

GENERAL STAFF MEETING
UNIVERSITY HOSPITALS
UNIVERSITY OF MINNESOTA



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I. CATHOLICS AND AUTOPSIES

In Hospital Progress for April 1931, Sister M. Patricia, writing on "Autopsies - How Obtained," concludes with a beautiful quotation from The Spirit of St. Francis de Sales, by Jean Pierre Camus.

"Since our blessed Father was not, like the martyrs, privileged to offer his body, both by living and dying, as a victim for God, he found out, with the ingenuity of love, a method of self humiliation and self-sacrifice to be carried out after his death. When quite young and still pursuing his studies at Padua, following a dangerous illness, he begged his tutor to see that when he was dead his body should be given into the hands of surgeons for dissection. 'Having been of so little use to my neighbor in life, I shall thus at least after my death be able to render him some small service.' "

Report of Committee on Autopsies 1931. American Hospital Association.

II. NEUROFIBROMATOSIS (VON RECKLINGHAUSEN'S DISEASE) Abstr. Pearson.

Abstract: Potter, P. C. and McWhorter, J. E., Ann. Surg. XC, 397-402 (Sept.) 29.

Report of single case showing most of the features of the complete picture of Von Recklinghausen's disease.

Checks indicate features present:

Congenital	X
Inherited (at least 20%?)	?
Subcutaneous tumors	X
Visceral tumors	X
Pigmentary anomalies	X
Painless lumps	X
Nerve pain	?
Deformity (elephantiasis-like) of extremity	X
Bone changes (extremities)	X
Scoliosis, kyphosis, etc.	X
Asymmetry of head, other anomalies	X
Psychic disturbances (often mental deficiency)	X
Eye changes	?
Ear involvement	?

Gradual and persistent development of signs and symptoms	X
Invasion of central nervous system	X
Prominent intrathoracic tumor	0
Frequent failure to show neurogenic origin of fibroma in microscopical sections	X
Malignant degeneration of one tumor	X
Unsuccessful surgical removal	X
Recurrence of growth	X
Metastases to lungs	X
Tendency for fatal cases to occur in young adults	X

The only feature in common to all forms of the disease is a fibroma (1 to thousands) of, attached to or near a nerve. Any or all of the other signs may be present.

Illustrative cases from our institution follow.

III. CASE REPORT

NEUROFIBROMA OF THE CAUDA EQUINAE. Abstr. Pearson.

The case is that of a white male, 53 years of age, admitted to University Hospitals, 1-15-32 and discharged 2-19-32 (35 days).

Pain, pigmentation

1926 - Developed shooting, stinging pain in region of right 6th intercostal space (anteriorly. Had occasional attacks at intervals for some time. Noted area of light brown pigmentation at this particular site.

Accident

1928 - Struck by automobile, then developed lumbar pain. X-rays at this time were negative. Was not well for about five or six weeks after accident.

More pain

1- -30 - Developed dull, gnawing pain in right hip. Later this became quite severe, sharp and knife-like. Also had pain from 2nd to 5th lumbar vertebrae. Pain was aggravated on straining, i.e., stool, and also severe at night.

Sciatica (Sciatic syndrome?)

4- -30 - Visited clinic where diagnosis of sciatica and prostatitis was made. Treatment prostatic massage, intravenous typhoid therapy and weight to leg. Later after returning from clinic, private physician injected alcohol into nerve trunk and continued prostatic massage.

Foci of infection?

Fall 1930 - Consulted physician who continued treatment as outlined above. Later, a submucous resection was done in effort to clear up foci of infection. A small, painless tumor was removed from medial aspect of right tibia. Following this, there was an area of numbness over medial aspect of right lower leg for about 6 months. Patient had known of existence of this tumor since childhood.

Nerve pain

1- -31 - Pain began to extend down leg. Had prickling sensation in 3rd, 4th and 5th digits. This was followed by similar sensation in upper part of calf. Sensation changed to intense pain. Saw chiropractor without avail. Lost about 30 pounds in weight.

Another tumor

Spring 1931 - Another tumor removed from left leg (not examined microscopically). Tried diathermy and ultraviolet treatment for pain.

12-14-31 - Went to bed because of severe pain.

Past History

Gastro-enterostomy and appendectomy, 6 years ago (ulcers of stomach). Obtained complete relief from these operations. Also hemorrhoidectomy, tonsillectomy, and sinuses washed. Has one brother who has lumps on scalp.

