CHRONIC ATROPHIC POLYCHONDRITIS*

A SOUTH AFRICAN CASE

JAMES MARSHALL, M.D., AND D. G. LE ROUX, M.R.C.P., *Departments of Dermatology and Internal Medicine, University of Stellenbosch and Karl Bremer Hospital, Bellville*

Chronic atrophic polychondritis (CAP) (polychondropathia; systemic chondromalacia; relapsing polychondritis; panchondritis, etc.) is an uncommon disease whose chief characteristic is recurrent inflammation in and eventually lysis or atrophy of cartilage. Cartilage throughout the body is attacked, but clinical signs may be localized or generalized. Recurrent inflammatory swelling of the ears is the most constant sign, although it may not be the first, and most patients complain of rheumatism. Affection of the cartilage in the respiratory system may cause death from pulmonary infection and suffocation.

Non-cartilaginous structures such as the inner ear, the sclera and the anterior uveal tract are often involved, and in a few cases there have been signs of the Gougerot-Sjögren syndrome with dryness of the mouth and eyes; electrocardiographic and histological signs of myocarditis have occasionally been discovered.

The first case was described in 1921 by Jaksch-Wartenhorst,[1] and Prof. H. Harders of Hamburg, Germany, informs us that he will shortly publish a report on some 50 cases that have since been seen.

The cardinal signs and mode of onset of the disease are portrayed in Table I. The case now reported is unusual in that the rheumatic symptoms were muscular more than arthritic, and unique in that LE cells were discovered on one occasion.

CASE REPORT

The patient, a White man aged 41 years, had the following history:

1956 - 1959: During this period he suffered recurrent attacks of pain in the left lumbar region, hip, and leg, diagnosed as fibrositis and cured by rest.

1960: Early in the year he had a severe attack. Since radiological examination revealed no abnormality he was treated for sacro-iliac strain; a supporting corset gave no relief, but the attack subsided after a month. At the end of the year another severe attack was successfully treated with sclerosing injections.

1961: The right lumbosacral area became painful; sclerosing injections were only palliative and he had attacks throughout the year. Radiological examination again showed no abnormality.

1962: Back pain persisted in lesser degree, but he began to have attacks of pain in the left great toe and instep, in the perineum and in the right shoulder and neck muscles.

When first seen by one of us (D.G. le R.) in October 1962 the main findings were sharply localized areas of tenderness along the sacrospinalis and elsewhere in the back and neck muscles with considerable stiffness and painful limitation of movement. There was extreme tenderness at the tendinous insertion of various muscles, the adductors of the thighs in particular. There was swelling of the metatarsophalangeal joint of the left great toe and redness and swelling of the left mid-tarsal joints.

Another phenomenon had appeared in April 1962. The right ear suddenly swelled and became painful, tender and dark red in colour; two months later the left ear was similarly affected. Attacks of acute inflammation of the ears interspersed with periods of relative improvement have persisted ever since. At the height of an attack the ears were swollen to about double their normal volume and the contours of the cartilage could not be distinguished; they felt rubbery and were darkly erythematous, painful and very tender to touch. The earlobes were unaffected. At the end of 1962 he was totally incapacitated for about a month by a severe attack chiefly characterized by pain in the neck and shoulder muscles, but any movement would also cause spasm and pain in the muscles of the back, thighs and abdomen. The ears were swollen and there was

*Paper delivered at the 7th Academic Staffday of the Faculty of Medicine of the University of Stellenbosch and the Karl Bremer Hospital, 4 October 1963.

