SUMMARY

In rheumatoid arthritis, it is supremely important that patients should be able to walk and to use their hands.

If flexion deformities of knees and wrists are prevented, arthritic patients will never become crippled.

Deformity of the knee can be prevented by keeping it extended in a caliper and encouraging the patient to walk.

Deformity of the wrist can be prevented by continuous immobilization in plaster splints which allow the fingers to be used and the hand to be rotated.

Arthritic joints which are immobilized for a few weeks do not ankylose.

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RÉSUMÉ

L'arthritique grabataire dans la plupart des cas est devenu impotent par déformation en flexion des genoux. Un cercle vicieux s'est établi ensuite puisque le repos au lit est désastreux pour les articulations, les os et les muscles. Les sujets atteints de polyarthrite chronique évolutive ne doivent donc s'aliter que pour des périodes aussi brèves que possible. Si les genoux sont immobilisés en flexion, l'auteur suggère de les étendre sous anesthésie et de les conserver en extension par l'application immédiate d'un appareil plâtré. Il faut se garder d'appliquer une force trop grande au cours de cette manœuvre dans la crainte de rompre les os devenus fragiles. On peut conserver le tonus des quadriceps par des exercices de contraction répétés plusieurs fois par jour. En cas de faiblesse de ces muscles il convient de les aider dans leur tâche par l'application de plâtres cylindriques.

Les poignets sont la principale source de douleurs dans la P.C.E. On ne doit pas permettre que ces articulations fléchissent en pronation; il faut au contraire les immobiliser en bonne position et laisser le pouce et les doigts libres. Cette immobilisation diminue beaucoup les douleurs dans les doigts.

L'immobilisation d'une articulation enflammée ne mène pas nécessairement à l'ankylose. Au contraire lorsque l'inflammation s'est calmée l'articulation gagne une plus grande mobilité. Le secret du succès est de ne pas forcer une articulation malade au delà du champ d'action que lui permet la lésion. Si le cartilage a été détruit, l'immobilisation dans un appareil plâtré permet à l'ankylose de s'établir dans la position la plus favorable et la plus utile.

Case Reports

PNEUMOCYSTIS CARINII PNEUMONIA*

REPORT OF A CASE DIAGNOSED DURING LIFE

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THERE HAS BEEN an increasing awareness of the interesting condition, pneumonia due to *Pneumocystis carinii*, in the past few years. There are possibly many instances of this disease hidden in autopsy files, and now that the condition is recognized as a clinical entity many more cases will doubtless be recognized in their true light.

The patient was an infant girl born October 6, 1958. Her first admission was to another hospital on October 31, 1958, at the age of 25 days, with a severe staphylococcal skin infection particularly affecting the abdominal wall, due to *Staphylococcus aureus*, coagulase positive, which was sensitive to chloramphenical (Chloromycetin), erythromycin and oxytetracycline (Terramycin). This cleared up on local application of neomycin ointment. During this admission a systolic murmur was noted which seemed to be maximal over the apex of the heart, but an electrocardiographic tracing was considered normal. At the time of discharge on November 14, she weighed 6 lb., having made no gain in the preceding 28 days. Her next admission to a second hospital was two weeks later. She then presented with vomiting, diarrhœa and skin pustules. This was treated with tetracycline (Achromycin) and penicillin. On the day before the present admission she had neck stiffness and irritability, but spinal fluid examination was negative. She was transferred to the Brandon General Hospital on December 9, at which time she had a severe staphylococcal skin infection.

Physical examination showed a slightly dehydrated pale, malnourished infant of about two months; weight 6 lb., temperature 104° F. The infant had a weak cry and lay with her head arched backwards. There were many pustules over the upper part of the trunk and several abscesses on the scalp, the largest measuring about 2.0 cm. in diameter.

On December 10, the large abscesses on the scalp were opened and drained, and culture yielded a profuse growth of *Staphylococcus aureus*, coagulase positive, sensitive to erythromycin and chloramphenicol. The white cell count at this time was 12,400 per c.mm. A diagnosis of staphylococcal pyodermia and malnutrition was made.

On treatment with hexachlorophene soap (pHisohex) and chlorhexidine (Hibitane) locally, and erythromycin

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systemically, the infant gradually improved. By December 28, the skin was clear, and the infant was discharged. At this time the weight was 7 lb. 7 oz.

