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SCHÜLLER-CHRISTIAN'S DISEASE

Volume XVII | Friday, December 14, 1945 | Number 9

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INDEX

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I.

UNIVERSITY OF MINNESOTA MEDICAL SCHOOL
CALENDAR OF EVENTS
Dec. 15 - Dec. 21, 1945
Medical Visitors Welcome

No. 94

Saturday, Dec. 15

9:00 - 9:50 Pediatrics Grand Rounds, I. McQuarrie and Staff; W-205 U. H.

9:15 - 10:20 Surgery-Roentgenology Conference; O. H. Wangensteen, L. G. Rigler, and Staff; Todd Amphitheater, U. H.

9:00 - 9:50 Medicine Case Presentation; C. J. Watson and Staff; M-515 U. H.

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221 U. H.

Sunday, Dec. 16

11:00 - 1:50 Obstetrics and Gynecology Grand Rounds; J. L. McKelvey and Staff; Station 44, U. H.

Monday, Dec. 17

9:00 - 9:50 Roentgenology-Medicine Conference; L. G. Rigler, C. J. Watson and Staff; Todd Amphitheater, U. H.

9:00 - 10:50 Obstetrics and Gynecology Conference; J. L. McKelvey and Staff; Interns Quarters, U. H.

12:15 - 1:15 Pediatrics Seminar; Irvine McQuarrie and Staff; 6th Floor Eustis.

12:15 - 1:15 Obstetrics and Gynecology Journal Club; M-435, U. H.

12:30 - 1:20 Pathology Seminar; Newer Cardio-Surgery; M. J. Shapiro; 104 I. A.

Tuesday, Dec. 18

9:00 - 9:50 Roentgenology-Pediatrics Conference; L. G. Rigler, I. McQuarrie and Staff; Eustis Amphitheater, U. H.

12:30 - 1:20 Pathology Conference; Autopsies; Pathology Staff; 104 I. A.

3:15 - 4:15 Gynecology Chart Conference; J. L. McKelvey and Staff; Station 54, U.H.

4:30 - 5:20 Ophthalmology Ward Rounds; Erling Hanson and Staff; E-534, U.H.

5:00 - 5:50 Roentgen Diagnosis Conference; Dr. Oscar Litschultz and Dr. Harry Mixer.

Wednesday, Dec. 19

8:00 - 8:50 Surgery Journal Club; O. H. Wangensteen and Staff; M-515 U. H.

9:00 - 10:30 Pediatrics Staff Rounds; W-205 U. H.

9:00 - 10:50 Neuropsychiatry Seminar; Staff; Station 60 Lounge, U. H.

11:00 - 11:50 Pathology-Medicine-Surgery Conference; Bronchogenic Carcinoma; E. T. Bell, C. J. Watson, O. H. Wangensteen and Staff; Todd Amphitheater, U. H.

12:30 - 1:20 Physiology Chemistry Journal Club; Staff; 116 M. H.

4:00 - 6:00 Medicine and Pediatrics Infectious Disease Rounds; W-205 U. H.

Thursday, Dec. 20

9:00 - 9:50 Medicine Case Presentation: C. J. Watson and Staff; Todd Amphitheater.

4:30 - 5:20 Ophthalmology Ward Rounds; Erling Hansen and Staff; E-534, U. H.

5:00 - 5:50 Roentgenology Seminar; Review of Recent Radiologic Literature; S. C. Peterson; M-515 U. H.

Friday, Dec. 21

9:00 - 9:50 Medicine Grand Rounds; C. J. Watson and Staff; Todd Amphitheater, U.H.

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221 U. H.

10:30 - 12:20 Otolaryngology Case Studies; L. R. Boies and Staff; Out-Patient Otolaryngology Department; U. H.

11:50 - 1:15 University of Minnesota Hospitals General Staff Meeting; New Powell Hall Addition Amphitheater.

1:00 - 2:00 Dermatologic Allergy - Dr. Stepan Epstein; W-312 U. H.

2:00 - 3:20 Dermatology and Syphilology; Presentation of Selected Cases of the Week; H. E. Michelson and Staff; W-312 U. H.

1:30 - 2:20 Roentgenology-Neurosurgery Conference; H. O. Peterson, W. T. Peyton, and Staff; Todd Amphitheater, U. H.