TABLE I. CARDINAL SIGNS AND MODE OF ONSET OF THE DISEASE

No.	*Authors*	*Ref. No.*	*Year*	*Sex*	*Age*	*First signs*	*Joints*	*Eyes*	*Nose*	*Ears*	*Respiratory*	*Tracheotomy*	*Died*
1	Jaksch-Wartenhorst	1	1921	M	32	Rheum.	+	—	+	+	+	—	—
2	Altherr and Von Meyenberg	2, 3	1936	M	14	Rheum.	+	—	+	+	+	—	+
3	Gordon *et al*	4	1948	F	34	Rheum.	+	+	+	+	+	+	+
4	Hilding	5	1952	F	59	Rheum.	+	+	+	+	—	—	—
5	Harders	6	1954	M	16	Dyspnoea	+	—	+	+	+	+	+
6	Rogers and Lansbury	7	1955	F	61	Rheum.	+	+	+	+	—	—	—
7	Bober and Czarniecki	8	1955	F	41	Rheum.	+	+	+	+	+	+	—
8	Harwood	9	1958	M	33	Ears	+	+	+	+	+	+	+
9	Klatskin and Katzenstein	10	1958	F	63	Rheum.	+	—	+	+	+	—	+
10	Bean *et al.*	11	1958	F	58	Nose, ears	+	+	+	+	+	—	—
11	Winer and Levin	12	1959	F	30	Ears	—	+	—	+	Cough	—	—
12	Winer and Kline	13	1959	M	32	Nose	+	+	+	+	—	—	—
13	Arundell and Haserick (1)	14	1960	F	22	Ears, Nose	+	+	+	+	+	+	—
14	Arundell and Haserick (2)	14	1960	F	New born	Rheum.	+	—	—	+	—	—	—
15	Pearson *et al.* (1)	15	1960	M	33	Nose	+	+	+	+	—	—	—
16	Pearson *et al.* (2)	15	1960	F	29	Ears	—	+	—	+	+	—	—
17	Pearson *et al.* (3)	15	1960	F	45	Ears	+	—	—	+	—	—	—
18	Pearson *et al.* (4)	15	1960	M	47	Ears	+	—	—	+	—	—	—
19	Degos *et al.*	16	1960	F	47	Eyes	+	+	+	+	—	—	—
20	Coste *et al.*	17	1961	F	56	Rheum.	+	+	—	+	Dysphonia	—	—
21	Domant *et al.*	18	1961	F	63	Eyes, Ears	+	+	—	+	—	—	—
22	Kaplan *et al.*	19	1962	F	43	Ears	—	—	+	+	—	—	—
23	Marshall and Le Roux		1964	M	41	Rheum.	+	—	—	+	—	—	—

pain on pressure on the thyroid cartilage. Oral corticosteroid therapy for 3 weeks during the attack had no effect; antibiotics were equally ineffective. He has had no treatment since this time.

1963: The severe attack subsided by the end of January, but he continued to have generalized muscular pain and recurrent swelling of the ears.

Extraction of his upper teeth for pyorrhoea in March was followed by marked improvement in his symptoms.

In July the ears were normal in size, appearance and consistency, but a few focal points of tenderness to pressure on the cartilage persisted, and there was tenderness in the neck muscles and dorsal spine.

In September extraction of some teeth in the lower jaw was followed 3 days later by transitory swelling of the ears, increase of pain in the back and redness and swelling over the right tarso-metatarsal joints. There was tenderness to pressure on the thyroid and on the costal cartilages. The submaxillary glands were slightly enlarged; the patient stated that he had noted recurrent swelling over the past 4 years, but had never suffered from dryness of the mouth, nose or eyes.

No abnormality was ever found in the erythrocyte and leukocyte counts. Total serum protein 6·6 G/100 ml.; albumin 5·6 G/100 ml., globulin 2 G/100 ml. Serum uric acid on 2 occasions 5·4 and 6 mg./100 ml. A blood culture showed no growth. The Latex test was negative.

The erythrocyte sedimentation rate (Westergren) varied between 23 mm. and 90 mm. in the first hour on 6 occasions between October and December 1962; in July and September 1963 it was 8 mm.