Final admission.—The final admission was on January 30, 1959, at the age of four months. The mother stated that four days before this admission the infant had developed recurrence of skin infection with small pustules on the scalp. She was seen by her own doctor at home, where treatment was again instituted with erythromycin orally. Two days before admission the baby began to cough and run a low-grade fever, and for 24 hours before admission had been taking very little by mouth.

The local doctor when contacted stated that the home conditions were very poor and the mother apparently had not always carried out the treatment recommended. One of the siblings had been treated for rat bites, and the condition of the home was quite unsanitary.

Physical examination.—The patient was a very small, scrawny, sick-looking infant appearing to be one month of age but actually four months old. There was marked lack of subcutaneous fat but no loss of skin turgor or other evidence of dehydration. Many small pustules were seen on the scalp, two of these being approximately 1.0 cm. in diameter. No other skin lesions were present. The infant had marked buccal moniliasis. Respirations were rapid and shallow and the alæ nasi flared with each inspiration. No dullness could be noted in either lung field and there were no definite adventitious sounds. Some retraction of the lower costal margin was present on inspiration. No other significant abnormalities were noted. The weight was 7 lb. 15 oz. An admission diagnosis of staphylococcal pyoderma, possible staphylococcal pneumonia, thrush, and severe malnutrition was made. A radiograph of the chest was reported as follows: "The heart and great vessel shadows are not remarkable. The diaphragms are within normal limits. The lung fields appear clear." There was no evidence of any pleural effusion or pneumothorax.

A vein was cut down upon and a hypotonic multi-electrolyte infusion started. Intramuscular chloramphenicol (Chloromycetin) 100 mg. every four hours was started on admission.

On February 1, the child's condition had deteriorated. No physical signs of pneumonia were found, but respirations were grunty and rapid. A radiograph showed widespread pneumonia but no pneumothorax or empyema. The radiological report was as follows: "Re-examination of the chest shows some infiltration at the right base. Some less extensive peribronchial infiltration is also shown at the right apex. There is some quite dense consolidation of the left upper lobe. The right lower lobe appears generally clear. The findings are consistent with an extensive pneumonia. There has been marked deterioration in the appearance of the chest in the interval since the last examination 48 hours previously." By 3:45 p.m. on February 1, her condition was worse, with grey cyanosis. The chest was still clinically normal.

On February 2, at 11:00 a.m., there was not much change; some crepitations were present over the right base. The infant lay with her head retracted because of air hunger, but there was no neck stiffness.

On February 3, 11:00 a.m., the infant was deeply cyanotic, requiring continuous oxygen in a croupette. If the croupette was opened the infant became almost

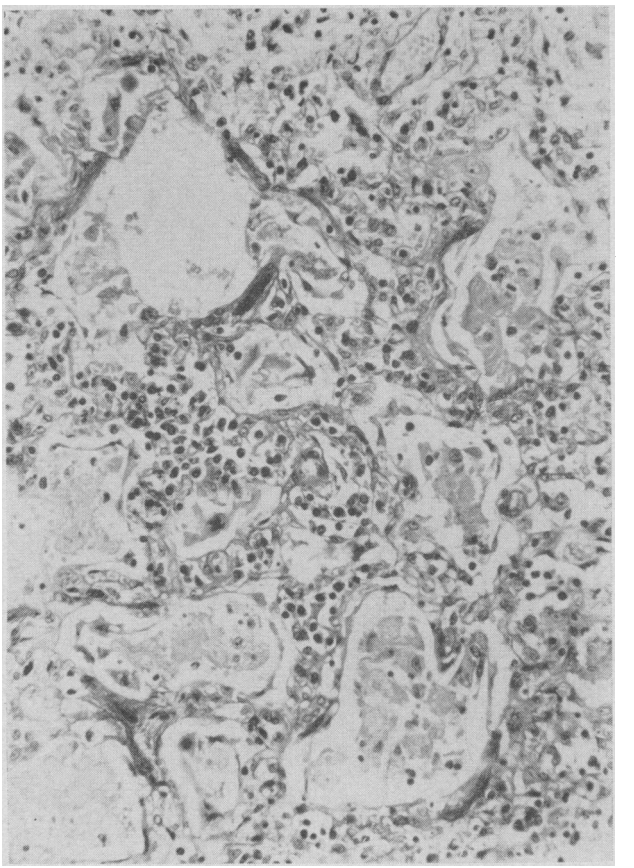

Fig. 1.—Histological section of lung showing interstitial infiltrate and alveolar exudate. Giemsa stain × 184.

black. Occasional fine crepitations and rales were heard over the right base. A bedside radiograph taken at this time was reported as follows: "A single AP film of the chest was made on the ward with the portable equipment. This again shows the extensive consolidation involving the major part of the right lung and the left upper lobe. The left lower lobe still appears generally clear. The findings are again consistent with a widespread pneumonia."

