

HYPERTROPHIC PULMONARY OSTEO- ARTHROPATHY (BAMBERGER- MARIE DISEASE)

REPORT OF A CASE FOLLOWING LUNG ABSCESS *

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WITH ROENTGENOGRAMS BY

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In the surgical service of the U. S. Army Base Hospital at Fort Sam Houston, Texas, I became deeply interested in a case of draining lung abscess associated with enormous enlargement of the hands and feet, with ossifying periostitis of certain long bones and with fluid in the knee joints, evidently a case of subacute infectious osteo-arthritis.

REPORT OF CASE

Clinical Picture.—March 20, 1919, I found a young man having the appearance of a chronic invalid, but he was not confined to bed. He was rather pale and slightly emaciated. His facial expression was listless and dejected. Speech and mentality were rather slow. He had a chronic cough which occurred in paroxysms with the production of moderate amounts of thick, tenacious, mucopurulent sputum, at times streaked with blood, with a musty but not offensive odor. Most striking was the enlargement of the hands and feet, the hands resembling huge, clumsy paws. The wrists were enlarged after the manner of the hands. The fingers were generally thickened and sausage-shaped, but were not "clubbed"; indeed, the greatest enlargement was over the proximal interphalangeal joint. The finger nails were large, square and flat, smooth and without any of the curving seen in "simple clubbed fingers"; the large nails were only in proportion with the huge fingers. The elbows were slightly enlarged. The feet and toes were correspondingly enlarged, and there was moderate edema of the legs, with pitting on pressure. The knees were markedly swollen and contained fluid. There was slight tenderness to pressure over the

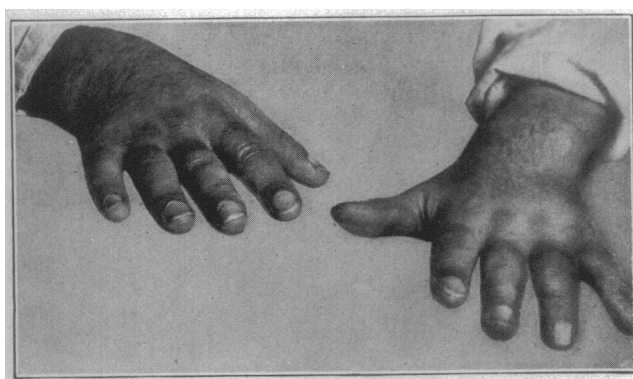


Fig. 1.—Condition of the hands.

affected bones and joints, but there was no redness. The bones of the skull and the face did not appear to be affected. From the surgical wound over the ninth rib in the posterior scapular line, there was slight purulent drainage, and examination revealed that the sinus communicated with a bronchus. In walking, the gait was clumsy and shuffling; the head was held slightly forward and the shoulders drooped, but no kyphosis nor scoliosis was detected.

* Owing to lack of space, this article is abbreviated in THE JOURNAL by the omission of certain paragraphs and illustrations. It will appear in full in the author's reprints.

History.—B. M. W., aged 24, first lieutenant, Flying Detachment Air Service, was admitted, Aug. 1, 1918, to the base hospital.

Family history: His father was alive, aged 54. He had had glycosuria for one year. His mother was alive and well, aged 43. All brothers and a sister were alive and well. There was no history of tuberculosis or cancer.

Personal history: The patient had had measles and mumps in infancy, and scarlatina at the age of 6. He gave a vague history of "rheumatism," saying that his feet had



Fig. 2.—Appearance of the feet of patient in November, 1918.

been swollen at times, but he was never confined to bed on this account. He had had occasional colds, but had never had tonsillitis until December, 1917. He had had no other illnesses. He stated that he had had no venereal disease.

Present illness: In December, 1917, while on duty with the Flying Corps, he "caught cold" and had tonsillitis. He was advised to have the tonsils removed.

Jan. 8, 1918, tonsillectomy was performed under ether anesthesia, in Philadelphia. He remained in the hospital only twenty-four hours. A few days later he developed a cough and pain in the left chest, at times referred to the left side of his abdomen.

February 1, he took to his bed and remained there for three weeks, suffering from what his physician called "pleurisy" of the left chest.

