Two weeks after admission the patient had a grand mal convulsion. This attack appeared to be different from the episodes he had had before admission. C.S.F. examination was negative. E.E.G. showed considerable generalized abnormality, the background activity being dominant from between 6 and 7 cycles per second.

By about the third week of admission the liver appeared to have diminished in size, and it was then possible to detect a separate ballotable mass in the left hypochondrium. A few pus cells and red cells were noted in the urine, and these findings led on to intravenous pyelography. This revealed some displacement and deformity of the calices of the left kidney, and possibly a mass arising from the midportion of that kidney.

A left nephrectomy was performed on 30 September 1965 (Mr. W. Campbell). The kidney was seen to be completely replaced by tumour tissue, and there was extension into the renal vein. Histology confirmed that the tumour was a hypernephroma. There was no evidence of hepatic secondaries, either grossly or on microscopy of a biopsy specimen.

The patient made a good recovery after the operation. He gained weight rapidly, the hepatomegaly disappeared, and the liver-function

tests reverted to normal (see Chart). Two months after the operation an E.E.G. was normal. Ten months after the nephrectomy the patient was still very well. Four months later he died quite suddenly. Necropsy was limited to determining the cause of death. This was due to coronary thrombosis. There was no macroscopic evidence of metastasis in the liver or other organs, but the brain was not examined.

COMMENT

Hepatomegaly and non-metastatic liver-cell dysfunction, with return to normal on removing the renal lesion, is now well documented (Stauffer, 1961; Greenberg *et al.*, 1964; Mohamed 1965a), our case being the fifteenth to be reported. It is important that this syndrome is recognized, for there is a tendency to refuse operation to patients with hepatomegaly and a raised alkaline phosphatase on the assumption that the patient already has liver metastasis and the condition is then irremediable (Sherlock, 1963). Patients with hypernephroma may have extreme cachexia and impairment of cell function without metastasis (Creevy, 1935; Mohamed, 1965b).

The reversal of the signs and symptoms after nephrectomy in the above case could also have been related to spontaneous regression of carcinoma at other sites, and this possibility cannot be definitely excluded, particularly in relation to the brain.

It is a pleasure to acknowledge the assistance given by Dr. John Laidlaw, Chalfont Colony, Bucks, with the interpretation of the E.E.G.s. We are grateful to Mr. W. Campbell, who performed the nephrectomy and liver biopsy, and to Dr. D. G. Rushton, who carried out the necropsy.

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Changes in E.S.R., haemoglobin, platelets, plasma alkaline phosphatase, B.S.P. excretion, plasma proteins, and body weight before and after nephrectomy have been plotted. The vertical dotted line indicates the day of operation. The improvement in haemoglobin level before operation was due to blood transfusion.

Relapsing Polychondritis

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Relapsing polychondritis is a disease of which the main features are episodic inflammation, followed by dissolution, of cartilage throughout the body. In addition, there is often inflammation of the iris and sclera. It should be noted that the disease has been described under various titles. These include "chondromalacia," "chronic atrophic perichondritis," "diffuse perichondritis," "polychondropathia," and "relapsing polychondritis." The latter designation has been favoured by most recent authors.

The first case was probably described by Jaksch-Wartenhorst (1923). Since then nearly 50 cases have been reported, but only two of them are in British literature (Rhys Davies and Kelsall, 1961; Thould, Stansfeld, and Wykeham Balme, 1965).

CASE REPORT

A housewife had been in previously good health apart from a cough which she said she had had all her life. Her family history was non-contributory. In 1956, when aged 41, her cough became worse, and some wheezing and mucopurulent sputum developed. The symptoms were all worse in the winter.

In 1961 painful swelling of the ankles and knees developed. The erythrocyte sedimentation rate (E.S.R.) at this time was 60 mm. in one hour (Westergren). The Rose-Waaler test was said to be positive but the titre was not known. She was given a short course of corticosteroids and her joint symptoms subsided.

In January 1962 she became short of breath and developed stridor. Her voice became hoarse. She developed swelling of both pinnae and her ears became "like cauliflowers." At the same time she had an episode of right-sided iritis. Effusions into both knee joints were noted. At this point some falling in of her nose was noticed. Laryngoscopy showed "swelling and spasm of the whole laryngeal inlet." A biopsy of the tracheal wall showed extensive chronic granulation tissue deep in the wall with dense hyaline fibrosis

of the lamina propria. The Rose-Waaler and Kveim tests were negative. Renal function tests were normal. Tracheostomy was performed because of the respiratory obstruction. A small dose of radiotherapy was given to the larynx.