Hospital

1-15-32 - Admitted to University Hospitals. Physical examination reveals white male, 53 years of age, lying quietly in bed. Does not appear to be acutely ill. Chest - shows area of pigmentation, very light tan in color, over anterior left chest, approximately 2 x 4" in size, (6th interspace, 7th rib and 7th interspace at costochondral junction). Abdomen - old, right, upper rectus scar.

Scars

(Lower) 4 cm. in length, over medial aspect of right thigh; one; 1 cm. above left knee, one, on medial aspect of right tibia in midpartion; and one, 3 cm. long on medial aspect of left knee from which tumors had been removed.

Nodules

(Lower) - one, 1 x 2 cm. on posterior aspect of lower one-third of left thigh. pressure causes tingling, sharp, painful sensation which passes to posterior aspect of leg; another, 1 x 0.5 cm. on lateral aspect of sole of left foot, pressure causes sharp, tingling pain radiating to 5th toe; (upper) - one, 1 cm. in diameter, on ulnar side of volar surface of left forearm at middle third -- smooth, firm with skin over it freely movable and sensitive to pinprick, pressure causing tingling, painful sensation radiating down ulnar surface of forearm to little finger (sensation similar to that of striking "crazy bone").

Tenderness

(Not very marked) Present from 2nd to 5th lumbar vertebrae. Marked tenderness between tuber ischium and great trochanter of femur on right side along course of sciatic nerve.

Reflexes

Babinski's - bilateral, negative; middle and lower abdominals not obtained; right knee jerk - distinctly less active than left; left ankle jerk - normal, right - not obtained; Laseque's sign - positive on right; slight atrophy of right leg.

Laboratory:

Urine - 1.011, few wbc's. Hb. 101%, wbc's 6,750, Pmn's 62%, L 34%, M 3%, E 1%.

Neurological Consultation

1-18-32 - We are in accord with opinion that patient has neurofibromatosis. It seems very likely that a neurofibroma is present in sciatic nerve in depression between right trochanter and ischial tuberosity, considering marked tenderness in region with referred pain into right sciatic

distribution. The possibility of an intrathecal tumor producing this sciatic pain is not ruled out. Since one's attention is rather forcibly directed to trochanter-ischial tuberosity region and since exploration at this point is hardly more risky than a lipiodol injection, we would suggest exploration of the nerve here first, and then if no relief ensues, further study from the standpoint of a possible cauda equinae lesion.

Laboratory

Urine - 1,006, occasional wbc's.
Stool - negative. Prostatic secretion - 20 to 30 pus cells per oil immersion field, few gram negative diplococci which resemble Neisserian organisms morphologically, but as they are extracellular, they cannot be definitely so diagnosed. Complains of severe pain. Morphine sulphate gr. 1/8. Later in day, given morphine sulphate gr. 1/6, atropine sulphate gr. 1/180 and codeine sulphate gr. 1-1/2. Small palpable tumor on anterior surface of left forearm removed for biopsy. Pathological diagnosis - fibroma (from nerve).

Operation

1-20-32 - Anesthesia - spinal, supplemented by gas. Procedure: Lower portion of gluteus maximus on right side was cut near its insertion and reflected medially, Exposing upper portion of sciatic nerve up to sciatic notch, and down to lower border of gluteus maximum muscle. Beyond this, the nerve is, of course, sufficiently superficial so that a tumor should be palpable without exposure of nerve. This portion of the nerve exposed appeared to be entirely normal and palpation revealed no other abnormality. The finger was then inserted through the sciatic notch on both the upper and lower surfaces of the nerve in the hope that the tumor mass might be palpable in this situation but none was found. Then the nerve was exposed practically up to nerve roots which make up the plexus (nothing found). The tumor must therefore be intradural. Returned from operating room in good condition. Pulse 118. Respirations 22.

Neurological Examination

1-28-32 - Left ankle jerk - 1, right - not obtained. Knee jerks - both within normal limits. Sensation to pin and cotton - quantitatively less on whole

right lower extremity up to approximately first lumbar segment, buttocks included. It would appear probable that there are multiple fibromata throughout the cauda equina. Complains of severe pain in right hip. Pyramidon gr. x times (4).

