arthritis in adults, the development of Sjögren's syndrome is not uncommon.3 As a complication of the juvenile form of rheumatoid arthritis, it is so rare that the complete syndrome of ocular, oral, and joint disturbance has not been adequately documented as occurring in any patient under age of 17 years.4 In children with juvenile rheumatoid arthritis, serious ocular complications are not uncommon (5-21%)5,6 when compared with adults similarly involved with the rheumatoid disease. The nature of the eye involvement usually comprises one of the "triad" of Smiley:5 iridocyclitis, band keratopathy, and cataract.

---

*Department of Medicine  
**Present address: University of Texas, Dallas, Texas  
***Present address: Montreal, Canada
This report presents details of the illness of a 12-year-old white girl with juvenile rheumatoid arthritis, keratoconjunctivitis sicca, and salivary involvement classical of Sjögren's syndrome.

Case History (H.F.H. #1208221): At the age of 10 years, this Caucasian child was taken to her family physician for symptoms of “fatigue and muscle pains.” She was found to be “anemic,” and oral iron therapy was prescribed. During the following 12 months, she developed intermittent swelling, stiffness, and pain in the peripheral joints. At times this lasted several weeks. There was no family history of anemia or joint disease.

From the age of 11 onward, the patient experienced continuous involvement of numerous joints. The polyarthritis ultimately affected both wrists, finger joints, elbows, shoulders, temporomandibular joints, knees, ankles, and feet. In the course of her illness there were occasions when many joints flared simultaneously. At other times only one or two joints would be disproportionately active. Shortly after the development of this polyarthritis, she noted that the joint flares were accompanied at times by a painless swelling over the face and jaws in the region of the parotid and submaxillary glands. This swelling was often associated with a persistent dryness of the mouth. Within the next 12 months, the patient began to experience eye irritation with redness, grittiness, and mild photophobia, worse at the end of the day. She also complained of a “lack of tears when crying.”

During the years of 11 to 18 (her present age), she had grown normally to a height of 62 inches and a weight of 106 pounds. She had experienced no rashes, no pulmonary infections, and no febrile episodes. Acetylsalicylic acid alone has been used in the treatment of her polyarthritis.

By the age of 16, she had developed the typical deformities and limitations in the finger and wrist joints seen in long-standing juvenile rheumatoid arthritis. The elbows could not be fully extended; several terminal interphalangeal joints had impaired ranges and were swollen, and flexion of the wrists was markedly impaired. She had a slightly retrognathic mandible and an associated restriction of mouth opening together with radiological evidence of damage to the temporomandibular joints. Cervical spine x-rays showed no significant changes. The liver edge and spleen were both palpable. These have shown no change in recent years.

The swelling of the salivary glands was usually symmetrical and could be produced by “acid or spicy foods,” particularly carbonated beverages. These the patient ultimately avoided because of the tightness which developed in the jaws together with the rapid change in the contour of the face. This latter feature was exaggerated by the patient's retrognathia. At times she also experienced great difficulty in eating dry foods because of an inadequate production of saliva. During the last six years, there has been a gradual overall deterioration in the production of saliva from the salivary glands.
This x-ray shows the ectatic changes in the left parotid gland during (a) the injection phase and (b) the secretory phase of a sialogram. Note the relatively short mandible (retrognathia) frequently seen in children with juvenile rheumatoid arthritis.
Sialograms of the patient’s parotid glands showed marked injection and secretion phase globular sialectasis and in some areas mild cavitary and destructive sialectasis (Fig. 1). Biopsy of the left parotid gland showed changes as reported previously typical of Sjögren’s syndrome. During the years of observation, there has been considerable variation in the intensity of the patient’s symptoms; at times several months of persistent swelling were followed by partial relief, also lasting some four to five months.

By age 12, the patient had developed signs of keratoconjunctivitis sicca. A recent detailed eye examination revealed no significant refractive error, and there was 20/20 acuity OU. External and slit lamp evaluations revealed mucous threads present, especially in the lower cul-de-sac, with ropey secretions and cellular debris. Instillation of fluorescein revealed only mild superficial punctate staining of the corneal epithelium and the exposed areas of the bulbar conjunctiva. Rose bengal 1% produced marked staining of the areas of the globes exposed by the palpebral fissures, with the protected areas free of stain. Characteristic triangles of staining on either side of the cornea were sharply outlined (Fig. 2). Applanation tonometry, and funduscopic examinations were normal. The staining varied in intensity, on one occasion lasting for two days following the examination. Variations in Schirmer’s test were also evident. On one occasion 25 mm (OD) and 30 mm (OS) of wetting of the filter paper occurred within a 3-minute period, although the subsequent rose bengal staining was intense and the symptoms were marked.

