Relapsing polychondritis

By N. H. Mahindrakar and L. J. Libman (Manchester)

Relapsing polychondritis is a disease of unknown aetiology affecting the cartilaginous structures in the body. The cartilages of the nose and ears are involved at an early stage of the disease and this is of direct interest to the otolaryngologist.

Case reports of this disease have appeared infrequently in the literature. Pearson, Kline and Newcomer (1960) summarized the findings in ten previously reported cases and added two of their own. Kaye and Sones (1964) reported fourteen cases and discussed the clinical findings and treatment.

The disease has been described under different names. Jaksch-Wartenhorst (1923) called it polychondropathia. Altherr and Von Mayenburg (1936) named it polychondromalacia. Bober and Czarniecki (1955) called it chronic atrophic polychondritis. Pearson (1960) called it relapsing polychondritis, and this seems to be the most appropriate, as the disease has periods of 'relapsing', and 'polychondritis' suggests that the basic pathology is the inflammation of more than one cartilage in the body.

The disease commonly affects adults between the second and the fourth decades though no age group is immune. Kaye and Sones (1964) reported two cases in whom the symptoms appeared at the age of two years. Both sexes are equally affected.

The common regions involved are the cartilages of the nose and ears. The predominant symptoms are nasal obstruction due to narrowing of the anterior nares following collapse of the alar cartilages. The softening of the septal cartilages leads to depression of the nasal bridge. The auricles present a classical picture of inflammation of the cartilage with collapse. During the acute episodes the auricles are red, swollen and tender, and can be deformed with little pressure. The external auditory canals may collapse leading to conductive deafness.

The involvement of the larynx may present as hoarseness and pain in the thyroid cartilage. Mirror examination may reveal swelling and congestion of the laryngeal mucous membrane. The thyroid cartilage may feel soft, and tenderness may be elicited. When the tracheal cartilages are involved stridor develops.

The costal cartilages may be involved. The cartilages of the joints of the limbs may be affected giving rise to joint pains.

The extrachondral lesions include the inner ear and the eye. The inner ear involvement can give rise to perceptive hearing loss, tinnitus or vertigo. The reason for the inner ear involvement is not known. The ocular lesions include conjunctivitis, iritis, episcleritis, and chorio-retinitis.

In addition to the above symptoms, the patient may present general non-specific symptoms such as lethargy, loss of weight, fever and a general feeling of being unwell.

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Case Report

W.M., a female aged 26 years, was admitted to the Manchester Royal Infirmary under the care of a Consultant Physician, in June 1965; she had been generally unwell for a year. During this time she had sore throats, nasal obstruction, and pain in the joints of her limbs. She was diagnosed as rheumatoid arthritis and was treated with small doses of steroids with little benefit.

Within a few months she developed hoarseness, pain in the region of the thyroid cartilage, increased nasal obstruction, cough and dyspnoea.

She was re-admitted to the hospital. On admission she was found to have inspiratory stridor with retraction of the supra-clavicular and the intercostal spaces. She developed pyrexia and episodes of cyanosis. There was narrowing of the anterior nares due to collapse of the alar cartilages. The nasal bridge was depressed, and the cartilaginous parts of the external auditory canals were collapsed.

The thyroid cartilage was soft and tender, and on mirror examination the laryngeal mucosa was uniformly red and swollen.

The investigations carried out showed the haemoglobin to be 72 per cent, white cell count 19,000/c.mm. and the E.S.R. 54 mm./hour. The serum albumen and globulin were 3·7 g. and 4·1 g. per 100 ml. of blood respectively. The electrophoretic pattern showed increase in the gamma globulin level. Repeated tests for lupus erythematosus cells in the blood were negative. The electrocardiogram, the chest X-ray, the blood urea and the electrolytes were normal.



FIG. 1.
Showing deformity of the nose.