II. SCHÜLLER-CHRISTIAN'S DISEASE WITH PULMONARY INVOLVEMENT

Solveig M. Bergh

One of a group of diseases usually occurring in children in which lipoid substances are accumulated in cells of the reticulo-endothelial system is most commonly known as Schüller - Christian's disease or Hand-Schüller-Christian disease. The term xanthomatosis has been used to include the entire group which Sosman[18] has classified under five headings: Gaucher's disease, Nieman-Pick's disease, Schüller-Christian's disease, Xanthomas occurring in icterus diabetes and pregnancy, and those called essential xanthomatosis.

Epstein and Lorenz suggested the term lipoidosis for the group with a modifying term to indicate the type such as lipoidosis, Schüller-Christian type.

Essentially the process consists of an invasion of the reticulo-endothelial system by lipoid substances. It may be well, therefore, to review briefly the distribution of the reticulo-endothelial cells. They are very widespread in the body but are most common in the blood forming organs. They also are located in loose connective tissue; in the alveoli of the lungs; in glands of internal secretion such as the hypophysis, thymus, adrenal, thyroid, and parathyroid; in the heart; intestine; central nervous system; and in the ovary and testicle.

These cells have the power to become free ameboid cells and their chief function seems to be phagocytosis. They have an affinity for lipoid substances.

The type of lipoid deposited varies as well as its location. This also plays a part in the classification of the diseases.

Gaucher's disease affects chiefly the spleen, but the liver, bone marrow and lymph nodes may be involved. The reticular cells in these organs contain a cerebroside, Kerasin, which normally is present only in the myelinated nervous tissue. This disease is about twice as common in females as in males and has a familial tendency. It runs a chronic course, death usually resulting from some intercurrent infection.

Niemann-Pick's disease is a rapidly progressing, fatal disease affecting young children, especially Jewish infants. About 70% of the cases have occurred in females. In this disease the reticular cells show an accumulation of the phosphatides, chiefly lecithin. The spleen and liver are the common sites of predilection, but the bone marrow, smooth and striated muscle, epithelial cells of the thyroid and kidney, and the ganglion and glia cells of the nervous system may be involved as well.

We are especially interested in Schüller-Christian's disease. The lipoid involved in this disease is cholesterol which may be present in pure form or as an ester, combined with fatty acid. According to most biochemists the cholesterol level is maintained in the liver and the lungs. This view is supported by the fact that more cholesterol is contained in blood from the portal vein than from the hepatic veins, and also there is more in the pulmonary artery than in the pulmonary veins.

In 1924, Letterer described a case of a child who had acute reticulo-endotheliosis with little lipoid storage. A number of similar reports have appeared in the literature. Early death occurred in all of these cases. This condition is now known as Letterer-Siwe disease. Wallgren, in 1940, expressed the opinion that little lipoid is stored because of the rapidity of the course. Farber believes that this is a variant of Schüller-Christian's disease but runs a more acute course.

Schüller-Christian's disease was first described by Schüller in 1915 and elaborated upon by Christian in 1919 and Rowland in 1928. In 1893, Hand described a case showing osseous defects in the skull, exophthalmos, and polyuria which he attributed to tuberculosis but it most likely represented Schüller-Christian's disease.

The diagnostic triad reported by Christian consists of the map-like defects in the membranous bones of the skull,

diabetes insipidus, and exophthalmos. All of this triad is not necessary, however, to establish the diagnosis. Strong believes the skull defects are an integral part of the disease process but Sosman emphasizes that no one of the triad is essential. Only one or two of the clinical signs may be present. Involvement may occur in any part of the reticuloendothelial system in the body and such clinical signs as lymphadenopathy, hyperpyrexia, splenomegaly and hepatomegaly, pulmonary fibrosis, involvement of the long bones, and icterus may be found.

Etiology

The etiology is somewhat obscure although it is thought to be due to a disturbance in lipoid metabolism, especially cholesterol metabolism. Rowland believes that excess lipoids in the blood act as foreign bodies which irritate the blood vessel walls and thus cause perivascular cellular infiltration. As the vessel walls break, fusiform cells develop and fill with lipoid. When they are completely filled, more cells form and a tumor mass is produced. The older cells are pushed to the periphery while the newer ones lie close to the source of the lipoid. Diagnosis by biopsy at this stage is relatively easy, but later on fibrous tissue infiltration and foreign body giant cells mask the appearance, leading to an erroneous diagnosis such as sarcoma, giant cell tumor, fibroma, multiple myeloma and so forth.

Pathology

Discrete nodules or a diffuse infiltration of granulomatous tissue which are firm in consistency and may have a yellow coloration are demonstrated in various sites. These include the bones of the skull, dura, mandible, heart, lungs, pleura, ribs, vertebrae, long bones, spleen, liver and lymph nodes.