Lipiodol of spine

2-4-32 - There is definite obstruction to passage of lipiodol in lower portion of spinal column with definite accumulation of solution at level of upper margin of 4th lumbar vertebra. Below this there is a definite rounded filling defect which is characteristic of a intraspinal tumor. Conclusions - Intraspinal tumor at level of 4th lumbar vertebra. Spinal fluid - 1 wbc, 7 rbc, Nonne +, Noguchi -.

Second operation

2-5-32 - Preoperative diagnosis - von Recklinghausen's disease. Tumor located at third lumbar spine. Anesthesia: Local infiltration, supplemented with ethylene inhalation. Procedure: Laminectomy, removing 1st, 2nd, 3rd and 4th lumbar spinous processes and laminae to expose corresponding portion of cord. The tumor was found at level of approximately the 3rd lumbar spine. It was about 1.5 x 2 cm. in vertical extent. Quite firm, and completely filled spinal canal so that very little spinal fluid was found until the canal was opened about it. It was well encapsulated, lying free within the spinal canal, but was attached to one of nerve roots. Tumor was removed. Part of nerve root was preserved. Pathological examination - neurofibroma. Returned from operating room in good condition. Pulse 136. Respirations 24. Intravenous of 2000 cc. saline started. Blood pressure 140/96. Morphine sulphate gr. 1/4 for pain.

Distension

2-6-32 - Developed abdominal distension. Pituitrin 1 ampule. Rectal tube inserted, with some relief. Nasal suction started. Very much relieved by suction. Complains of severe pain in hips.

2-10-32 - Bothered with hiccoughs. Finally stopped with bromides, chloral, morphine and H.M.C. tablets. Since

then, patient has felt much better and quite comfortable.

2-13-32 - Improved markedly. Complains of tenderness in left thigh and foot.

2-19-32 - Discharged in good condition. To return for follow-up.

Note: Type of neurofibromatosis in which intranural tumors cause pain, pigmentation and interference with function. No painless tumors present.

IV. CASE REPORT

NEUROFIBROMATOSIS. SARCOMATOUS DEGENERATION. Abstr. Pearson.

The case is that of a white male, 28 years of age, admitted to University Hospitals 4-7-31 - and discharged 6-7-31 (61 days).

Blow (Age 12)

1915 - Kicked in lower part of spine by a boy at school. Later noted right hip was black and blue but not swollen. Later area began to swell.

Tumor removed (13)

1916 - Developed hard tumor of right hip region, which was tender. Tumor could be moved about. Operated upon, and tumor size of grapefruit was partially removed with uneventful recovery. Tumor increased in size and became tender.

Other tumors (19)

1922 - Operated upon at a clinic for removal of similar tumors, one on dorsum of left arm, another above left clavicle. Similar tumor masses developed on other parts of body after this: Tumor of hip steadily increased in size. Masses were not tender when small but more so when larger.

Pain (24)

Summer 1930 - Began to have pain in hip. Legs ached especially after walking, so he had to sit down and rest frequently. Condition increased in severity.

9- -30 - Treated for pain medically.

Groin Operation (28)

1- -31 - Operated upon for removal of mass, size of grapefruit in right groin. Tumor in hip was not touched but shortly afterward became very painful. All of these tumors were diagnosed fibromas.

Past history

No history of fibromatosis in family

Physical examination

4-7-31 - Admitted to University Hospitals. Poorly nourished white male, 28 years of age. No special complaints except tenderness and pain in large mass over right hip. Head - small, soft mass over right occiput (about size of bean). Chest: Heart - B.P. 132/80. Spine - Slight tenderness over lumbar spine on pressure. Scoliosis. Extremities: Large, hard, tender tumor over right hip apparently attached to head of femur; soft, pendulous, round tumor over right buttock. Skin: Many, small, soft masses, size of lima bean, not especially tender, situated just below skin and freely movable. They seem to be along course of peripheral nerves. Laboratory: Urine - negative. Hb. 73%, rbc 4,360,000, wbc 11,400, Pmn's 70%, L 28%, E 2%.

Operation

4-9-31 - Preoperative diagnosis - neurofibromatosis. Anesthesia - spinal, very satisfactory. Procedure - Elliptical incision over external border of tumor combining two skin flaps, long one being on medial side. Through this incision, tumor was gradually uncovered. Upper portion was found to be encapsulated, but firmly adherent to ilium beneath. Gluteus muscle was gradually dissected away. The greater portion of gluteus maximus had to be sacrificed. The sciatic nerve was defined below tumor.

