(35 per cent), the abductor digiti quinti (10 per cent), the opponens digiti quinti (10 per cent), and the 4th dorsal (35 per cent), and 3rd palmar interossei (20 per cent). Sensory deficit was limited to the ring and little fingers (Fig. 1, A).

Six months after admission the sensory pattern was unchanged. Percussion of a small neuroma under the scar caused severe pain in the ring and little fingers. All intrinsic muscles of the hand were stronger than 70 per cent, except the abductor and adductor digiti quinti which had increased in strength to 50 per cent (Fig. 1, B and C). In August 1950 a scarpplasty was done and the ulnar nerve explored. The nerve ends lay about 1½ inches apart and were bound in dense adhesions beneath the scar. Neither response in the intrinsic hand muscles, nor sensation in the little finger was demonstrable on stimulation of the distal segment. An anterior transplant and neurorrhaphy were performed (by Gordon T. Wannamaker, Captain, M.C.). No aberrant branch from the median to the ulnar nerve could be found within 6 inches below the elbow. There was no change in the sensory pattern or strength of the hand muscles following this procedure. The postoperative course was uneventful. The strength in all the intrinsic hand muscles reached 70 per cent before he returned to full military duty in October 1950.

All the intrinsic hand muscles in this instance were evidently innervated through an unusual distribution of the median nerve. This anomaly illustrates an extreme instance of a commonly encountered variation which one must consider in diagnosis of lesions of the ulnar and median nerves.

REFERENCES


NEUROLOGIC COMPLICATIONS ASSOCIATED WITH HEREDITARY DEFORMING CHONDRODYSPLASIA

REVIEW OF THE LITERATURE AND A REPORT ON TWO CASES OCCURRING IN THE SAME FAMILY

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Hereditary deforming chondrodysplasia is described as a congenital abnormality of osteochondral development, resulting in irregularity and arrest in growth at the metaphyseal ends of long bones, causing deformity and the occurrence of multiple cartilaginous masses which with calcification become osteo-cartilaginous.

Various names have been given to this condition. The most frequently used are: hereditary deforming chondrodysplasia, multiple cartilaginous exostoses, dyschondroplasia and diaphysial aclasia.

A difference of opinion exists as to the etiology of this disorder but the general consensus points to a disturbance of the process of proliferation and ossification in the intermediary cartilage period of skeletal growth.

The first description of a case of multiple exostoses is credited to Key in Guy’s Hospital Reports of 1825. However, in the Surgical Essays by Astley Cooper and Benjamin Travers in 1821, an account is given of the case of Mr. Abernethy “of
a boy who came out of Cornwall, who was so excessively afflicted with an apparent predisposition to exostosis, or an exuberant deposition of bone matter, that a very trifling blow would occasion a bony swelling in any bone of his body. His ligamentum nuchae was ossified, . . . so that he was as it were completely pinioned; besides all this, the subject in question had numerous other exostoses in various parts of the body."

Familial Incidence. The familial nature of the disease was first noted by Stanley in 1849. Reinecke in 1891 reported on 176 cases occurring in 36 families running through to the fifth generation. Gibney in 1875 described the first case in the American literature and the familial incidence in 1876. In 1917, Ehrenfried reviewed over 300 articles and found about 600 cases of this condition. Leucutia and Price, in 1929, found more than 700 cases but their review is rather incomplete because Stocks had reported 765 cases in 1925. Reports of other cases of familial involvement have followed since then. One of these described four members of a negro family who were afflicted by this condition.

Differential Diagnosis. Hereditary deforming chondrodysplasia must be differentiated from:

(1) Achondroplasia (chondrodystrophia fetalis)—a congenital dystrophy of the growth cartilage occurring in utero and characterized by dwarfism, deformities and a bull-dog facies.

(2) Morquio’s Syndrome—a disorder of epiphyseal development with frequent absence of heads of femora and deformity of the spine, with crippling and dwarfism.

(3) Maffucci’s Syndrome—a dyschondroplasia associated with multiple hemangiomas and roentgen manifestations of multiple small cystic areas, particularly of small bones.

(4) Ollier’s Disease—a unilateral development of single or multiple exostoses. Some writers believe that this disease is similar to hereditary deforming chondrodysplasia, especially since in Ollier’s original case there was actually bilateral involvement. However, others consider this a separate entity because of its non-hereditary nature, generally unilateral distribution and roentgenographic signs of cystic changes similar to those of osteitis fibrosis cystica.

(5) Single bony exostoses, which are of frequent occurrence and pathologically are generally osteomas.

Neurologic Complications. Approximately 1000 cases of hereditary deforming chondrodysplasia have been reported. In 1925 Stocks reviewed 765 cases reported in all literature. In these and in cases reported subsequently in the English literature, neurologic complications were found to be rare. In many instances the reported complica-
tion was secondary to a single exostosis not necessarily of chondrodysplastic origin; in others the complication appeared to be the result of an associated neurologic disease and not secondary to an exostosis.

Neurologic complications have been reported by various authors. We have reviewed the available case histories of patients reported as having neurologic complications and these appeared to fall into two groups. Group I consisted of cases in which, in our opinion, the findings did not support a diagnosis of hereditary deforming dyschondroplasia or in which the neurologic disturbance suggested an unrelated disease associated with the dyschondroplasia. There were 11 cases in this group. Group II consisted of 19 cases in which the neurologic complication was directly related to an encroaching osteochondral exostosis. For the sake of brevity we have given the reference numbers only to these reported cases.