The xanthoma cell or "foam" cell is the most characteristic feature of the infiltration. The usual cellular arrangement shows the more mature cells toward the periphery of the lesion while the smaller mononuclear cells occupy the center of the lesion. As the lesion develops there is some tendency toward fibrosis. The cells which acted as macrophages are converted into fibroblasts so the tissue develops a nodular character with fibrous tissue, giant cells, fibroblasts, and lipoid deposits which are both free and intracellular. The granulation tissue can spread easily in areolar tissue and perivascular spaces. An apparent infiltration of an organ actually is an extension along perivascular channels.

Pathologists have been aware of xanthomatous changes in the lungs for some time but this fact has not been noticed clinically. Bell[2] states that in the early stages of the disease, xanthoma cells are seen in the alveoli and in the septa which are increased. He also states that in late stages an extensive fibrosis is noted which may cause right heart failure.

Clinical Picture

The age incidence shows this disease to be primarily one of the first decade and it is about twice as common in males as it is in females.

The onset is insidious and the disease is usually far advanced before a physician is consulted.

Symptoms are dependent upon the location of the lesions and the mechanical effects resulting from pressure and bone destruction. In order of frequency, the symptoms are: skull defects, diabetes insipidus, exaophthalmos, next a combination of these three, and then involvement of the pelvic bones, skin lesions, arrest in growth, and others of less frequency. There usually is a lack of any pulmonary symptoms unless the disease is very far advanced.

Children who have this disease usually are underweight and poorly developed. They usually are irritable and cross as well. The head may be deformed by soft subcutaneous swellings or defects may be felt in the skull.

Polydypsia and polyuria occur when xanthomatous lesions within the skull either invade the pituitary gland or cause pressure upon it.

Exophthalmos occurs when the granulation tissue extends anteriorly through the superior orbital fissure and optic foramen into the retrobulbar space.

Loosening of the teeth may occur when lesions are present in the alveolar processes.

Usually an arrest in growth occurs with the onset of diabetes insipidus. About 30% of the cases show skin lesions. These usually are manifest as small fatty lesions with crusts and are diffuse over the entire body.

In late stages, the children may develop respiratory difficulties. X-ray examinations of the chest may show increased density of the lungs which is diffuse but most marked near the hilus, or small, multiple areas of density resembling metastases or miliary tuberculosis. The increased induration of the perivascular spaces in the lung may cause resistance to blood flow and thus hypertrophy of the right side of the heart may occur.

Prognosis

Schuller-Christian's disease usually is considered to be a chronic disease with a slow and irregular course. The prognosis depends somewhat upon the stage in which the patient is seen and therapy instituted. If the disease is recognized early, the prognosis is relatively good. However, in cases which are unrecognized or those which fail to respond to treatment, death may be due to intercurrent infection or heart failure.

Treatment

Insulin is of some benefit in that it stimulates the appetite and thus favorably influences the weight and strength of the patient. Intranasal or hypodermic administration of pituitary extract may control the diabetes insipidus. Endocrine therapy, otherwise, is not beneficial.

Radiation therapy is the only known means of effectively treating the bone lesions. A medium dosage, about 400 r to 600 r in air is given to local areas of involvement. This amount of radiation usually will arrest the process, and in many cases cause filling in of the bony defect. Lesions, however, may appear in other regions of the body.

Heavy doses are not desirable because of the possibility of causing an arrest in growth. There also may be harmful effects on surrounding normal tissue such as the brain.

Diabetes insipidus has shown a favorable response to irradiation also, but exophthalmos is not reduced.

Xanthomatosis of the Lungs

Although well known to pathologists, changes in the lungs as a result of Schuller-Christian's disease have not been recognized clinically probably because symptoms were not produced. The observation of rather destructive changes in the roentgenogram of the lungs in a patient with this disease directed our attention to the possibility that pulmonary involvement in this disease might not be uncommon.

An increase above the normal in the shadows produced by the bronchovascular lymphatic systems of the lungs may be observed in a wide variety of conditions. Such changes are often nondescript and nonspecific but in many cases they are the result of chronic bronchitis, asthma, non-specific emphysema, interstitial pneumonitis, sarcoidosis, infiltrating carcinoma, and miliary tuberculosis. More recently it has been shown that changes in the lungs associated with cystic fibrosis of the pancreas may produce a similar picture. In tuberous sclerosis likewise pulmonary infiltrations have been observed which produce similar roentgenologic findings. To this long list we wish to add Schuller-Christian's disease.