Tumor was found to be densely adherent to sacrum posteriorly and ilium below, and to extensor muscles of thigh at inferior margin of wound. By dissecting here and there around crest of tumor, tumor was gradually mobilized except beneath. It was found so densely adherent to muscle and bone that it was just cut across, some of tumor tissue being left there. At upper portion superior gluteal artery was cut across with considerable hemorrhage. Vessels were divided fairly close to origin from parietal division of hypogastric artery and hemostasis was somewhat difficult to effect. Inferior gluteal artery was cut and ligated without much trouble. Lower portion of tumor and sciatic nerve seemed fused. Tumor mass extended

through sacrosciatic notch into pelvis. Tumor tissue was therefore cut across, and a few loose pieces of tissue excised. Bleeding points were tied. Endotherm was used as a spark to a few small bleeding points. Two vaseline strips, two small Penrose drains were inserted. Because of patient's lowered blood pressure, saline solution was given during operative procedure, and later 700 c.c. of whole blood was given, intravenously. Also Morphine sulphate and fibrogen (1 ampule) times 2. Ephedrin 40 mgs. (once).

Laboratory

4-12-31 - Hb. 54%, rbc's 3,140,000. Pathological diagnosis - spindle cell sarcoma (degeneration of neurofibroma).
4-18-31 - Wound irrigated with 10 c.c. Dakin's solution. All sutures removed, and some necrotic tissue excised. Aspirin gr. x and pyramidon gr. v.

Operation

5-4-31 - Preoperative diagnosis - defect right buttock region from previous partial excision of neurofibrosarcoma. Local anesthesia. Procedure - an area, about 5 cm. square, was denuded of its epidermis. This epidermis was divided into small pieces, each about 1 mm. square, and transplanted on blunt ends of needles beneath the surface of the granulations.

Operation:

5-25-31 - Preoperative diagnosis - Has been quite well since attempted excision of tumor. A small tumor mass has come up in the lower angle of wound. The greater portion of the wound is epithelialized. It was felt that if some of the tumor could be coagulated today that perhaps the skin could be gotten to close with smaller margins. Anesthesia - spinal, very satisfactory. Procedure - with a finger the greater portion of the subcutaneous tumor was shelled out. 3000 mch. of radium was inserted about 10 days ago. Tumor has gone down somewhat in size. It was impossible to shell the entire tumor out due to tendency to bleed. After manual shelling out of a good portion of the tumor, the base was electrocoagulated. There was some hemorrhage necessitating the leaving in situ of 3 forceps and 2 small packs. Vaseline gauze pack and another dry gauze pack were placed over it and a few grafts were plated into the area which were not com-

pletely epithelialized. (Implantation method). Patient left the table in very good condition.

6-7-31 - Dichloramin-T packs to wound. Patient is up in a wheel chair and feels fairly well. Discharged.

Death occurred about 7 months later. No autopsy obtained.

Note: Malignant degeneration of one tumor through fibrosarcoma to spindle cell tumor. Unsuccessful attempts at removal.

V. CASE REPORT

NEUROFIBROMATOSIS (ELEPHANTIOSIS-LIKE DEFORMITY OF LEG).

The case is that of a white male, 26 years of age, admitted to the University Hospitals 3-5-32, and still is a patient in the Hospital.

Congenital deformity

Has had deformity of left lower leg and foot since birth. No difficulty in walking or getting around during first 15 or 16 years of life. Went to school, walking a mile a day (to and from school. Finished 6th grade. I.Q.?

Growth

1930 - Noted great increase in size of mass, causing patient to seek medical aid. No pain or tenderness in mass. No injury. Has several pea-size nodules over right leg, chest, arms, face, abdomen (some are soft, red, pendulous). Skin is now quite pigmented. Nodules have been present for about two years, and are not painful. Does light work on a farm.

Tumors, pigmentation

3-5-32 - Admitted to University Hospitals. Physical examination - well-nourished and developed, white male, 26 years of age, sitting up in bed. Seems quite nervous. Heart - B.P. 120/72. Extremities - Left lower leg and foot are markedly deformed; enormous soft, pendulous mass hanging down from tibia; all toes present. Skin - covered with numerous, brown pigmented areas; small, brown areas of pigmentation on chest and arms; heavier pigmentation in inguinal region; blotch of pigmenta-