His cough persisted for about ten weeks.

March 19, he reported for duty at Kelly Field No. 2, Texas, and began flying. About this time his cough left him and he says that it did not return until after an accident in May in an aeroplane.

May 29, his aeroplane dropped from a height of 700 feet, but he was not seriously injured; he did not lose consciousness, but he was bruised and considerably shaken up. His chin was lacerated and his upper central incisor tooth was knocked out. This tooth was not found, and he wonders whether he may have "aspirated" it. There was no roentgenologic evidence that such was the case. No bones were fractured, and he remained in the hospital for only four days, reporting for duty on the sixth day after the accident.

His cough returned, and except for short remissions has persisted up to the present time. After his fall he was not again permitted to fly and was soon given a sick leave for a month.

About July 20, on his return from sick leave, he first noticed "swelling of his feet and toes"; his cough was troublesome, and he was sleepless and easily tired on exertion. The legs and feet felt heavy, weak and stiff as if they had been "jammed."

August 1, he entered the base hospital at Fort Sam Houston where he has remained up to the present time.

About August 15, the hands began to enlarge rapidly, attaining their maximum size (so far) within a couple of weeks. The paroxysmal cough, especially on lying down at night, caused violent frontal headache and insomnia. The headaches lasted only for a few weeks, and there have been none during the past six months; but the sleeplessness has persisted and is still a distressing feature despite the use of sedatives.

Clinical Course.—During August, there were slight rises of temperature during the afternoon, the maximum being 100 F., but usually about 99.4. The pulse ran between 98 and 108. There was much cough, sleeplessness, nervous irritability and depression of spirits.

Weight on admission was 133 pounds, which was about normal.

The physical and the roentgenologic examinations pointed to an involvement of the left lower lobe of the lungs. The heart was normal. The systolic blood pressure was 114. The enlargement of the hands and feet was as described above. The epitrochlear nodes were enlarged. Otherwise the physical examination was not significant.

Blood Wassermann tests were negative, August 6 and 9, and September 6.

The sputum was negative for tuberculosis in approximately twenty examinations.

Repeated examinations of the urine disclosed no albumin, no sugar and no casts. The specific gravity averaged from 1.009 to 1.020.

Blood examination, Aug. 2, 1918, revealed red blood cells, 4,740,000; hemoglobin, 70 per cent.; leukocytes, 11,200; polymorphonuclears, 72; large mononuclears, 3; small mononuclears, 25. There was no poikilocytosis nor chromatophilia.

The feces were negative for parasites.

In September, 1918, efforts were made to locate pus which was suspected in the left chest, and several unsuccessful aspirations were made.

September 13, a small amount of thick pus was aspirated from the left chest posteriorly at a depth which seemed to indicate a lung abscess, and at once costectomy of the eighth or ninth ribs in the posterior scapular line was performed and the parietal and the visceral pleura were sutured together, as the first stage of a two-stage operation for lung abscess. The operator was Major Russell.

September 17, through the costectomy wound, an abscess deep in the left lower lobe was opened with the cautery and a considerable amount of pus was evacuated and the abscess was drained.

From September, 1918, up to the present time, there has been more or less drainage of thick pus from the wound. Surgical solution of chlorinated soda (Dakin's solution) was not well borne on account of the communication with the bronchus.

At one time in February, 1919, the discharge gradually ceased and the wound closed but opened again spontaneously about sixteen days later, and discharged a bit of greenish, gangrenous tissue.

At the present time, there is still slight purulent drainage

and free communication with the bronchus, namely, a chronic bronchial fistula.

During these months whenever the drainage seemed insufficient there occurred an exacerbation of the cough, mental depression and above all an increase in the size and in the aching pains along the enlarged bones and joints of the extremities, but without local heat or redness.

In January, 1919, the knees became more swollen and filled with fluid.

The treatment during this time has been surgical care of the wound in the chest, symptomatic treatment for the pains and the insomnia, and during December, 1918, the injection of an autogenous vaccine prepared from the pus.

This vaccine contained *Staphylococcus albus*, and a gram-negative typhoid-like bacillus. No effect was noted from its use.