Until this time it had been considered likely that the infant had a staphylococcal pneumonia. Because of the radiological appearance, the very marked cyanosis and air hunger associated with minimal, if any, local signs of pulmonary involvement, a diagnosis of *Pneumocystis carinii* pneumonia was now considered probable.

On February 4, the infant appeared slightly improved, although she was quite distended. The cyanosis did not appear quite as marked as on the previous day. On examination of the chest no adventitious sounds were heard. At 4:00 p.m. on February 4, the cyanosis suddenly became very marked, and the baby ceased to breathe.

Laboratory Examination

December 10, 1958. Hb. - 8.6 g. %. Hæmatocrit - 27%. M.C.H.C. - 31%. Leukocyte count - 12,400. Differential: Neutrophils - 66%. Lymphocytes - 30%. Band forms - 4%. Smear - essentially normal other than slight hypochromia.

January 31, 1959. Hb. - 11.8 g. %. Leukocyte count - 18,200. Unfortunately a differential count was not done at this time.

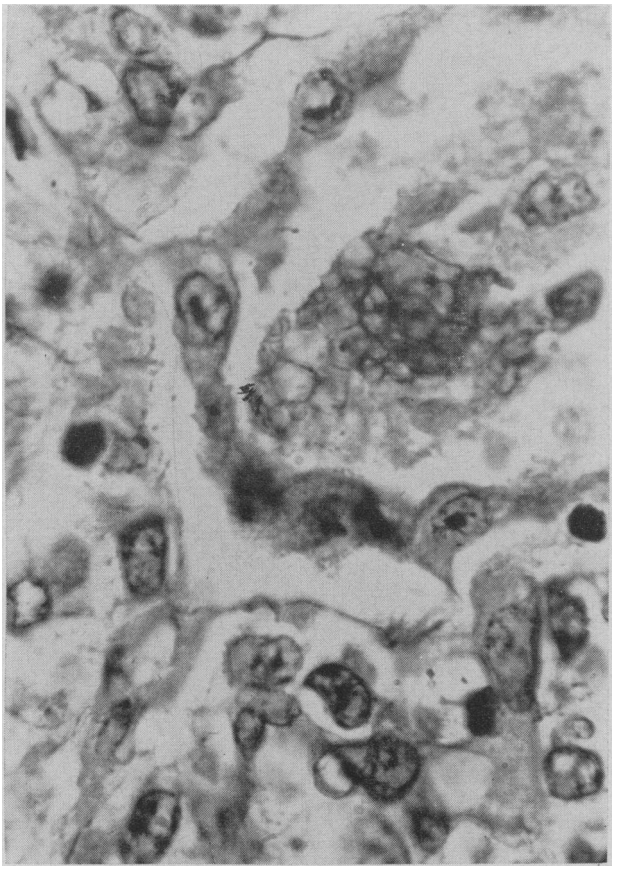

Fig. 2.—Histological section of lung showing typical foamy intra-alveolar exudate. Giemsa stain × 1150.