ion in right arm, several on back and legs (more dense over sacrum). There are small elevations over body, lipocystic in nature. There is cystic nodule on right side of abdomen. Upper left leg shows atrophy. X-ray - (2-27-32) chest - left leg. Enormous enlargement of soft tissues of left leg involving chiefly foot and lower portion of leg. Tibia and fibula are markedly deformed. This is secondary no doubt to soft tissue lesion. The foot is extremely deformed, all metatarsals being very narrow, their proximal ends in several instances being completely absorbed. Bones of tarsus are also markedly absorbed and very much deformed. The whole appearance suggests pressure from tremendous soft tissue mass, the exact nature of which is not apparent. No involvement of knee. Upper portion of tibia and fibula look fairly normal. Appearance of metatarsals, phalanges, and tarsal bones suggests process similar to leprosy, multiple areas of ulceration and diffuse absorption being present. Some type of trophic change might also account for this appearance. Single plate of chest shows multiple rounded densities, three in left chest, one large one at right hilum, one large one near base of right chest. These suggest sarcomatous metastases by their appearance. The possibility of metastases from a teratoma of testicle must be considered, although they are not entirely characteristic of this. Conclusion - Tremendous soft tissue lesion, left leg and foot. Secondary erosion and destruction of bones of foot and leg. Probably sarcomatous metastases to both lungs.

Laboratory

3-7-32 - Skin temperature (C°)

Right knee, 36.1, left 32.5; right ankle 35.1, left 32.7; right big toe 31.3, left 31.5. Oral 36.9. Room 22. Urine - negative, very occasional hyaline casts. Blood - Hb. 85%, wbc's 7,000, Pm's 72%, L 22%, M 6%.

Blood chemistry - carbon dioxide, left leg 50 volumes %, right 66 volumes %. Question of arterio-venous aneurism is considered.

Operation

3-10-32 - Leg amputated in middle third of thigh as palliative measure. Good postoperative response. X-ray showed no definite arterio-venous communications.

Dissection revealed enormously thickened nerve trunks (flexiform) and one large lobulated greyish-red necrotic malignant tumor in lower portion.

VI. ABSTRACT

NEUROFIBROMATOSIS (von Recklinghausen's disease).

Abstr. Pearson.

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(Excellent bibliography and discussion of Chest Tumors).

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(Excellent early review)

General Statement:

Although neurofibromatosis has been known since 1800 (Alexander), it is only recently that there has been a general recognition of the widespread nature of the process. Many of us have thought of it as a fibroma connected with nerve trunks sometimes becoming malignant or necessitating operation because of growth within the nervous system. It is our purpose today to indicate some of the possible changes to be encountered in von Recklinghausen's disease.

Historical

Smith, R. W. (Dublin, 1849) is usually given credit for the first complete anatomical description of neurofibromatosis. Von Recklinghausen (Berlin 1882) pieces together the data which existed in his day into a clinical syndrome and the disease is now generally known by his name. Aside from individual case reports, the probable number of contributions from our country is not very large. The recent article by Kienbock and Rosler has a very large bibliography (many of the earlier references taken from Adrian?). The paper by Brooks and Lehman is a splendid contribution on the subject of bone involvement in this disease. Hosoi contributes to our knowledge concerning malignant degeneration. Heuer has written extensively on the subject of "Hour-Glass Tumors of the Spine." The monograph by Antomi (1920) is also a splendid contribution.

3. Classification

The mesodermal origin of multiple neurofibroma from the epineurium, perineurium and endoneurium of the nerve trunks is now generally accepted. Verocay expressed the belief that they were ectodermal (arising from cells of the sheath of Schwann.) Various names are given to these tumor formations, neurinoma, Schwannoma, glioma, etc. According to Penfield, multiple neurofibromatosis is a generalized fibrous connective tissue reaction showing both nerve fibers and connective tissue elements. He classifies all of the preceding and acoustic neuromas under the general heading of "perineural fibroblastoma" (see discussion of spinal cord tumors, Vol. III, No. 14). Neurofibromata may arise from the cerebral spinal or sympathetic nervous systems (neuro-

fibroma and ganglioneuroma). They are usually benign but may undergo malignant degeneration. They are usually multiple but may be single. Occur commonly in skin, viscera, nervous system and also involve special sense organ.