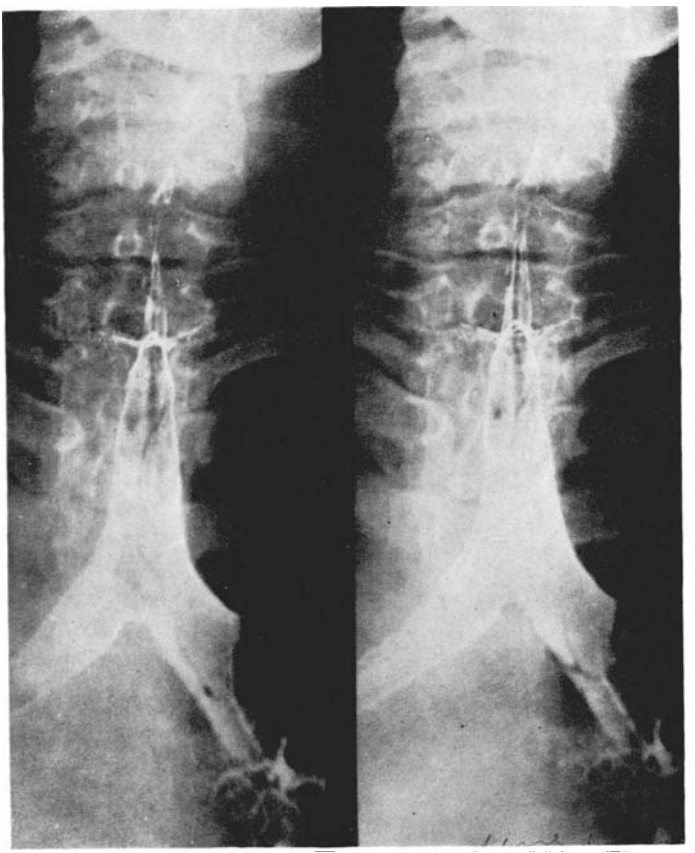
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The inspiratory stridor gradually became worse and a tracheostomy was performed under local anaesthetic to relieve the obstruction of the airway. At the same time a bronchoscopy was performed and collapse of the cervical trachea was observed.

The histopathological study of a piece of tracheal cartilage removed at the time of tracheostomy showed changes in keeping with the diagnosis of polychondritis.

Subsequently, a tracheo-bronchogram was performed and this showed marked narrowing of the cervical trachea. The lumen was reduced to half a centimetre in diameter and narrowed further on inspiration. The thoracic part of the trachea and the bronchi were of normal calibre.

With 100 mg. of prednisolone a day the symptoms markedly improved and the E.S.R. fell to 9 mm./hour in two months. The dose of prednisolone was gradually reduced to 30 mg. a day over eight months. The shape of the



(a) FIG. 2. (b)

Tracheo-bronchogram showing marked narrowing of the cervical trachea. The thoracic trachea and bronchi are of normal calibre.
(a) Inspiration. (b) Expiration.

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patient's auricles and nose showed improvement. The pain in the auricles and the thyroid cartilage was relieved. However, the patient developed moon face, marked increase in weight, and severe backache due to osteoporosis of the dorsal spine. These side-effects of the steroid therapy made her life rather miserable. In view of this, the prednisolone dosage was reduced to 20 mg. a day. Unfortunately, the patient developed recurrence of symptoms and the E.S.R was raised to 50 mm./hour.

At this stage, the patient having developed symptoms of steroid toxicity, it was felt unwise to increase the dosage of prenisolone.

Presuming that polychondritis could be an auto-immune disease, she was put on Imuran, an immuno-suppressive drug. Imuran suppresses the production of antibodies to auto-antigens.

For the last four months, the patient has been on 50 mg. of Imuran daily. Her symptoms have improved a great deal and the E.S.R. fell to 7 mm. in the first hour.

At present the patient is progressing satisfactorily on 150 mg. of Imuran and 20 mg. of prednisolone daily.

Discussion

The aetiology of polychondritis remains unknown. The most accepted theory is that it is a collagen disease, because it is a primary and generalized



FIG. 3.
Showing improvement in the shape of the nose.

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non-specific inflammation of cartilaginous tissue, with raised E.S.R., and usually it responds to steroid therapy.

Pearson, Kline and Newcomer, suggest that it may be a hypersensitivity reaction as many observers have shown that the softening and the collapse of rabbits' ears is associated with a loss of basophilia, reduction of chondro-muco-protein, and chondroitin sulphate in the cartilages following intravenous injection of papain. However, the exact relationship between these observations and the disease in human beings is not clear. They also raise the possibility that the disease could be due to a generalized metabolic or enzymatic defect, the nature of which is as yet unexplained.

Could it be a collagen disease? The clinical response of the above reported case to Imuran may suggest this possibility.

Dolan, Lemmon and Teitelbaum (1966) performed direct fluorescent studies on the serum of two patients with polychondritis to find out the presence of antibodies to the cartilage. However, their result was inconclusive.