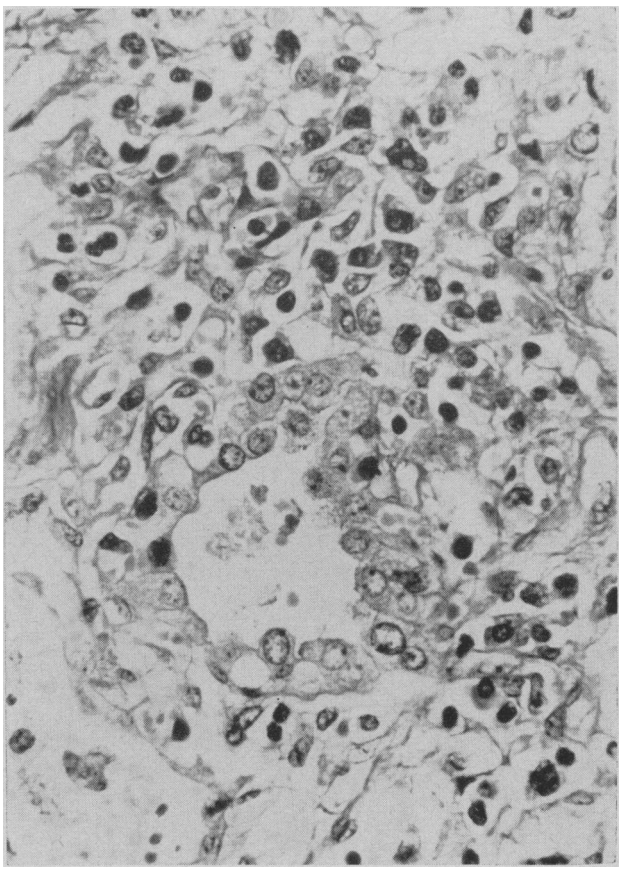

Fig. 3.—Histological section of lung showing alveolar lining hyperplasia. Giemsa stain × 460.

Autopsy Findings

The body was that of an extremely pale infant girl, four months of age, weighing 7 lb. The anterior fontanelle was widely patent and somewhat depressed. There was cyanosis of the lips and nailbeds, and several pale, circular, old scars were present over the anterior aspect of the chest and abdomen. The abnormal gross findings were confined to the thoracic cavity. There was no free fluid, no pleural adhesions and no evidence of pneumothorax. The right lung weighed 110 g. and the left lung 90 g. The pleural surfaces were of a deep reddish-blue colour showing slight mottling. There were no subpleural hæmorrhages or petechiæ. Both lungs felt indurated throughout all lobes. The bronchi contained some muco-purulent material. On section the lungs cut with the consistency of liver and showed no obvious areas of aeration. The cut surfaces showed a mottled appearance, some areas being greyish in colour while others were dark red or bluish. The lungs had the appearance of hepatic tissue. The right side of the heart was slightly distended with blood clot. There was no ventricular hypertrophy and the heart weighed 35 g. No cardiac congenital abnormality was found.

Microscopic Findings

On routine hæmatoxylin and eosin sections, the lungs showed a striking picture (Fig. 1). There was little similarity to normal lung tissue and the main features were the interstitial infiltration and the material present in the alveoli. The alveolar interstitial infiltrate was exclusively mononuclear and consisted predominantly of lymphocytes and plasma cells. Many of the alveoli appeared completely collapsed by the dense interstitial infiltrate, while most of the others showed varying degrees of distortion and compression. For the most part the alveoli were filled with a slightly eosinophilic fibrin-like material among which were moderate numbers of mononuclear cells, foamy macrophages and alveolar lining cells. The typical honeycombed material was extremely scanty in this case when compared with previous illustrations of this disease (Fig. 2). Occasional alveoli showed marked hyperplasia of their lining cells (Fig. 3). With the Giemsa stain the honeycombing became more apparent, and the typical punctate or angular, darkly staining protozoa could be identified (Fig. 4). The lungs were markedly hyperæmic but there was no evidence of bronchopneumonia. The liver showed a moderate fatty metamorphosis and several areas of superficial ulceration of the œsophagus were present.

DISCUSSION

It is not proposed to give a comprehensive report of the literature or the many aspects of this disease since they have been fully covered in recent papers. Recently two papers appeared in the same issue of this journal. Berdnikoff[1] reviewed the Canadian picture, and with the further case reported by Junger and Wyllie[2] there would appear to be 25 cases reported in Canada. The first report of this condition appeared in the United Kingdom literature in 1955,[3] and in the same year the first United States report was published,[4] followed by the first Canadian paper.[5]

Pneumocystis carinii pneumonia has been recognized for some time in Europe and many cases have

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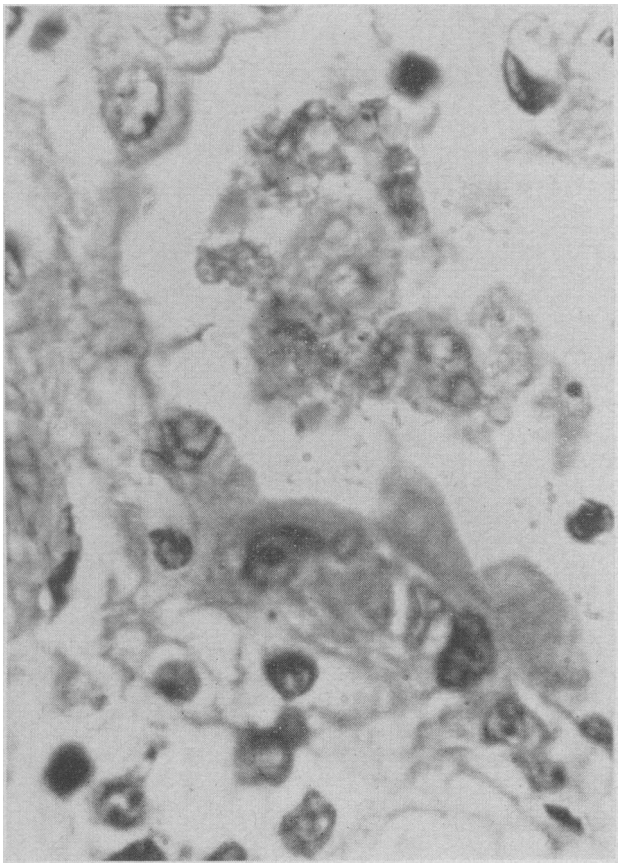