In many cases, a differential diagnosis cannot be made by roentgenogram alone, but clinical findings and the associated lung findings together will lead to the correct diagnosis.

Emphysema frequently plays a part in many of these conditions. The lungs are hyperaerated and appear darker than

normal with the usual exposure. The blood vessels and bronchi thus appear very sharp in contrast with the dark lung fields. In addition, in asthma, chronic bronchitis, etc., there is thickening of the peribronchial, perivascular, and interstitial connective tissue which is evidenced on the roentgenogram as a fine network distributed throughout both pulmonary fields. If the increase in connective tissue becomes marked, the capillaries are compressed sufficiently to cause obstruction to blood flow. This leads to widening and thickening of the pulmonary arteries, enlargement of the pulmonary conus, and finally cardiac enlargement.

Miliary densities diffusely spread throughout the lungs may be present in miliary tuberculosis, bronchitis, emphysema, pneumonitis, etc. The densities are minute in size, usually fairly uniform but they may be slightly larger in the upper lobes. Enlarged mediastinal nodes frequently are present in miliary tuberculosis.

Recently a case of Schuller-Christian's disease was reported in which findings similar to miliary tuberculosis were present in the lung. The condition persisted for years so the possibility of miliary tuberculosis was ruled out. At postmortem examination the lungs showed distended alveoli, increased connective tissue and thickened alveolar septa resulting from the xanthomatous infiltration.

We have collected five cases of Schuller-Christian's disease showing pulmonary involvement and four of these will be presented here in detail.

Case Reports

1. ____, male, 3 years.

History

Born 8-20-40, weight 8#. Full term pregnancy, instrument delivery. He apparently was in perfect health until he was 9 months of age. At that time he developed a seborrhea on the trunk which spread to the head and the rest of the body. Ordinary symptomatic methods of therapy used did not improve the lesions.

On 3-10-43 he was hospitalized elsewhere. Coal tar ointment was applied to the scalp, sulfathiazole ointment to infected areas, and ultra-violet light treatments were given. There was improvement, but the skin lesions did not entirely disappear.

While hospitalized, about 2½ weeks before admission here, he developed an infection under the left thumb nail. This spread to all the fingers of both hands. There was drainage of yellow material with a foul odor. He also developed a gum infection and a generalized lymphadenopathy.

Several months before admission--exact date unknown--an exophthalmos of the left eye was noticed.

Physical examination

Temperature, pulse, and respirations: normal.

Head:
Scalp: Covered with a seborrheic crust with a few pustules.
Occipital nodes: Large and tender.
Eyes: Moderate exophthalmos on left.
Ears: Purulent fetid discharge, bilateral.

Neck: Submaxillary, anterior and posterior cervical, and the posterior auricular lymph nodes enlarged, hard, non-tender.

Chest: Normal to physical examination.

Abdomen: Liver palpable 2 cm. below costal margin.
Spleen palpable 1 cm. below costal margin.

Extremities: Enlarged, hard, non-tender axillary and inguinal nodes.

Laboratory Findings

Urine: Negative. Hgb.: 9 grams.
Rbc: 3,800,000; Wbc: 19,750,
87% prm's, 13% lymphocytes.
Blood Cholesterol: 189 mgms %.

Plasma protein: 6.1 gms. %.
B.U.N.: 5 mgm%. Blood Wasserman: Negatime. Sed. rate: 11 mm. in 1 hr.
Schick test: Negative.
Mantoux test: Negative.

X-ray Findings

Skull: large defect in the left supra-orbidal fissure and characteristic defects in both mastoids, and at the symphysis of the mandible.

Pelvis: Negative.
Chest: 5-17-43. Small miliary lesions scattered throughout both lung fields but more marked on the left. It was reported that the exact etiology was not apparent and reexamination was requested. This was done a few days later. In addition to the miliary densities, enlargement of the hilar glands was noted bilaterally.

Another chest film was taken shortly before the patient expired. This showed no essential change in the fine mottled densities. The diagnoses suggested at this time were interstitial fibrosis or infiltration with Schuller-Christian's disease.

Biopsy: Left cervical lymph node: Enormous reticulum hyperplasia destroying the architecture of the node. Consistent with Schüller-Christian's disease.

Treatment: X-ray therapy
7 treatments in 9 days 5-25-43 to 6-2-43.
Left mastoid--800 r in air.
Left orbit--500 r in air.
Factors used were 220 K.V., 50 cm. distance, .5 mm. Cu. filter, H.V.L. 1.35.