The sputum, March 4, 1919, was thick, mucopurulent, and streaked with blood. Culture shows *Micrococcus catarrhalis* and a green-producing nonhemolytic streptococcus. There was no elastic lung tissue. The odor was musty but not very offensive.

Blood examination, March 2, revealed: red blood cells, 5,740,000; hemoglobin, 80 per cent. (?); leukocytes, 11,400; polymorphonuclears, 54; small mononuclears, 39; large mononuclears, 4, and transitionals, 3.

The systolic blood pressure was 110; the diastolic, 64.

Fluid from the left knee was aspirated, March 6. Smear and culture were negative.

Ear and throat examinations were negative.

Eye examination, March 11, 1919, revealed: vision right eye 20/20, left eye, 20/20. The media were clear. The fundi were normal. The field of vision for form was normal in extent in both eyes.

The weight was 133 pounds Aug. 1, 1918, and 122 pounds, March 15, 1919, a net loss of 11 pounds.

The height was 64 inches, June, 1917, and 64½ inches, March 18, 1919.

The thyroid gland appeared to be normal.

There has been practically a normal temperature for months with occasional elevations to 100 F. at times when the drainage has been temporarily blocked.

The pulse still averages from 96 to 104.

Summary.—There was an involvement of the left lung, probably the effect of an aspiration infection following tonsillectomy under ether. There was a chronic productive cough. There was a rapid enlargement of the feet and hands during a period of six months. A lung abscess was drained, September, 1918, with a resulting persistent, bronchial fistula. Mental depression and insomnia were present. There was a slight loss of weight. There was no evidence of syphilis or tuberculosis. The pathologic condition in the enlarged hands and feet and other regions is best revealed by the roentgenograms which show, besides the enlargement of the overlying soft parts, a definite striated production of new bone in the periosteum of the shafts of the metacarpals and the first two rows of the phalanges, also along the lower ends of the radius and ulna. The

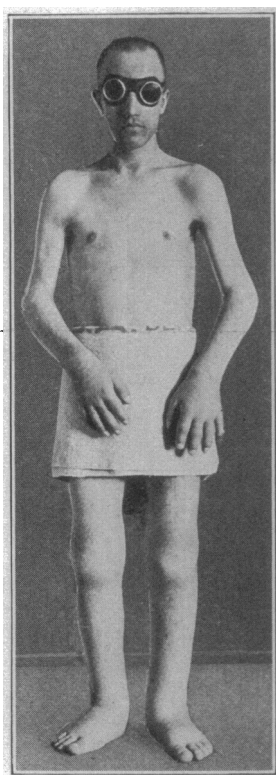


Fig. 3.—Condition of patient, March 20, 1919.

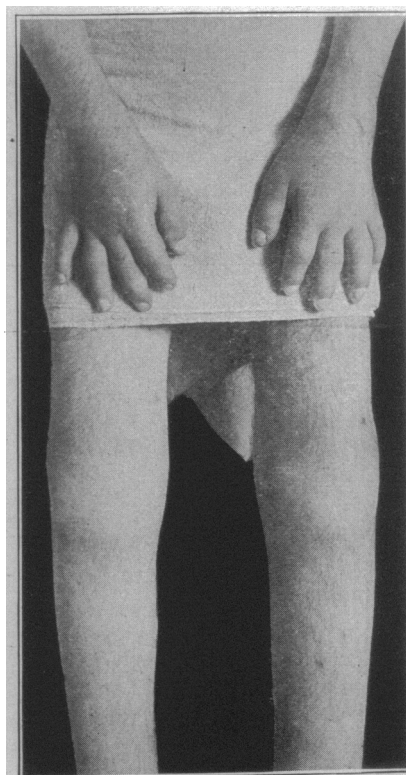


Fig. 4.—Appearance of hands and legs, March 20, 1919.

corresponding bones of the lower end of the femur; also the clavicles and some of the ribs, are to a degree affected. The joint surfaces appear unaffected. There is fluid in the knee joints.

Negative Findings.—The fingers are not "clubbed." No pituitary or thyroid changes are present. There are no

periosteal changes in the long bones, but we do not see the reverse.