In October 1963 scanty, but typical, LE cells were found; subsequent searches were negative. This finding misled us for some time into believing we were dealing with an atypical case of systemic lupus erythematosus.

Biopsy was consistently refused.

The patient is convinced that his improvement is attributable to the extraction of his teeth; we consider the partial remission to be probably coincidental.

DISCUSSION

CAP may begin at any age, but is commonest around the forties. It has been found at birth in a baby born of a woman with the disease; symptoms were acute for 6 weeks and the child had rheumatism for 2 years, but eventually recovered.[14] The mode of onset is variable, but rheumatism is often for a long time the only symptom, and the diagnosis is established only when inflammation of the ears or nose occurs. Women are affected oftener than men, but race, climate, occupation and heredity have no apparent influence. The erythrocyte sedimentation rate is almost always accelerated, but no specific alterations in the blood picture or constituents have been noted. The Latex or Waaler-Rose tests have been negative in cases where they have been performed. Our case is apparently the only one in which LE cells have been discovered.

Inflammatory swelling of the ears, usually both, is a cardinal sign of CAP and may be present from the start. The ears (lobes excepted) become red, swollen, firm and tender with an appearance suggestive of erysipelas, and there may be fever. Attack succeeds attack with eventual diminution in intensity. The natural contours of the cartilages disappear leaving ears as big as, or bigger than, they originally were, but deformed and rubbery; in fact, cauliflower ears (Fig. 1). The external canal may be narrowed, but hearing is not usually disturbed thereby.

The inner ear seems to have been affected in several patients who had symptoms of Ménière's syndrome and diminished hearing.

Polyarthritis affecting the large joints and the extremities is common, involvement of the intervertebral and sacro-iliac joints less so. In some cases there is simply pain, but swelling, deformity and ankylosis may occur. Diminution of joint spaces, osteoporosis and a little juxta-articular periosteal reaction may be demonstrable. The picture closely resembles that of rheumatoid arthritis.

The nasal cartilages may be affected either insidiously or, like the ears, in recurrent attacks of acute inflammation. In either case the result is a pug nose (Figs. 2 and 3).

The costal cartilages and the xiphisternum may be involved discretely or in acute attacks of inflammation. Total lysis leaves the ribs and sternum floating free in a flail chest.

Ocular involvement is quite frequent with recurrent attacks of episcleritis, iridocyclitis or both; blindness may be caused by cataract or by corneal lesions. Xerophthalmia has been noted in cases with the Gougerot-Sjögren syndrome.

Respiratory involvement may be manifest early or late, its onset insidious or abrupt. Hoarseness, cough, dyspnoea, tracheobronchitis and asthma are early symptoms, and latent involvement may be found in nodular thickening of the epiglottis or tracheal rigidity demonstrable in soft X-ray photographs. Stenosis of the air passages may require tracheotomy, but this is only of temporary benefit. Slow collapse of the bronchial cartilages impedes ventilation and usually leads to atelectasis, although bullous emphysema may have arisen in one case where spontaneous pneumothorax occurred.[5] The terminal episode is usually a bronchopneumonia.

The disease progresses in bursts of activity alternating with quiescent spells, adding symptoms as it goes. It may run its course to a fatal end in a few months, or grumble slowly for many years.

At autopsy changes are found in all the cartilages, which are soft and shrunken; the trachea loses its rigidity and its walls collapse.