He also received 7 treatments to the cervical areas in 15 days 9-24-43 to 10-8-43 using the same factors. Total dosage:
Right lateral cervical--600 r in air.
Left lateral cervical--400 r in air.
Right anterior oblique cervical--200 r in air.
Left anterior oblique cervical--150 r in air.
The right supraclavicular area received 750 r in 4 treatments over 8 days, 10-11-43 to 10-18-43.

In addition, he received some superficial therapy to both inguinal regions. Four treatments were were given in twenty days 8-20-43 to 9-4-43. A total of 300 r in air was given using 140 K.V., 30 cm. distance, 2 mm. al filter, with a H.V.L. of .17 mm. Cu.

To skin lesions: 5% sulfathiazole ointment in aquaphor.

Course

This patient was carefully followed in the outpatient department and was admitted to the hospital on 5 different occasions. These were for treatment of various fluctuating lymph nodes.

Patient was last admitted on 12-22-43 with severe abscesses in both groins with practically a beginning gangrene of the tissues around the groin. There was very foul, purulent and fatty discharge. Patient's condition became critical on the evening of admission and he expired on the following day.

Postmortem Examination

Gangrenous lymph nodes in both inguinal regions were the immediate cause of death. A necrotic defect 3x7 cm. in right inguinal. Another 4 cm. in diameter in left inguinal region.

Lungs: Left--240 grams, right--200 grams. "Both appear somewhat fibrotic on cut section. Definite emphysematous blebs up to 2 mm. in diameter are present throughout."

Microscopic: "The interalveolar tissue is enormously increased consisting of fibrous connective tissue and lipid-filled giant reticulo-endothelial cells. The alveoli show epithelialization in the larger blebs. Macrophages are found within them. The pleura, too, is enormously thickened."

In addition, there was extensive xanthomatous infiltration in the spleen, lymph nodes, periadrenal fat, and sternal marrow.

Comment

A typical case of Schuller' Christian's disease without pulmonary symptoms but with the characteristic roentgen findings is here presented. It should be noted that physical signs were absent. The autopsy confirmed the roentgen evidences of pulmonary involvement in addition to the usual findings of xanthomatosis elsewhere. Study of the microscopic sections of the lungs explains well the roentgen findings since perivascular and alveolar infiltrations of small size but widely scattered were present together with emphysema of fairly marked degree.

Case 2.

____, male, 2 yrs. 10 mos.
Admitted 1-5-43, expired 1-30-45.

P.C.: Exophthalmos--6 months.
Anorexia--2 months.
Polydipsia--2 months.
Polyuria--2 months.
Listlessness.
Limp and disability of left leg.
Waxy, yellow appearance of skin.
Dyspnea--one week.

History

Born 4-9-40, full term--6# normal delivery, breast fed--6 weeks, developed normally, walked at 9 months. Bronchitis at 6 months, occasional colds, no contagious diseases.

Physical Examination

Revealed an acutely ill child. The skin was waxy and yellow and patient was edematous.

Head: Asymmetrical; defect in left parietal region; defect above left orbit.
Eyes: Bilateral exophthalmos, more marked on the right.
Ears: Foul smelling discharge from left ear which was found to be coming from just anterior to the drum. Ear drums normal.
Throat: Mucous membranes pale; tonsils enlarged.
Chest: Breathing labored; breath sounds accentuated; no rales; heart--systolic murmur at apex.
Abdomen: Liver 4 cm. below costal margin; mass in L.U.Q. 4 cm. below costal margin, thought to be spleen.
Extremities: A few petechiae on legs; slight pitting edema.

Laboratory Work:

Urine: Negative.
Blood: Hgb: 1.3 grams.
Rbc: 800,000.
Wbc: 14,600 pmn's 21%, lymphocytes 73%, normoblasts 6%.
Cholesterol: 107.
Serum proteins: 4.2
Albumin: 2.1
Globulin: 1.7
Bleeding time: 24 minutes.
Clotting time: 3 min. 41 sec.
Wassermann: Negative.

X-ray Findings

Chest: (1-5-43) Prominent lung markings, destructive areas in several ribs, and a lesion of the left scapula.

Skull: Multiple large, irregular, sharply defined areas of destruction involving the calvarium. Wings of the sphenoid and roofs of the orbits also were involved. The medial end of the right petrous tip showed destruction, as well as areas in the mandible.

Pelvis: Extensive areas of destruction in left ilium, right ilium, right ischium and pubis.

Long bones:
Left femur: Small area in proximal end, large area distal end.
Right femur: Area in distal end.
Later chest film (1-20-43) showed areas of density radiating from the hila into the upper lobes but more marked on the right.