She had a persistent cough with much sputum, which required to be aspirated many times daily. Her ears gradually returned to their normal size. Apart from several further episodes of iritis her condition remained otherwise virtually unchanged until February 1965, when she experienced some difficulty with inserting the tracheostomy tube. Bronchoscopy showed narrowing of both the trachea and the origins of the main bronchi (see Table) and the mucosa was granular and irregular. Biopsy of the tracheal mucosa showed squamous metaplasia with marked fibrosis. At this point prednisolone 20 mg. daily was given.

Corrected Measurements of Tracheal and Bronchial Diameters

	Normal Control	Patient
Trachea 2 cm. above carina	2 cm.	0.74 cm.
Right main bronchus 1 cm. below carina ..	1.2 "	0.67 "
Left " " 1 " " " " ..	1.0 "	0.67 "

She was admitted to the Churchill Hospital in April 1965, under the care of Dr. J. M. K. Spalding, because of difficulty with her tracheostomy. The depression of the middle of her nose was pronounced (Fig. 1) and the nasal cartilages were very soft. The right ear was somewhat soft and flabby. Her hearing was normal to ordinary ward testing. There was no objective evidence of past iritis. The tracheostomy tube was in situ and appeared to be satisfactory. There was some tenderness over the sternomanubrial joint. The other joints were objectively normal.

Investigations showed a mild hypochromic anaemia. The E.S.R. was 11 mm. in one hour (Westergren). The Wassermann reaction and Rose-Waaler test were negative. L.E. cells were not seen. No antinuclear factor was demonstrable. The total serum proteins

FIG 1.—Photograph showing depression of middle of nose.

FIG. 2.—Tracing of inclined frontal plane tomograph taken through the plane of trachea and main bronchi.

were 6.6 g./100 ml. Serum electrophoresis showed a slight increase in the alpha-2-globulin and gammaglobulin. A midstream specimen of urine was normal. There was no excess of mucopolysaccharides in the urine. The blood urea was 26 mg./100 ml. The maximum urinary specific gravity was 1020. The creatinine clearance test was 82 ml./min.

Plain x-ray films of the chest were normal, but tomograms revealed gross narrowing of the trachea and main bronchi (Fig. 2). Lung-function tests were performed by Dr. R. Marshall, who reported as follows: "In this patient there is considerable increase in airway resistance. At normal flow rates (up to 0.5 l./min.) the resistance during inspiration is approximately the same as during expiration. At higher flow rates the resistance increases on expiration and the maximum possible expiratory flow is about 0.7 l./sec." There was sclerosis of the sternomanubrial junction. X-ray examination of the cervical and lumbar spines showed only non-specific degenerative changes. X-ray films of the hands and knees were within normal limits.

Since April the patient had remained reasonably well and was taking prednisolone 5 mg. t.d.s. In October 1965 she had a mild relapse involving painful swelling of both ears. This subsided without a further increase in her steroid therapy.

DISCUSSION

This patient shows the classical features of relapsing polychondritis. Most authors have reported single cases. Kaye and Sones (1964) reviewed 14 personal cases, together with 27 cases collected from the literature.

The disease is characterized by episodes of activity in which cartilage in various sites becomes inflamed. The episodes are often accompanied by low-grade fever and malaise. Recurrent episodes of activity ultimately produce softening of the affected cartilage. The pinnae become soft and flabby and the nose assumes a "saddle-nose" deformity. Involvement of the middle and inner ear may lead to deafness.

The cartilages of the respiratory tract are often involved, and ultimately narrowing of the trachea and bronchi may occur. This involvement of the respiratory tract is the usual cause of death. Cartilaginous joints such as the symphysis pubis and the manubrium sterni are sometimes affected. Synovial joints are affected in some cases, and occasionally a polyarthritis resembling rheumatoid arthritis may occur (Thould *et al.*, 1965). Three of the cases reported by Kaye and Sones (1964) fulfilled the criteria for definite rheumatoid arthritis laid down by the American Rheumatism Association (1959). The eye is involved in about one-third of reported cases. Scleritis, conjunctivitis, and iritis may all occur.

The E.S.R. is often raised during an exacerbation. Tests for rheumatoid factor are usually negative. During an exacerbation there may be an increase in urinary mucopolysaccharide excretion. The cause of the disease is unknown.

Steroids appear to suppress the inflammatory reaction, and if the respiratory tract is involved, long-term steroid therapy is probably indicated. Treatment is otherwise symptomatic.

My thanks are due to Dr. J. M. K. Spalding for permission to publish this case; and to Dr. D. Davies, Physician-Superintendent, Ransom Hospital, Rainworth, near Mansfield, Notts, for referring the patient and for the early investigations. I also wish to thank Dr. R. Marshall for undertaking the lung-function studies.

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