The matrix of the cartilage becomes progressively more acidophilic, and the chondrocytes lose their cytoplasm until eventually only nuclear remains are found in the areas most affected. The tissue around affected cartilage is at first oedematous with a perivascular granulomatous infiltrate; later come signs of organization with many condensed elastic fibres and a mononuclear infiltrate. Eventually most of the cartilage is replaced by fibrous tissue, and what remains shows irregular distribution of chondrocytes which are totally lacking in places. Calcification and even ossification may occur. Rheumatic granulomas have been found in the heart muscle and in the neighbourhood of joints.[2]

Two major theories on the origin of CAP have been propounded. Bean[11] suggested that it might be an acquired metabolic error and that the mechanism involved was perhaps analogous to the dissolution of cartilage and release of chondroitin sulphate produced by papain in experimental animals. Thomas and others[20, 23] demonstrated that the

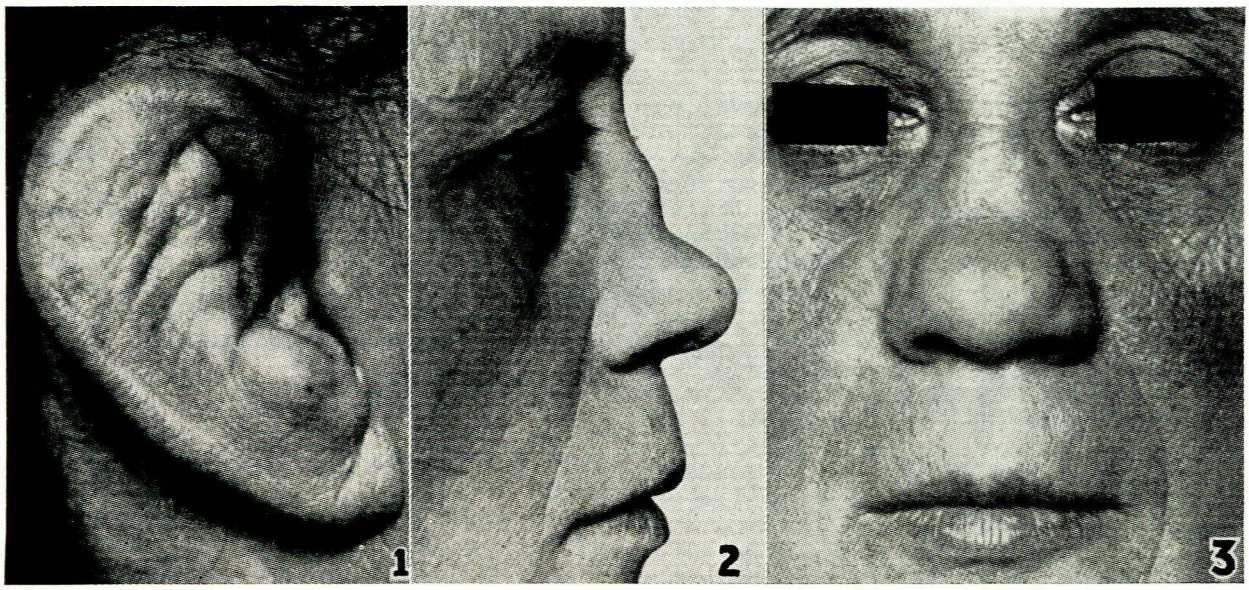

Fig. 1. Chronic atrophic polychondritis. Deformity of ear (De gos).
Fig. 2. Chronic atrophic polychondritis. Deformity of nose (De gos).
Fig. 3. Chronic atrophic polychondritis. Deformity of nose (De gos).

intravenous injection of crude papain or of inactivated (by sulphydryl blocking agents) crystalline papain causes softening of cartilage in the ears and elsewhere in rabbits; the cartilage loses its basophilic staining characteristics and chondroitin sulphate is released into the blood and excreted in the urine. The cartilage is slowly restored if papain is withheld; but continuous administration of corticosteroids inhibits repair. Potter *et al.*[24] produced a similar attack on rabbit cartilage by Vitamin A intoxication.

An autoallergic mechanism was invoked by Harwood,[9] although he was unable to produce any reaction in guinea-pigs by injecting or implanting suspensions or pieces of their own cartilage. Glynn and Holborow[25] however, produced hypersensitivity to chondroitin sulphate with lesions of the ears in rabbits by the intravenous inoculation of chondroitin sulphate-streptococcal antigen. Streptococcal infections figure in the history of a number of the reported cases of CAP.