Biopsy: Bone marrow aspiration showed hypoplasia of the marrow with some large reticulo-endothelial cells measuring 30

to 60 microns. The cytoplasm was very pale and was filled with lipoid. Conclusion--Schuller-Christian's disease.

Treatment

Multiple transfusions were given with a total of 3000 cc. of whole blood. Hemoglobin ranged from 6.5 to 9 grams.

X-ray therapy was started in 1-15-43. He was given eight treatments to the skull in eleven days using 220 K.V., .5 mm. Cu. filter, 50 cm. distance, with a H.V.L. of 1.35.

Total dosage:
Right lateral skull--217 r
Left lateral skull--200 r
Superior skull--100 r
Anterior skull--200 r
Posterior skull--100 r

Autopsy: Autopsy showed a grade III jaundice with petechiae over the entire body. Exophthalmos was very marked and bony defects could be palpated in the skull. Abdomen distended, liver and spleen palpable.

Pleural cavities contained a small amount of straw colored fluid.

Right lung, 170 grams; left lung, 150 grams. "Congestion is minimal and some fibrosis is apparent."

Microscopic--"The visceral pleura is somewhat thickened. Immediately beneath it there is a sheath of xanthomatous tissue with rich capillary blood supply. Xanthomatous infiltrate is present throughout the lungs, more marked about the blood vessels and bronchial tree. The alveolar walls are thickened, and many of the alveoli contain lipid-filled macrophages."

In addition, xanthomatous tissue was found in the epicardial fat, lymph nodes, liver, thymus, bone marrow, kidney pelvis, choroid of the eye, retrobulbar tissue, interstitial connective tissue of the pancreas, skin, gallbladder, dura mater, peri-adrenal tissue, and in the pituitary gland.

Comment

Here again the lung findings were confirmed by the changes found at autopsy. It should be noted that some dyspnea was present but few physical signs were manifested.

Case 3.

, male, 5 yrs.

History

This patient was first seen in the clinic on 6-10-43. Presenting complaint was pain in the right groin for 2 days after stepping into a hole and hitting the right inguinal region in the fall. Pain was aggravated by motion. Fracture was suspected and patient was sent for an x-ray examination of the pelvis. This revealed an extensive defect in the right ilium above the acetabulum and extending into the acetabulum itself. Xanthomatosis was suggested and films of the skull requested.

The patient was admitted and more careful examination revealed:

Head: Several soft, tender spots palpable where there apparently were defects in the bone. These varied from $\frac{1}{2}$ inch to $1\frac{1}{2}$ inches.

Eyes: Medial strabismus; no apparent exophthalmos.
E.N.T.: Negative.
Neck: Small lymph nodes palpable in cervical chains but none markedly enlarged.
Chest: Marked funnel chest. Lungs clear.
Abdomen: Liver palpable 3 F.B. below costal margin.
Marked tenderness over right ilium and trochanter.
P.H. -- Full term baby weighing $9\frac{1}{2}$ pounds, delivered by Caesarean section.

Treatment:
X-ray therapy:
Right lateral skull--400 r,

½ mm. Cu.
Left lateral skull--400 r, ½ mm. Cu.
Anterior right hip--400 r, 1 mm. Cu.
Posterior right hip--400 r, 1 mm.Cu.

Given from 6-17-45 to 6-23-43, using 220 K.V., 60 cm. distance, 1.35 H.V.L.

The patient was discharged and was followed in the outpatient clinic. In August of 1943, he was found to have a 2 + Mantoux test so a chest film was ordered. This showed a diffuse increase in the bronchovascular markings suggestive of peribronchial or perivascular infiltration. Later chest films have also shown the same process in the lungs, and in addition some enlargement of the heart suggesting the possibility of involvement of the cardiac muscles.

The patient was seen at frequent intervals and he had no complaints until June, 1944. Then there was an erosion at the base of the lower right first bicuspid. He received 800 r in air to this area in 4 treatments over a 10 day period.

Since then, patient has been doing very well, is active, and he is attending school.

In this case the accidental presence of a positive skin tuberculin test led to the roentgen examination of the lungs with the resultant diagnosis of pulmonary xanthomatosis. Although there has been no confirmation by microscopic examination, the findings are so characteristic there seems oittle doubt of the validity of the observation. Here again the absence of pulmonary symptoms or physical signs should be noted.

Case 4.

___, male, 6 yrs.

This patient was first seen 1-9-43. Presenting complaint was polydipsia and polyuria for 1 month.