The organs most commonly involved are the nose and the ears, and the patient may be referred to the otolaryngologist in the early stages of the disease because of nasal obstruction, deformity of the nasal bridge or painful and deformed auricles.

The important change in the blood is the raised E.S.R. It is a direct index of the activity of the disease and is a guide in controlling the steroid dosage.

The radiological investigations are helpful in assessing the extent of involvement of the tracheo-bronchial tree. The state of the tracheo-bronchial tree is of paramount importance prognostically, as the involvement of the distal part of the trachea or bronchi often leads to fatal pulmonary complications.

Tomograms often do not give a clear outline of the lumen of the tracheo-bronchial tree. The most helpful method is by contrast X-ray studies using a radio-opaque substance; Dianosyl is the one commonly used. The technique is the same as for doing a bronchogram and the dye is allowed to outline the trachea as well. In the case described, the lower part of the trachea and bronchi are wide open and rigid, while the upper collapsed part of the trachea has been bypassed by a tracheostomy.

The histopathological changes in relapsing polychondritis show inflammation with replacement by fibrous tissue. There is absence of the usual basophilic staining of the tissues. There is also perichondral oedema and infiltration with lymphocytes, plasma cells, eosinophils and macrophages. The cartilage may show fragmentation followed by replacement with granulation tissue and subsequent fibrosis.

The majority of the published cases responded well to 20–30 mg. of prednisolone daily. The amount of steroids given is controlled by the E.S.R. reading.

The reported case did not respond to 30 mg. of prednisolone daily. To bring the disease under control, she received as much as 100 mg. of prednisolone a day for two months. This large dose of prednisolone did bring the E.S.R. to a near normal level. Unfortunately, she developed symptoms of steroid toxicity.

With the belief that relapsing polychondritis could be an auto-immune disease, the patient received 150 mg. of Imuran daily, and this has improved the clinical condition.

Imuran is chemically a 6–(1 Methyl 4 Nitro–imidozol 5 ythio) purine. It

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is a cytostatic agent and can cause bone marrow depression. It has been shown to suppress the development of haemoglutinins to sheep cells in mice. It has also been found to be effective in facilitating the survival and function of renal transplants.

The average dose of Imuran is 150 mg. a day. Regular checks on white cell count and platelet counts should be done to avoid the complications of bone marrow depression.

Summary

A case of relapsing polychondritis is described. The early symptoms were referred to the nose and the ears. Subsequently, she developed joint pains, hoarseness and inspiratory stridor. The nasal bridge was depressed and the auricles were soft and collapsed. The thyroid cartilage was tender and the laryngeal mucosa was uniformly red and swollen. The investigations showed anaemia, leucocytosis, high E.S.R. and raised gamma globulin level in the serum. The tracheo-bronchogram showed marked narrowing of the cervical trachea.

The patient did not respond to the usual doses of prednisolone. She received 100 mg. of prednisolone daily to bring the disease under control. Subsequently, the large steroid dosage had to be reduced to minimize the toxic effects. Presuming on emperical grounds that the condition could be an auto-immune disease, the patient was put on 150 mg. of Imuran daily to which she responded favourably. In view of the satisfactory responses to Imuran, the suggestion arises that relapsing polychondritis may well be an auto-immune disease.

The commonest manifestations of relapsing polychondritis involves the nose and the external ears. Many reported cases in the literature had unnecessary multiple sinus and nasal operations before a correct diagnosis was made. In view of this, the early recognition of the disease may be the responsibility of the otolaryngologist.

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REFERENCES

Bober, S., and Czarniecki, W. (1955) *Schweiz. med. Wschr.*, **85,** 448.
Dolan, D. L., Lemmon, G. B. and Teitelbaum, S. L. (1966) *Amer. J. Med.*, **41,** 285.
Jaksch-Wartenhorst, R. (1923) *Wien. Z. inn. Med.*, **6,** 93.
Kaplan, H. L., Norris, J. E., Freeman, B. S., and Brown, W. G. (1962) *J. Amer. med. Ass.*, **180,** 164.
Kaye, R. L., and Sones, D. A. (1964) *Ann. int. Med.*, **60,** 653.
Pearson, C. M., Kline, H. M., and Newcomer, V. D. (1960) *New Engl. J. Med.*, **263,** 51.
Thomas, L. (1956) *J. exp. Med.*, **104,** 245.
Von Meyenburg, R. (1936) *Schweiz. med., Wschr.*, **17,** 1239.

Royal Infirmary,
Manchester.

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