The occasional finding of the Gougerot-Sjögren syndrome with CAP together with the discovery of LE cells and muscular involvement in our case supports the second hypothesis and suggests a relationship to the collagenoses rather than to the disorders of metabolism. The nature of the target substance common to cartilage, eye and inner ear is unknown. The diversity of tissues attacked is not made manifest in any of the titles proposed for the disease and it will obviously have to be renamed in time.

In CAP only corticosteroids have any effect, and that simply suppressive. Pearson *et al.*[15] report that prednisone, 30 mg. daily, controlled the acute phase in their cases and that 10 - 12·5 mg. daily was required for maintenance; exacerbations repeatedly followed reduction of dosage to, or below, 7·5 mg. daily.

SUMMARY

A case of chronic atrophic polychondritis with the unusual features of predominantly muscular rheumatism and the discovery of LE cells is reported. The literature is reviewed.

We are indebted to Prof. R. Degos, Saint-Louis Hospital, Paris, for the photographs illustrating the end phase of the attack on the cartilage of the nose and ears.

REFERENCES

1. Jaksch-Wartenhorst, R. (1921): Med. Klin., **17**, 1341.
2. Altherr, F. (1936): Arch. Path. Anat., **297**, 445.
3. Von Meyenberg, H. (1936): Schweiz. med. Wschr., **66**, 1239.
4. Gordon, E. J., Perlman, A. W. and Schechter, N. (1948): J. Bone Jt Surg., **30-A**, 944.
5. Hilding, A. C. (1952): Arch. Intern. Med., **89**, 445.
6. Harders, H. (1954): Schweiz. med. Wschr., **89**, 712.
7. Rogers, F. B. and Lansbury, J. (1955): Amer. J. Med. Sci., **229**, 55.
8. Bober, S. and Czarniecki, W. (1955): Schweiz. med. Wschr., **85**, 448.
9. Harwood, T. R. (1958): Arch. Path., **65**, 81.
10. Klatskin, G. and Katzenstein, R. E. (1958): Resident Physician, **4**, 75.
11. Bean, W. B., Drevets, C. C. and Chapman, J. S. (1958): Medicine (Baltimore), **37**, 353.
12. Winer, L. and Levin, H. (1960): Arch. Derm., **81**, 469.
13. Winer, L. and Kline, H. (1960): *Ibid.*, **81**, 469.
14. Arundell, F. and Haserick, J. (1960): *Ibid.*, **82**, 439.
15. Pearson, C. M., Kline, H. M. and Newcomer, V. D. (1960): New Engl. J. Med., **263**, 51.
16. Degos, R., Guilaine, J. and Decaudin, A. (1961): Presse méd., **69**, 1117.
17. Coste, F., Laurent, F., Basset, F. and Chaouraki, L. (1961): Ligue française contre Rhumatisme, séance 18 janvier.
18. Domant, A., Duperrat, B., Hazard, J., Hillemand, B. and Catton, D. (1961): Bull. Soc. franç Derm. Syph., **68**, 177.
19. Kaplan, H. L., Norris, J. E., Freeman, B. S. and Brown, W. G. (1962): J. Amer. Med. Assoc., **180**, 164.
20. Thomas, L. (1956): J. Exp. Med., **104**, 245.
21. McClusky, R. T. and Thomas, L. (1958): *Ibid.*, **108**, 371.
22. Spicer, S. S. and Bryant, J. H. (1957): Amer. J. Path., **33**, 1237.
23. Tsaltas, T. T. (1958): J. Exp. Med., **108**, 507.
24. Potter, J., Weissmann, R., McClusky, R. T. and Thomas, L. (1959): Proceedings of the meeting of the American Rheumatism Association, Detroit, 11 December.
25. Glynn, L. E. and Holborow, E. J. (1952): Lancet, **2**, 449.