The characteristic findings are multiple pedunculated or flat soft tumors distributed over the entire body surface, associated with areas of pigmentation. The tumor locations correspond to the distribution of the cutaneous nerves or deep nerve trunks. The pigmentation is irregular, coffee-colored blotches or freckled areas in the skin. Associated with the classical picture already described, there is frequently noted other changes of widely varying character such as mental deterioration, congenital developmental defects, (spina bifida, hypospadias, glaucoma, elephantiasis, scoliosis) and other soft tissue and skeletal deformities. One gets the impression that these various anomalies have heretofore been considered as being accidentally associated with von Recklinghausen's disease. The disease, however, appears to have the stamp of congenital anomaly in the broadest sense and heredity has been noted in the considerable proportion of cases (approximately 20%).

4. Cause

The condition is apparently both congenital and hereditary in a large percentage of cases. In an increasing number of reported cases, the tumors are noted to have been present at birth as well as the other deformities noted.

5. Gross

Tumors vary from microscopic size to that of head. Tendency is to be egg-shaped and smooth. On cut section, they are gray to reddish-brown, show a fibrillar structure and are frequently gelatinous or slimy. They may be found on or in any nerve in the body, i.e., viscera, tongue, tonsils, intestines, stomach, mesentery, bladder, thorax, peritoneal cavity, extremities, central nervous system, etc. The changes in the bone have been studied and are apparently part of the same

process. As the tumor grows along the nerves in the bone, it causes destruction, cyst-like formation and deformity. It may actually stimulate the growth of the bone so that both long and short bones are accounted for on the same basis. Scoliosis is so frequently encountered in this condition may be a compensatory change.

7. Histology

The structure may resemble fibrous tissue (mature or very cellular). There are long narrow nuclei with cells often arranged in palisade formation sometimes in whorls. In addition, in the type commonly seen in association with the sympathetic nervous system, ganglion cells are found. At times the stroma is so edematous that it gives the appearance of a myxomatous tumor.

8. Ganglioneuroma

These tumors which belong to the general group only arise in the sympathetic nervous system, are benign, generally solitary, occur at the base of the neck, in the thorax, along the sympathetic plexuses and also in the suprarenal. Tumor in the thorax and abdomen may become large and frequently are of the hour-glass type. They are seldom multiple although cases of multiple tumors associated with subcutaneous nodules have been described. Horner's syndrome may be due to pressure of tumor in the thorax. May cause pain, or be silent. Fever is also noticed. Sometimes recurrent pains cause operations to be done for gall-bladder disease. Tracheal, esophageal stenosis - recurrent paresis of recurrent laryngeal nerve are noted. Frequent symptoms are cough, signs of pleuritis, sometimes paraplegia. Usually occur behind pleura in costovertebral angle and not in mediastinum. In their experience, ganglioneuromas have been most common in females, neurofibromas apparently equal in males and females. Average age is 40. Tumors have been present from 3 weeks to 17 years. (Keinbock and Rosler collected 58 cases of intrathoracic new growths (37 neurofibroma, 19 ganglioneuroma).

8. Age

May appear at any age. Probably all present at birth but do not appear until varying times of life. The differential diagnosis of disease is frequently

made by biopsy and many conditions must be considered before this information is received. Sometimes a biopsy reveals what is apparently a neurofibromatosis, i.e., something different (lipomatosis, angiomatosis, etc.)

9. Bones

Changes in the bones are less frequent in the ribs and spine (except scoliosis). Others may show defects, breaking down of tubular bones, partial hypertrophy, atrophy, erosion, development of exostoses, and cystic formation. Condition is frequently confused with osteitis fibrosa cystica and may at times resemble a Paget-like formation because of granular changes. There is frequently asymmetry of the head. The skull may show hydrocephalus, microcephalus, etc.

10. Hour-glass Tumors

The term is applied to a group of tumors which arise along the spine from the higher cervical to the lower sacrals, either from within spinal canal, or the vertebra. They are thought to grow outward through intervertebral foramina or between adjoining laminae to form paravertebral tumor or to arise in the paravertebral location primarily. Heuer has collected 64 cases from literature (including his own). Hour-glass tumors may at times be due to other causes than neurofibroma. By location, 18 in cervical spine, 37 dorsal spine, and 8 in the lumbosacral. The outstanding symptoms with only two exceptions were due to compression of the spinal cord.

