STILL'S DISEASE AND RHEUMATOID NODULE OF THE SCLERA*†

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THE following report concerns a patient suffering from Still's disease who developed a rheumatoid nodule of the sclera and subsequent scleral thinning of the right eye, a finding not previously recorded in the literature.

Case Report

A woman aged 31 years first developed joint symptoms at the age of 11 years. Stiffness and pain in the joints increased, until within 9 months all her joints were affected. She has undergone numerous orthopaedic operations, including arthroplasty of both elbow joints at the age of 13 years, a right transtrochanteric osteotomy at the age of 15 years, bilateral mandibular condylectomies at the age of 16 years, and further arthroplasties of the elbows at the age of 18 years.

At present there is gross deformity with limitation of movement in the hands, elbows, shoulders, hips, knees, and ankles. She is undersized and underweight. Nodules are present on the fingers, elbows, and knees. A nodule removed from the scalp proved histologically to be a rheumatoid nodule. There is no lymphadenopathy and no skin rash and the liver and spleen are not palpable. She has been treated with prednisolone for the past 12 years in doses varying from 5 to 12.5 mg. daily.

Investigations.—Hb varies between 9.6 and 12.8 g. per cent. with a hypochromic anaemia.

ESR has varied between 16 and 46 mm. 1st hr (Westergren.)

Sheep cell agglutination positive 1 : 512.

Latex-fixation test positive 1 : 10,240.

Serum protein estimation shows a reversal of the normal albumin : globulin ratio, with an increase in the gamma globulin fraction on electrophoresis.

L.E.-cells were not detected on six occasions.

Blood urea normal.

No evidence of renal failure.

X rays of the joints of both upper and lower limbs, temporo-mandibular joints, and cervical spine show erosive bone destruction and cystic changes.

Ophthalmic History.—About 18 months before this report was drawn up the patient noted variable intermittent diplopia. This symptom was thought to be due to weakness of the left superior rectus, and there was an indurated swelling over the insertion of this muscle. The condition was treated with prednisolone drops, no biopsy was taken, and the symptoms and swelling resolved within 3 weeks, but 9 months later the patient noticed redness of the right eye, most marked supero-temporal to the limbus, and examination showed a generalized episcleral injection, most intense supero-temporally.

* Received for publication January 3, 1968.

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During the next 3 months an intensely painful, large, reddish-yellow nodule appeared on the temporal aspect of the right globe adjacent to the limbus (Fig. 1). Adjacent to and above this nodule an area of scleral thinning with dilated superficial scleral vessels was already present. The condition was treated by four sub-conjunctival injections of methyl prednisolone (Depo-Medrone), 20 mg. being given over a period of 5 weeks, as well as atropine and prednisolone drops used intensively. The nodule and scleritis gradually resolved (Fig. 2) over a 4-month period, leaving a large area of scleral thinning which is now ectatic. The area of scleral thinning superiorly has also increased.

FIG. 1.—Rheumatoid nodule on temporal side of right globe, 3 months after onset of symptoms.

FIG. 2.—Site of resolving nodule and adjacent scleral thinning (5 weeks later than Fig. 1).

The patient has been maintained on prednisolone drops locally to the affected eye in addition to the systemic steroids. No recurrence of inflammation has occurred but there has been a gradual extension of the scleral thinning which now involves about a third of the perilimbal circumference (Fig. 3). The visual acuity has remained at 6/6 in both eyes with correction throughout the course of this episode, although there have been transient changes in refraction, probably due to ciliary spasm. The fundi are quite normal.

FIG. 3.—Site of resolved nodule, showing further extension of scleral thinning with ectasia (5 months after Fig. 1).

Discussion

Although Still (1897) first described a condition consisting of an acute systemic upset with fever, skin rash, splenomegaly, and lymphadenopathy, with accompanying or subsequent joint involvement, the term Still's disease is now often applied to all patients developing polyarthritis in childhood.

Such patients may be grouped into three categories:

(a) "Juvenile rheumatoid arthritis type": patients having joint involvement similar to that seen in adult rheumatoid arthritis, except that the changes are less severe and tend to affect the larger joints. A small proportion of these patients only show positive serological tests for rheumatoid factor, and these tend to have more severe changes affecting the peripheral joints. It is these patients who often have involvement of other systems, as do adults with rheumatoid arthritis accompanied by a positive serology, and it is to this group of patients that our case belongs.

(b) "Ankylosing spondylitis type": a group of patients, the majority being males, who have involvement of one or two large joints and who often suffer changes in the spine and sacro-iliac joints later in the course of the disease.

(c) "Systemic lupus erythematosus type": the group originally described by Still, presenting with pyrexia, skin rash, splenomegaly, and lymphadenopathy. A small number of these patients have positive tests for L.E.-cells and ultimately develop frank systemic lupus erythematosus.

Although no ocular complications were mentioned by Still in his original description apart from prominence of the eyes, ocular involvement has since been recorded by many authors. These ocular symptoms comprise a triad of anterior uveitis, band-shaped keratopathy, and secondary cataract. Franceschetti, Blum, and Bamatter (1951) described fourteen patients with Still's disease with ocular complications, all of whom showed the features described above. Similarly, in the series of Smiley, May, and Bywaters (1957) and Laaksonen (1966), an overall incidence of ocular complications of 5.5 per cent. was recorded. The ocular involvement again consisted of unilateral or bilateral anterior uveitis, keratopathy, and secondary cataract. Chadwick and Rosen (1968) described the association of papillitis without anterior uveitis with Still's disease.

No case of Still's disease with a rheumatoid nodule of the sclera has been recorded as far as we can determine. Scleral thinning following the development of a scleral nodule is seen in a small number of adult patients suffering from rheumatoid arthritis, Sevel (1965). These nodules have a histological structure similar to rheumatoid nodules occurring elsewhere and there is a strong correlation between the presence of nodules and positive serological tests for rheumatoid factor. It is perhaps not surprising, therefore, that a similar picture should be seen in a patient with Still's disease of the "juvenile rheumatoid arthritis" type.

Summary

A patient is described suffering from Still's disease who developed a rheumatoid nodule of the sclera.

A scleral nodule was present adjacent to the limbus supero-temporally which resolved to leave an area of scleral thinning which has subsequently become ectatic.

No such association has been recorded previously.

Our thanks are due to Dr. H. T. Howat for permission to examine his patient.

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