In our case we cannot accept the last clause of Sternberg's statement, for we have a case with pronounced changes in the long bones and yet without "clubbing of the fingers," the fingers being very generally enlarged throughout and the nails, while proportionally large, not curved more than normal.

ETIOLOGY

The French writers were the first to attribute the origin of this rare disease to the absorption of toxins from a septic focus, usually in the lungs. This is the explanation now generally accepted; and in order of frequency the following foci have been encountered: bronchiectasis, empyema, abscess of the lung, pulmonary tuberculosis with cavity formation, toxic cirrhosis of the liver, and malignant tumors of the lung.

Sternberg mentions such other causes as pyelonephritis, dysentery, pneumonia, pleurisy, influenza, alcoholism and congenital heart disease; but Osler remarks that Sternberg must have included many doubtful cases. The tuber-

culosis theory is upheld by Poncet, Thorburn, Ball and Alamartine. Syphilis has also been advocated by certain French writers as the cause.



Fig. 5.—Thickening of the periosteum with irregular deposit of subperiosteal bone on metacarpals; also on proximal and middle phalanges (February, 1919).

headaches, disturbance of vision nor drowsiness; on the contrary, very marked insomnia. There are no changes in face, lips or skull. There is no evidence of abnormal thirst or hunger, and there are no alterations in the special senses.

HYPERTROPHIC PULMONARY OSTEO-ARTHROPATHY

In 1890, Marie first reported a case which he described as "osteo-arthropathie hypertrophique pneumique," a distinct clinical entity, and yet, according to Babcock, this very case turned out later to be a case of acromegaly.

I have been unable to make a complete review of the literature, stationed as I am on military duty, but all the available authorities have been consulted and many and great are the discrepancies encountered.

From many of the articles it would appear that the writer never had seen a genuine case of this rare disease and had been obliged to draw on and still further confuse the already confused descriptions of preceding authors.

It is doubtful whether the whole literature up to the present time contains records of 100 cases; it is certainly the only case of its kind which has ever come under my observation.

One is struck by the emphasis laid on the clubbing of the fingers, which is not present in our case. We agree very heartily with Sternberg when he says that "one must suspect that many cases, so-called, are not all the same, and it is a question whether or not 'clubbed fingers' and the periosteal changes are related." Sternberg says that we see clubbed fingers without



Fig. 6.—Subperiosteal bone deposits on lower ends of tibia and fibula; posterior aspect of os calcis; anterior surface of astragalus and of metatarsals (March, 1919).

Cyanosis, if it were constantly present, might be called on to explain simple clubbed fingers, but it fails utterly to account for the bone changes.

The bone and joint involvement in our case indicate conclusively that the process is a subacute infectious.

osteo-arthritis, and in our case, as in the vast majority of genuine cases reported, the focus of infection is in the lungs.

PATHOLOGY

In 1906, Alexander collected seventy-seven instances from the literature which he regarded as genuine cases of this disease, and he reviews the pathology of all the cases in which complete necropsies were performed (sixteen cases).

We quote Alexander's findings almost verbatim from Osler, and the description fits our case very perfectly:

Symmetrical deposits of new subperiosteal bone on the shafts of the long bones. Most frequently affected are the lower ends of the radius and ulna, the metacarpals and the first two rows of the phalanges; more rarely the lower end of the humerus and the upper ends of the radius and ulna. The new bone begins abruptly from 4 to 5 inches above the wrist joint and forms a sheath covering the lower ends as far as the epiphyseal line. The circumference of the bone is about equally affected, the thickest deposit being at the junction of the shaft with the lower extremity of the bone. The carpal bones are not affected, but the metacarpals are ensheathed and appear uniformly enlarged. The first two rows of the phalanges are similarly affected but less than the metacarpals. The terminal phalanges are apparently unaffected.