History

Born 1-4-37, wt. 10½#. Forceps delivery. Cyanotic for ½ hour. Difficult to start respiration. Normal development.

Previous diseases:
Measles--4 years.
Whooping cough--5 years.
Osteomyelitis left tibia-- 5 yrs.

The left tibia drained for six months and then healed.

Physical Examination

Essentially negative except for scar 8-10 cm. long on anterior surface of upper 1/3 of left tibia.

Laboratory Findings

Urine: Specific gravity 1000 otherwise negative.

Blood:
Hgb.: 116
Wbc: 8,100, pmn's 58%, lymphocytes 42%.
Cholesterol: 140 mgms. %.
Wassermann: Negative.
Mantoux: Negative.

X-ray Findings

Skull: Single small defect in right parietal area.

Five months later the patient developed a lesion in the left ischium just below the acetabulum.

Chest: Roentgenograms all show some increase in bronchovascular markings suggestive of diffuse infiltration. While hospitalized, the patient developed a right lower lobe pneumonia but this resolved with treatment.

Treatment

The patient received pitrossin intramuscularly at first, and later intranasally. He was given sulfadiazine for the pneumonia. X-ray therapy was used for the skull lesion and the left hip, 800 r to right parietal lesion using 140 K.V. To the left hip he received 400 r posteriorly in 4 treatments using 200 K.V., 60 cm. distance, ½ to

1 mm. Cu. filter.

Course

After completion of x-ray therapy, the patient was discharged from the hospital and followed in the outpatient clinic. Progress was satisfactory until May, 1944, when a small mass was noted on the forehead. Skull films showed a lesion in the frontal bone. X-ray therapy again was given and he received a total of 800 r in 4 treatments over 6 days, 5-23-44 to 5-29-44, using 220 K.V., 50 cm. distance, ½mm. Cu. filter with a H.V.L. of 1.35.

The patient has been active and seemed perfectly well until October, 1945. He then complained of some pain in the back. Films at this time showed marked collapse of the body of the sixth thoracic vertebra. X-ray therapy was instituted. 4 treatments in 8 days, 10-8-45 to 10-15-45, were given to the area totalling 800 r using 220 K.V., 60 cm. distance, ½ mm. Cu. filter.

Before he was discharged from the hospital he was placed in a short body cast to prevent anterior flexion. Patient has not been seen in clinic but has an appointment this month.

Discussion

We have been chiefly interested in the pulmonary changes in Schuller-Christian's disease. In both of the cases which have had postmortem examinations, positive findings were present in the lungs. These consisted of an increase of interalveolar tissue consisting of fibrous tissue and lipid-filled reticulo-endothelial cells, emphysematous blebs, lipid-filled macrophages in the alveoli, and thickening of the pleura.

Changes in the lungs seen on x-ray examination consist of increased bronchovascular markings and exaggerated root shadows. More severe cases may show miliary densities throughout both lung fields which may resemble miliary tuberculosis.

Many cases of Schuller-Christian's disease have been reported in the literature, but until recently very little has been written about pulmonary involvement.

Rowland reported a case of a boy 3 years of age whose lungs on x-ray examination showed a diffuse bilateral fibrosis. There was cardiac enlargement as well. Before death, he had dyspnea, a severe cough, cyanosis and edema. Cause of death was due to impaired circulation from extensive pulmonary fibrosis. Postmortem examination revealed vesicular emphysematous cavities varying in size from a pin head to a pea, the septa between these cavities were fibrous and inelastic with hyperplasia and lipoidosis of the reticulo-endothelial cells. There was xanthomatous infiltration into the lung and some fibrotic nodules were present in the parenchyma as well as an increase in connective tissue around the vessels and bronchi and in the interlobar septa.

Still another case was that of a 2 year old girl presented by Mallory.[14] Chest films showed a moderate amount of diffuse mottled peribronchial infiltration. This became more severe, then remained the same for 3 months when the lung fields became emphysematous. Hampton stated that if he had had only a single film of the chest, the most probable diagnosis would have been miliary tuberculosis in its terminal stage. Clinical diagnosis in this case was chronic non-specific pulmonary fibrosis. There was generalized cardiac enlargement.

A similar case in a 9 year old boy was reported by Thompson, Keegan, and Dunn.[20] A roentgenogram of the chest showed a diffuse increase in density as in chronic interstitial pneumonitis. This boy died suddenly from cardiac failure due to impaired circulation caused by the extensive pulmonary involvement. Microscopic examination showed extreme interstitial fibrosis with a moderate mononuclear cell infiltration.