11. Sarcomatous degeneration

Hosoi found 466 reported cases in literature; 13% had undergone malignant change. Change most frequently observed in deeper nerve trunks. Partial removal or any operative trauma of neurofibroma may activate the tumor into sarcoma? It is very uncommon for more than one tumor at a time to become malignant. After the removal or attempted removal of one tumor, another may show malignant transformation. As a general rule, recurrences are frequent and may be local or regional after operative measures. Such recurrences

may be repeated several times before distant metastases take place. Reports from literature indicate apparently 22% metastasized elsewhere, the lungs are the favored place. Death is usually due to cachexia resulting from many recurrences, metastasis and repeated operations. The tumors are usually of the spindle-cell variety and may be traced from fibrosarcomas to this stage. Sex ratio shows exactly the same as the reported series 1.6 to 1. 72% of the cases occurred in the 3rd, 4th and 5th decades, the youngest 15 and the oldest 70. The age of onset of von Recklinghausen's disease in the group varies from birth to 25 years. When sarcomas develop in viscerae where such tumors are not commonly encountered, a search should be made for evidence of neurofibromatosis elsewhere. In some instances, the development of a tumor is associated with the history of trauma. (See our case.) The elapsed time during which repeated attempts to remove the tumors are made may be very great.

Impressions

1. von Recklinghausen's disease or neurofibromatosis is a condition which may affect practically any of the tissues in the body. The fundamental process is essentially one of tumor growth.

2. Abortive forms probably occur but in the well-developed type the nervous system, special senses, skin, bones and viscera are involved.

3. It is suggested that in the future all phases of the disease should be studied. This should include a cooperative effort on the part of the Departments of Dermatology (pigmentation and tumors), Ophthalmology (eye changes), Otolaryngology (auditory involvement), Neurology and Psychiatry (changes in nervous system and psychic disorders), Surgery (biopsy, malignant degeneration, resectable tumors causing pressure symptoms), Radiology (study of bones and chest for tumors and metastasis), and Pathology (examination of excised tissue). A routine investigation of all cases would add greatly to our knowledge of the condition.

4. The presence of sarcomatous growths in unusual locations, e.g., bladder, intestines, etc. should suggest the possibility of neurofibromatosis elsewhere.

5. In every instance, an attempt should be made to determine when the tumors and

other defects were first noted. A careful family history is also indicated, because the disease is not only congenital but familial in many instances.

6. The necessity of biopsy is emphasized because many other conditions may resemble it or be associated.

7. Cases of osteitis fibrosa cystica should be considered neurofibromatosis until proven otherwise. With the increased interest in parathyroid adenomata as a cause of bone changes, this is especially important. The possibility of association of adenomata with many types of bone disorders has been suggested by Erdheim.

VII. THE DAY WE CELEBRATE

St. Patrick came to the shores of Ireland in 432 (1500 years ago). In 60 years he converted the people to Christianity, organized their laws, developed the annals of their nation and left behind him at his death a force that was destined to play a leading role in the salvation of ancient culture. Even more significant has been the effect of their religion, through all these years, on their social philosophy. For 1500 years this has been one of the most striking racial characteristics ever exhibited - probably shared only by the Jewish Race.

In the sixth and succeeding centuries, Western Europe from Iona and Lindisfarne to the banks of the Danube, Irish missionaries poured forth and evangelized the people. As a result, the patron Saint of hosts of European countries is an Irishman. In the 19th century, millions of immigrants came to English speaking countries as the result of economic conditions. The literature of the Irish people has a great cultural tradition (A Treasury of Irish poetry, MacMillan); their scientific contributions are many, and their names (in various forms) are to be found in the histories of practically every nation on the globe.

Their racial characteristics are so closely intermingled with their religious faith that it has been apparently impossible for them to lose either. One of the most adaptable of all races to new environments, they remain fundamentally the same wherever they go. Suspected of having an unusually well developed sense of humor, analysis reveals that this is probably not true; instead they probably do not take themselves as seriously as

other races. Emotional, improvident, charitable, their songs and stories reflect their attitude toward their daily life. Their pugnacious qualities are in sharp contrast to their tender sympathy for others in distress. The Irish mother has been eulogized in song and story for centuries. From the time her children are old enough to understand, she very carefully points out to them the necessity of living well so that they may die well. Few children start life so well equipped with a philosophy which is not primarily directed to achieving worldly success.

For more than 1400 years the world has paused to celebrate with them the memory of St. Patrick. It is suggested that their remarkable social philosophy - the direct outgrowth of the practical application of their religion to everyday affairs - is probably the reason for this widespread recognition.

For one day the world stops to enjoy with the Irish in song, story and social gathering, the joy of living which they have known through oppression and freedom for 1500 years.