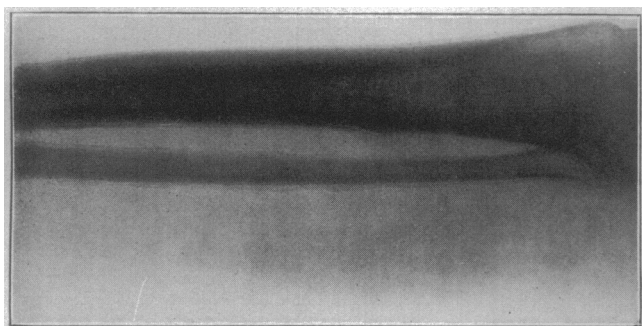


Fig. 9.—A sheath of new bone may be seen surrounding the shafts of the tibia and fibula. The cortex and medullary canal are apparently normal (March, 1919).

Similar bones of the lower extremity are affected, especially the lower tibia, which may be thickened by a layer of new bone one quarter of an inch thick. The phalanges of the toes are less affected than those of the fingers.

Deposits have been found on the iliac crests, on the clavicles and on the anterior surface of the patellae.

The symmetrical distribution is truly remarkable.

The cortex of the bone may show sclerosis and thickening, with diminution in the size of the medullary canal. The periosteum is very vascular and thick, and the main nutrient canals are large and many additional vessels enter the compact layer.

The bone changes are due to a chronic inflammation, causing new bone formation, also to atrophy and rarefaction and, to a much less degree, to osteophyte building.

The long bones do not bend. The skull is little, if any, affected.

In many cases there is excess of fluid in the joints, wrists, ankles, knees and fingers. In these affected joints the synovial membrane shows inflammatory changes, but there are no cartilaginous changes or lipping, no eburnation nor osteophyte building.

The bone changes are only part of the process, for there is much thickening of connective tissue, especially in the vicinity of the wrists, ankles and fingers.

Barker describes the affair as a slowly ossifying periostitis and osteitis, but he states that the joints are free.

Buck³ says that there is "an outside layer of dense, newly formed periosteal bone all along the shaft; this layer is striated and does not completely obscure the old cortical line. In pronounced cases all the bones in the body may be involved; the flat bones are thickened and there is marked kyphosis."

Fraenkel adds that it is "a true hypertrophy in consequence of periostitis, with new bone formation and at the same time a rarefaction of osseous tissue with dilatation of the natural interstices as well as increase of the medullary substance."

I make no attempt to harmonize the little discrepancies in the descriptions quoted above.

The thyroid and the pituitary glands are not altered.

TREATMENT

The treatment for the pains in the bones and joints is symptomatic. The only treatment which will logically have any effect on the progress of this rare disease must be directed toward the cure or improvement of the primary focus in the lungs.

CONCLUSIONS

The history, the clinical findings and the roentgenologic findings in our case all point quite conclusively to the diagnosis of hypertrophic pulmonary osteoarthropathy, by which we mean a subacute infectious osteo-arthritis, secondary to a septic focus in the lungs. However, we must acknowledge that the roentgenogram of the skull shows an abnormally small sella turcica, and the possibility of some change in the pituitary must be admitted.

Is it possible that our case, like the first case reported by Marie, may later develop into an irregular type of acromegaly?

CONGENITAL BILATERAL STRICTURE OF THE URETER*

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If we may judge from the literature on the subject, congenital stricture of the ureter is not rare; but a *bilateral* congenital stricture of the ureter is a rare defect, only eight cases, exclusive of the one here recorded, having been described. From the few cases recorded it would appear that these strictures are most often discovered at the extremes of life, most of them having become evident before the age of 5 years. Two cases have occurred in adults, one at the age of 25 years and the other at the age of 62.

No apparent reason for this congenital anomaly presents itself in studying the development of the urinary system. The ureters are primary derivatives of the bladder, growing cephalad to meet the metanephric tissue. Their lumina appear either as a result of central absorption of the epithelial outgrowth or are present primarily in the process of their evagination from the bladder¹. The failure of these processes to be completed would give rise to the obliteration of the lumen. Another explanation rests on an anatomic characteristic of the ureter in the embryo, namely, the presence of valves. In the case reported by Watson and Cun-

3. Bryant and Buck: American Practice of Surgery 1 and 3: 1906.

* From the Pathological Laboratory, Lincoln Hospital and Home, G. L. Rohdenberg, M.D., Director.

1. Huntington: Harvey Lectures, New York, 1906-1907, p. 222.