Pertinent blood studies included: hemoglobin 11.9 gm/100 ml, white cells 3,100 to 8,600/cu mm with a normal differential count, latex fixation test reactive, ASO titre 12 Todd units, C reactive protein positive, protein bound iodine (PBI) 4.5 μgm/100 ml, LE cell preparations negative, antinuclear factor (ANF) strongly positive threads, VDRL nonreactive, very low serum complement (14.03 units/ml), and the gamma globulins elevated at 3.7 gm/100 ml. The immuno-globulin was distributed as follows: IgA, 87 (normal 30-135) mg/100 ml; IgM, 240 (30-120) mg/100 ml; and IgG, 3,384 (600-1,400) mg/100ml. The urinalysis was normal.

Discussion

With the exception of a short report by O’Neill in 1965 describing Sjögren’s syndrome (without eye involvement) in a child of 10 years of age, other references to this disease occurring in children are rare and indirect. Available information indicates that the polyarthritis, keratoconjunctivitis sicca, and salivary gland involvement may not all be present in the same patient. In 1965, a comprehensive study of Sjögren’s syndrome reported from the National Institutes of Health indicated that 3 of 62 subjects with this syndrome developed symptoms prior to the menarche. Although specific details are not available, it has been indicated that none of these patients had salivary gland involvement. In reviewing the literature up to 1950 Henderson noted that the youngest case of Sjögren’s syndrome reported was age 17.
Figure 2
These photographs show an eye of this patient (a) before and (b) after the instillation of rose bengal (1%) dye. Note the punctate staining of the sclera with this red dye. Normally no staining occurs, while in Sjögren's syndrome the discoloration may last for hours or days.
Duncan, Epker and Sheldon

and the youngest of his own series of 121 cases was 19. Again, the coexistence of polyarthritis was not mentioned. Stenstam's report on 495 subjects primarily involved with rheumatoid arthritis revealed an incidence of keratoconjunctivitis in 9.5% (46 cases). While the minimum age of examination in this series was 15 years, Stenstam indicated that Sjögren's syndrome did not develop within the 12 months following the onset of the polyarthritis.

Our present case, a patient with classical juvenile rheumatoid arthritis, developed the complete Sjögren's syndrome with keratoconjunctivitis sicca and xerostomia between one and two years after the first recognition of the joint disease. Symptoms of the sicca syndrome have been intermittent and variable in their intensity; with eye discomfort and mouth dryness less troublesome at some times than others. All symptoms were intensified occasionally at the time of a flare-up of polyarthritis; at other times, the joints and the sicca symptoms were independent of each other. There has been some amelioration of the eye symptoms with the use of artificial tears, although their efficacy is somewhat uncertain because of the natural variability mentioned. On the other hand, we have observed considerable variation of the intensity and distribution of conjunctival staining with rose bengal when this was performed in identical circumstances but on different occasions. This occurrence of exacerbations and remissions was originally recognized by Sjögren.12

Recent studies, from an ophthalmological clinic in San Francisco on the association of eye lesions with joint disease, failed to report any instance of Sjögren's syndrome in patients under 16 years. Ansell and Bywaters had not observed Sjögren's syndrome in any of 316 juvenile rheumatoid patients followed for over a period of eight years in England, and the sicca syndrome was not recorded in any of the 544 children with rheumatoid arthritis reported by Laaksonen from Finland.13

The laboratory studies in our patient showed changes frequently seen in the Sjögren's syndrome: mild anemia, a depressed white cell count, and marked changes in the gamma globulin levels, particularly with a low IgA, a variable IgM, and a grossly elevated IgG. The antinuclear factors were present and showed strong homogeneous fluorescence. There is evidence which suggest that antibodies to different organs and tissues are quite numerous in this condition. Also, the reported presence of antibodies to thyroid, lacrimal gland, salivary gland, and gastric mucosal cells represent some degree of auto-immune reaction.

The prognosis of Sjögren's syndrome is dependent upon the associated condition. In the presence of lupus erythmatosus or polymyositis, the outlook is poorer than if it is associated with rheumatoid arthritis. In patients with no polyarthritis, there appears to be no increase in mortality due to the condition. Corticosteroids have no place in the systemic management of symptoms, and topical corticoids have been associated with little definite or lasting effect.

40
Sjögren's Syndrome

Abstract

The rare occurrence of Sjögren's syndrome in a child is reported. The complete syndrome described by Sjögren usually develops after the age of 30 years and comprises a triad: keratoconjunctivitis sicca, xerostomia and arthritis. Many published reports have included cases with only two components of the triad and where arthritis is absent the term sicca syndrome has been used synonymously.

This report describes a patient who developed keratoconjunctivitis and xerostomia with parotid swelling at the age of 12 years — two years after the onset of juvenile rheumatoid arthritis. There were fluctuations of the intensity of symptoms, at times synchronously with each other — at times independent. Sialograms showed distortion of the salivary ducts and eye staining with rose bengal demonstrated typical punctate corneal and scleral change.

Recent studies have indicated that circulating antibodies to many tissues — thyroid, lacrimal, salivary glands and gastric mucosal cells — are frequently found in subjects with Sjögren's Syndrome.

REFERENCES

Duncan, Epker and Sheldon