Fig. 4.—Histological section of lung showing alveolar "honeycombing" with scanty protozoa. Giemsa stain × 1150.

been reported from Switzerland, Germany, Austria, Yugoslavia, Czechoslovakia, Hungary, Italy and Scandinavia. Gajdusek[6] in a comprehensive review of the disease reports on his personal visit to a special isolation ward in Ljubljana, Yugoslavia, where 11 beds were set aside for infants with this disease. He reported that much of the time the ward was occupied, often full.

The etiological agent causing this disease is probably a protozoan parasite, although some consider it to be a fungus. The disease was first described in 1909 by Chagas in Brazil in the lungs of guinea pigs. Subsequently the organism has been found in the lungs of rabbits, rats, guinea pigs, dogs and monkeys.

The disease usually has its onset between the sixth and the sixteenth week of life, and has been reported in adults, usually associated with some chronic debilitating illness. The incubation period is thought to be about 40 days. The disease usually has a slow insidious onset, and tachypnœa and cyanosis of the lips and under the eyes may be the first indications of respiratory tract involvement. Physical findings are minimal and completely out of line with the degree of dyspnœa, sternal retraction, abdominal respirations, cyanosis and tachypnœa. Radiologically a diffuse ground-glass appearance with emphysema is the usual picture. Grossly the lungs are grey or greyish-pink and firm, and do not collapse when the chest is opened. Mediastinal emphysema or pneumothorax may be found. The heart may be slightly enlarged but other organs show no significant changes. Histologically there is an extensive mononuclear cellular infiltration with a striking absence of polymorphonuclear leukocytes. This infiltration fills the alveolar septa. Plasma cells are usually prominent in this infiltration. The alveoli are often lined by large cuboidal epithelial cells which are sometimes found lying free in the alveolar lumina. The characteristic feature of the disease is the foamy honeycombed material present in the alveoli amongst which small compact masses, one or two microns in diameter, are seen. These small masses are sometimes difficult to identify in hæmatoxylin and eosin sections, but can readily be identified by special stains, in this particular case using Giemsa stain. The condition has been associated with hypo- or agammaglobulinæmia and with cytomegalic inclusion disease. The mortality is in the region of 20 to 25%.

The epidemiological aspect of the disease is still not clear. It has been suggested that the agent passes to the newborn from an inapparent infection of the mother's genital tract; however, airborne dissemination seems most likely. It is possible that the organism is a saprophyte in human lungs and only under certain debilitating conditions such as cytomegalic inclusion disease, malnutrition, prematurity or malignant neoplasm is the organism able to become clinically manifest. In the present case the vermin-infested home raises the possibility of a rat or dog being the source of infection.

Laboratory aids do not materially help in the diagnosis of this condition. Lung puncture would be pathognomonic, but it is unlikely that this procedure would be carried out very often. Retropharyngeal swabs have been suggested as a means of diagnosis, but culture of the parasite on media or in experimental animals has not yet been developed as a diagnostic tool. Skin testing and complement fixation have been used with variable success. Therapy is purely supportive, and as yet none of the many antibiotics, steroids or antiprotozoal agents have proved their value, although antifungal agents are worth a trial.

SUMMARY

A further case of *Pneumocystis carinii* pneumonia diagnosed during life is reported. This case would appear to be the twenty-sixth reported in Canada. Attention is drawn to the paucity of organisms seen, and the scanty alveolar "honeycombing", even though the other microscopical features and gross and clinical findings were typical of the condition.

Our thanks are due to Dr. Jan Hoogstraten, pathologist, The Children's Hospital, Winnipeg, for his interest in this case, and to Dr. J. Stratton, the family physician who referred the patient to this hospital.

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