Cervical Cord Compression due to Exostosis in a Patient with Hereditary Multiple Exostoses

Case Report

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Hereditary multiple exostosis, or diaphyseal aclasis, is not a rare disorder. Almost 2000 cases have been recorded since 1849 when Stanley first described it. However, lesions affecting the central nervous system in this disorder are uncommon. This report describes a patient with an exostotic lesion compressing the cervical spinal cord.

Case Report

This 13-year-old girl was admitted to the Neurological Institute on June 28, 1964, with complaints of progressive weakness in the right leg of 4 months' duration and in the right arm for 3 months, loss of sensation in the left arm, and intermittent neck pain on turning her head to the right. Multiple exostoses had first appeared when the child was 4 years of age. Her mother also had numerous exostoses, but no other relative or sibling was known to be affected.

Examination. The patient was petite, slight, and of immature appearance. Height was 144 cm (average at 13 years is 152.2 cm) and the arm span was 125 cm (average 151.8 cm). Secondary sex characteristics were poorly developed. There were large palpable exostoses at knees, iliac crests, wrists, and elbows bilaterally. The right radius was bowed, with ulnar deviation of the wrist. There was spastic hemiplegia on the right side, with hyperactive reflexes, Babinski sign, and sustained ankle clonus. Pain and temperature sensation were decreased on the left side below the upper cervical region, and touch and vibration were decreased on the left side of the neck and left upper arm. The gait was markedly ataxic.

Head turning to the right was limited and painful.

The hemogram and urinalysis were normal; alkaline phosphatase was 39 King-Armstrong units. Lumbar puncture pressure was normal with cerebrospinal fluid protein 49 mg% and 1 WBC/mm. A RISA scan revealed no focal collection, but the electroencephalogram showed diffuse slowing. Skeletal x-rays confirmed the bony abnormalities described above (Fig. 1). Cervical films showed a poorly calcified density at the second cervical level which was visible only on lateral views. Myelography demonstrated an extradural type of filling defect at the C-2 level, with the cord displaced towards the right; a mass lesion at this level was outlined in the lateral supine view (Fig. 2). In view of the multiple lesions, the mass was believed to be a bony exostosis.

Operation. On July 15, 1964, laminectomy was carried out on the right from C-1 through C-4. A large bony mass was seen under the hemilamina of C-2. This mass had displaced the cord from the left to the right and had compressed it to approximately one-half its normal width (Fig. 3). The bony mass was smooth in appearance and was covered with a layer of cartilage. The cartilaginous cap and underlying thin cortical bone shelled out easily, exposing soft spongy bone. Postoperative laminograms revealed the origin of the mass at the junction of the lamina and pedicle at C-2 on the left.

Postoperative Course. There was immediate and continuous improvement of right-sided motor functions, which by 5 months postoperatively were almost normal. By then there was no Babinski sign although ankle clonus persisted; the left-sided sensory defects were only slightly diminished. The patient was attending school and traveling by public conveyance unassisted.

Discussion

Diaphyseal aclasis is a disorder of bone development characterized by heaping up of bone at the region between the diaphysis and epiphysis. The disorder is also referred to as hereditary multiple exostoses.

The genetic nature of this disorder is well established. In their large series, Stock and
Hereditary Multiple Exostoses

Fig. 1. X-ray of right forearm, demonstrating typical deformity. The radius has diaphyseal thickening and exostoses. The ulnar is shortened, causing ulnar deviation of wrist.

Fig. 2. Cervical myelogram, lateral supine view. The filling defect is seen at the level of the C-2 pedicle (arrow).

Fig. 3. Operative photograph. The exostotic lesion (arrow) is seen under the lamina of C-2 (end of instrument). The cord is markedly compressed from left to right.