Thoracic cord compression due to chondrosarcoma in two cousins with hereditary multiple exostoses

Report of two cases

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Two teen-age male cousins with hereditary multiple exostoses developed cord compression secondary to chondrosarcoma. The clinical presentation, diagnostic work-up, surgical treatment, pathological findings, and postoperative course are described in each patient.

KEY WORDS - spinal cord - chondrosarcoma - hereditary multiple exostoses

PATIENTS with hereditary multiple exostoses rarely develop chondrosarcomas or spinal cord compression. The literature contains a single case of cord compression by a pathologically documented chondrosarcoma in a patient with multiple exostosis. We now present two cousins with multiple exostoses and chondrosarcoma producing thoracic spinal cord compression in the second decade of life.

Case Reports

Case 1

A 13-year-old boy was admitted to the Massachusetts General Hospital on August 6, 1970, because of progressive weakness in the legs of 3 months' duration. The family history was remarkable because of hereditary multiple exostoses in several relatives on the father's side.

Examination. There was marked weakness of both legs, and the patient was unable to hold either leg off the bed for more than a few seconds. Hypalgesia was present below the nipple line. Position sense was present but impaired in the toes, ankles, and knees. Deep tendon reflexes were absent in the legs. Long bone films showed exostoses of the right ulna and of the humerus, radius, and femur bilaterally (Fig. 1). A thoracic myelogram revealed a total block at T-2 (Fig. 2).

Operation. A posterior thoracic laminectomy of T-1, 2, and 3 was performed on August 9, 1970. A glistening gray mass was found in the right lateral spinal gutter impinging upon the dura at the T-2 level. All gross tumor, which extended into the T1-2 interspace anteriorly, was removed. It was also necessary to remove the transverse process and right facet at T-2. A multiple layered sheet of gold foil was left in the tumor bed.

Pathological Examination. The tissue was hypercellular with sheets of chondrocytes. The size and shape of the lacunae and chondrocytes varied (Fig. 3). Osteoid formation, calcifications, and hemorrhages were pres-
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Postoperative Course. After surgery, rapid improvement in sensory and motor performance was noted. By the 7th postoperative day, the patient could walk with assistance. At discharge on August 22, 1970, there was mild weakness of the tibialis anterior and extensor hallucis longus bilaterally. Minimal hypalgesia to the nipples persisted. Position sense was excellent throughout, the deep tendon reflexes brisk, and the toes downgoing. Postoperative tomograms revealed a question of minimal destruction of the bodies of T-1 and T-2, probably consistent with operative removal of bone. Follow-up neurological examination on December 1, 1970, was normal except for a slightly waddling gait. Repeat sagittal and coronal tomograms showed no change from those done 4 months earlier.

Case 2

An 18-year-old boy was admitted to the Massachusetts General Hospital on August 24, 1970, with a 4-month history of “buckling” in the knees, episodic tingling in the low back and lateral aspects of both legs, and occasional numbness of the feet. He denied pain of any sort. The family history revealed hereditary multiple exostoses in the boy’s father and paternal uncle. A search of family records demonstrated that the patient was a cousin, via the father’s side, of the patient described in Case 1.

Examination. There was moderate weakness of all muscle groups in the lower extremities. Sensation to pinprick and fine touch was diminished below the level of the umbilicus, with relative sparing of the perianal region and genitals. Vibration and joint position sense were absent in the feet and diminished at the knee and hip. The knee and ankle jerks were 3+ and symmetrical; the plantar responses were extensor bilaterally. The patient walked with a wide-based gait, and the Romberg sign was present. Long-bone films showed exostoses on the proximal right humerus and on the distal portion of both femurs. A myelogram showed a complete block by an extradural mass at T-8. Plain films and laminograms (coronal and sagittal) of the thoracic spine were normal.

Operation. On August 27, 1970, bilateral
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the preoperative status, and he walked unassisted. Perception of pinprick and fine touch was normal except for diminution in the feet and calves. Vibration and position sense was unchanged, and the toes were downgoing.

Discussion

Although cases of hereditary multiple exostoses are relatively common, 2-11 neurological abnormalities in these patients are unusual. 1-7,9,10,12,13 Sarcomatous degeneration in cases of hereditary multiple exostoses is estimated at 10%. 4 Therefore, the incidence of hereditary multiple exostoses with chondrosarcoma causing spinal cord compression must be quite small. A review of the literature disclosed only one such case. 8

Genetic investigations have shown that hereditary multiple exostoses are inherited as an autosomal dominant with full penetrance. 7 Thus, the existence of the condition in two cousins is not unexpected. On the other hand, the coincidence of hereditary multiple exostoses is rare, and the occurrence of a chondrosarcoma in one of the patients is even more unusual. The combination of these factors suggests that the occurrence of a chondrosarcoma in one of the patients may be due to chance.

Posterior laminectomies at T-7 and T-8 were carried out by Dr. H. A. Richter, Neurosurgical Service, Massachusetts General Hospital. A grayish mass presented anterior to the posterior longitudinal ligament and beneath the T-7 and T-8 nerve roots on the right. The mass was about 1.5 inches in diameter, felt firm and slippery to the touch, and did not bleed after being incised. All of the lesion that was visible was removed. The tumor was separate from normal bone except at the pedicle of T-8, and this structure was therefore removed as well. Satisfactory decompression of the spinal cord was attained.

Pathological Examination. The tumor histology was similar to that observed in Case 1. The pathological diagnosis was low-grade chondrosarcoma.

Postoperative Course. Sensory and motor performance gradually improved. On the 10th postoperative day, the patient began to walk in a back brace and with a walker. At the time of discharge on September 15, 1970, motor power in the legs was improved over the preoperative status, and he walked unassisted. Perception of pinprick and fine touch was normal except for diminution in the feet and calves. Vibration and position sense was unchanged, and the toes were downgoing.

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ostoses with chondrosarcoma producing spinal cord compression in two cousins is extremely unlikely. In this respect, our cases are unique in the reported literature.

The clinical picture in our two cases was atypical for vertebral chondrosarcoma. Pain is the commonest presenting complaint, but our two patients were both free of pain. Plain spine films often show osseous destruction with mottled areas of partial calcification and ossification, but in Case 1 there was only a question of erosion of the bodies of T-1 and T-2, and in Case 2 the plain films and tomograms were entirely normal. The previously reported case of multiple exostoses with vertebral chondrosarcoma and spinal cord compression had no pain and normal plain films of the spine.

Total surgical extirpation is the treatment of choice for chondrosarcoma, but complete removal of the tumor with good margins is rarely possible in the vertebral column. Radiation therapy does not appear useful since chondrosarcomas are relatively radioresistant, and cancerocidal dosage would result in radiation damage to the spinal cord. Chemotherapy may have something to offer, but experience is lacking. Frequent recurrence leading to death is the rule in vertebral chondrosarcoma. In the Mayo Clinic series, death or recurrence was reported in 20 of 26 cases by 6½ years, but several long-term survivals (up to 22 years) were observed. Reoperation, in some cases through an anterior approach, may be of benefit when recurrence occurs.

References

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