Merritt and Paige[15] reported pulmonary involvement in a boy 3 years of age. The chest film showed an increase in the

bronchovascular markings with increased density near the hilar regions. Autopsy showed thickening of the pleura which was replaced by granulomatous tissue with mononuclear and foam cells.

In these cases as well as ours, there was a lack of pulmonary symptoms. For this reason, we wish to impress upon the clinicians the importance of examination of the chest by the x-ray method in spite of lack of symptoms or physical signs.

It is probable that the apparent increase in bronchovascular markings and the so-called pulmonary fibrosis really represents granulomatous infiltration but the ultimate result probably is pulmonary fibrosis.

Summary

1. A review of Schuller-Christian's disease has been presented.

2. Four cases are reported in two of which autopsy findings are available to illustrate pulmonary involvement in this disease.

3. The changes in the lungs previously known to pathologists but concerning which clinicians apparently have been unaware have been discussed. The x-ray findings are detailed and the pathology of the lungs which produces such findings is described.

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III. GOSSIP

Today is the last General Staff Meeting until after the Christmas holidays, when meetings will be resumed, January 11. Christmas festivities start in the hospital Wednesday, December 19 when the Traffic Club trims the tree; their regular Christmas luncheon will be held in their quarters Thursday, Dec. 20; that evening, the gifts will be wrapped. The program for the children will be held Monday, December 24 in the afternoon. This club, composed of railroad men and women and shippers, has taken care of us at Christmas time for nearly 20 years. Our children have been visited by Santa Claus' helpers, assisted by Dorothy Jones to learn the gifts they would like to have. Each child will receive 2, and the other patients will be given an attractive fruit basket. Santa Claus is not feeling too well this year, and after many years, his place will be taken by a substitute. Christmas morning the Traffic Club have arranged for carols. The North High Choir have sung for us for many years and they will be back. The Staff Christmas Party will be held in the new dining rooms, Friday, Dec. 21, from 2:30 to 5:00 P.M. Every staff man and employee of the hospital is urged to attend. Refreshments will be served. The nurses' party and several departmental parties will also be held during the season. The University of Minnesota Hospitals staff and patients are grateful to the Traffic Club for the fine spirit which they display to us. Years ago when we were adopted by them, they apparently made up their minds to put on the best hospital Christmas celebration in the Twin Cities, and they realized their ambition year after year....Staff meetings this fall have reached new highs in attendance and interest. It has been difficult to receive a steady supply of bulletins for mailing, but we hope this can be corrected in the coming year. The return of so many men who have been in the service has added greatly to the scientific value of these sessions. We have much to be thankful for this year, for even if the war is not officially over, things are beginning to look up....The Continuation Course in medicine will start January 7 at 8:30 a.m. Registration and orientation sessions will be held January 4 and 5 at the Center for Continuation Study. Visitors for the first week will be W. Barry Wood, St. Louis, Duckett Jones, Boston, and Francis Rackeman, also of Boston. At the same time, the course in the Basic Sciences will start. Both groups will be registered and given orientation at the same time. Priority has been allotted to University of Minnesota graduates, residents of Minnesota, and states in our area without medical schools. Courses are also being offered at Wisconsin, Marquette, Iowa, Nebraska and Creighton, which will help to ease the situation. Maximum registration in both groups has been fixed at 40. Weekly programs will be issued so that the interested Staff Members may attend. Graduate students are welcome, without the payment of extra fees. The special course in otolaryngology will start January 14. Visiting staff members will include Frederick T. Hill, Waterville, Maine; Arthur W. Proetz, St. Louis, Missouri; Louis H. Clerf, Philadelphia; Julius Lempert, New York City; Members of the staff of the Medical School and Mayo Foundation, and others will serve. Course in Hospital Administration will start January 21 and last five days. It will deal with "Recent Developments in Social Welfare, Public Health, Medical Service, and Hospital Service". A one month course in Pathology of Diseases of the Skin will be offered by Professor Pincus, starting January 21. This has been arranged for dermatologists and graduate students. The extra demands which will be made upon the staff during the next six months will receive adequate compensation in the gratitude of the many men returning from the services who receive this special instruction. Most of the residencies now occupied by men on inactive military status will be vacated by July 1. The present crop of residents and interns will be back for assistance with their educational programs as soon as the older veterans are cared for. The demands for training after graduation will continue and the Foundations are interested in those universities planning permanent programs in this field. One reason Minnesota has been so generously provided for by the Commonwealth Fund and the Kellogg Foundation is because of our attitude toward this training....The editor extends to all best wishes for a pleasant holiday season and the best of everything for the New Year.....