Because of the rarity of actual neurologic complications of so common a condition we are presenting 2 personally observed cases in which the etiology was proved and which occurred in the same family. In one case an exostosis of the cervical vertebrae compressed the spinal cord. The second patient, a brother of the first, had an exostosis of the femur compressing the sciatic nerve. Two children of the second patient also bear uncomplicated stigmata of the disease.

Case I. File No. 210621. P.N., a 33-year-old unmarried white man, was admitted Oct. 8, 1949 to the Buffalo General Hospital on the Neurologic Service of Dr. Burton M. Shimmer. He complained of paresthesias of the left 3rd and 4th fingers that had persisted since January 1949, followed by numbness of the hand, progressive weakness of the fingers and forearm, and impaired finger dexterity. In April 1949, he noticed a descending loss of sensation of the right lower extremity, urgency, frequency, dribbling of urine and suprapubic discomfort. Cough, sneeze and other forms of straining aggravated the pains and paresthesias.

Examination. He was a well developed man with obvious multiple skeletal deformities (Fig. 1). A large irregular mass was palpable in the left cervical area; a second mass was palpable in the left upper arm; smaller masses were found over the 5th and 6th ribs on the right; and a 10 cm. mass was in the left posterior tibial area. There was atrophy of the left supra- and infraspinatus, biceps and deltoid muscles. The circumference of the right upper extremity was greater by 1.0 to 4.5 cm. than the left, while the left mid-calf circumference was 44.5 cm. as compared to 37.5 cm. on the right. The left forearm was 3.5 cm. shorter than the right.

Positive neurologic findings were: a depressed left biceps reflex, hyperactive triceps and radial responses, and a left Hoffmann sign. The left lower deep tendon reflexes were more active than the right, with a Babinski sign and unsustained patellar clonus. Vibration sense was impaired on the left and position sense bilaterally. Hypalgesia was present over the left C6 dermatome distribution (deltoid area), the left forearm and proximal thumb areas (C5-6). Hypalgesia was present on the right side below D10 and decreasing hypalgesia from
D10 to C6. Touch sensation was preserved. The sensory changes on the right and pyramidal tract signs on the left suggested an incomplete Brown-Séquard syndrome at C5 cord level.

Roentgenograms on Oct. 8, 1949, showed multiple broad-based osteochondromas most evident on the left femur and leg (Fig. 2), and over C5 and C6 vertebrae (Fig. 3 A and B). Spinogram showed a complete subarachnoid block at C6 (Fig. 3 A).

Operation. At laminectomy at C5–C6 level, the vertebral canal was found narrowed in the anteroposterior diameter; the dura was elevated and displaced to the right by a grumous bony tumor. The intervertebral part of the tumor was removed but the paravertebral extension was too massive for removal at that time.

Histologic Report. A lobulated mass of hyaline cartilage with myxomatous degeneration in many areas (Fig. 4 A).

Course. After operation the patient improved subjectively but at discharge 12 days later the neurologic status was unchanged. He has not returned for examination but is known to be working daily as a stock clerk and is asymptomatic except for some shoulder pain.

Case 2. File No. 214985. A.N., a 37-year-old man, brother of Case 1, was admitted April 6, 1950, to the Buffalo General Hospital on the Neurologic Service of Dr. Burton M. Shinnors. During the past 8 months he had suffered paresthesias and cramping of the left posterior thigh and recurrent attacks of radiating sciatic pain.

Examination. He was a well developed man of small stature who had a large palpable mass over the left femur. Roentgenograms showed diffuse widening of the lower femoral shaft and a large cauliflower-like osteochondroma of the upper femur (Fig. 5 A) in the region of the lesser trochanter. Forearms and legs (Fig. 5 B) showed similar widening of the upper and lower shafts with shortening of the forearms and multiple osteochondromas.

Operation. The nodular femoral osteochondromas were removed later by Dr. Benjamin Obletz of the Orthopedic Staff.

Histologic Report. Osteochondroma (Fig. 4 B) with a broad base, having a thick cap of hyaline cartilage and covered by a thick fibrous perichondrium. Many areas of mucoid degeneration were seen in the cartilage.
FIG. 4. (A) Case 1. Osteocartilaginous tissue with fairly uniform chondrocytes and areas of myxomatous degeneration. (B) Case 2. Thick fibrous capsule surrounding osteocartilaginous tissue with large uniform chondrocytes.

FIG. 5. Case 2. (A) Pelvis and femora with changes typical of hereditary deforming chondrodysplasia. The osteochondral tumor arising from the left upper femur produced the sciatic pain. (B) Changes in distal and proximal ends of left tibia and fibula, increased density of osteochondral growth at proximal tibiofibular articulation, and exostoses demonstrating the diffuse bony involvement.
Fig. 6. (A) Roentgenogram of both legs showing the broad bases and exostoses typical of hereditary deforming chondrodysplasia in 7-year-old son of Case 2. (B) The left forearm and both legs show similar changes in the 2½-year-old son of Case 2.

Course. The patient had complete relief of paresthesias and pain. The roentgenograms of his two sons, aged 2½ and 7 years, are presented in Fig. 6.

SUMMARY

Nineteen cases of neurologic complications of chondrodysplasia have been found in the literature. Two new cases occurring in brothers are described, making a total of 21. Two children of one of our patients manifest asymptomatic stigmata of chondrodysplasia. The male sex was involved in 14 of 15 reported cases where the sex is mentioned. The spinal cord was involved in 10 cases, the sciatic nerve in 6, the peroneal nerve in 2 and the brachial plexus and ulnar nerve in 1 case each. One case was reported without specific nerve involvement. A brief review of the literature is presented.

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NEUROLOGIC COMPLICATIONS OF CHONDRODYSPLASIA