PARAPLEgia FROM RUPTURED LUMBAR DISCS IN ACHONDROPLASTIC DWARFS

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A herniated nucleus pulposus can be especially serious in the achondroplastic dwarf because of his abnormally narrow spinal canal. Two such dwarfs were operated on by us during the past year with relief of pain and paralysis caused by a ruptured nucleus pulposus between the 2nd and 3rd lumbar vertebrae. Although a number of cases of achondroplastic dwarfs with paralysis of the lower extremities are recorded in the literature, we could find no previous report of a ruptured disc being the cause of paralysis in these cases.

In appearance the achondroplastic dwarf differs from the normal because of the marked shortening of his extremities and the unusual shape of his large head. In utero there are some disturbances of cartilaginous centers especially of the epiphyses of the long bones, causing a retardation in long-bone growth later in life. By x-ray examination at birth, the spinal column of the achondroplastic is normal in length and curvature. However, the entire spinal canal is narrower than normal because of the shortness of the pedicles. As these dwarfs continue to subject their spinal column to the stresses and strains of everyday life, many will show changes in spinal curvature.
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Fairbank\(^8\) pointed out that in achondroplastic dwarfs the spines are extremely lordotic, but often the dorsolumbar spines are surprisingly flat. Sometimes the apparent lordosis results from the unusual prominence of the buttock.

Donath and Vogl\(^2\) described more or less marked changes in the physiologic lordosis of these dwarfs at the dorsolumbar region. From the 12th dorsal to the 3rd lumbar segments the vertebral bodies show a definite wedging of varying degree. Thus the achondroplastic dwarf has an inherent weakness of the spinal column manifested by dorsolumbar kyphosis. Whereas some show flat backs, others reveal distinct gibbous formation at that level.

There are earlier cases in the literature of achondroplastic dwarfs with neurologic symptoms and signs suggestive of cord compression. However, Albrecht and Ranzi,\(^1\) in 1926, reported a case of compressive myelitis upon which they performed a laminectomy. Not only was the diagnosis confirmed but the achondroplastic dwarf regained many lost functions. Donath and Vogl felt that, in their case, arthritic changes above the level of the gibbus caused the formation of osteophytic spurs which encroached upon the spinal cord. Freund,\(^4\) then Kennedy et al.,\(^5\) subsequently demonstrated similar findings in case studies. Vogl and Osborne,\(^7\) in 1949, reported another example and developed further the pathogenesis of varying degrees of transverse myelitis in achondroplastic dwarfs as being caused by exostoses encroaching upon a constricted spinal canal. They showed that the entire spinal canal is narrower than a normal adult canal. Since the achondroplastic dwarf’s spinal cord is of normal diameter, the cord just fits into the canal with no room to spare. Hypertrophic arthritic changes appear adjacent to and above the gibbous deformity because of the increased motion compensating for the lack of motion at the level of the gibbus.

REPORT OF CASES

Case 1. W. B., a 33-year-old white male auto worker, was well until mid December of 1950 when he noticed pain in both hips and his “feet gave way” while lifting materials and suddenly turning his body. He was taken to a hospital where he was put on bed rest. His hip pain diminished at rest but became very severe with radiation down both legs when he was on his feet. He also noticed numbness, tingling and weakness of his legs. He was admitted to Harper Hospital Jan. 8, 1951, on the orthopedic service.

Examination. The patient was an achondroplastic dwarf. He was in no acute distress. Positive findings were: an absent left knee jerk, an absent right ankle jerk and hypalgesia of the right little toe and lateral aspect of the right thigh. Both legs were weak.

Course. Because he was thought to have a mechanically unstable back, his spine was manipulated under sodium pentothal and a plaster body cast was applied. The patient did well until he was on his feet and then his pain and paresis recurred. He was referred to the neurosurgical service.

A lumbar puncture was done on Jan. 23, 1951, with injection of pantopaque. A few drops of yellowish CSF were obtained; total protein was 9269 mg. per cent. The myelograms revealed a complete block at the level of the intervertebral disc between L2 and L3. The pantopaque did not flow up or down with the patient on the tilt table. This was thought to be consistent with an arachnoiditis.

Operation. A laminectomy was done with removal of the lower four lumbar spines and laminae. The spinal canal throughout was much shal-
lower than normal. The dura was blue and nonpulsating. Upon opening the dura, the cauda equina was found to be inflamed and tightly squeezed in the dural sac. A large resilient protrusion between the 2nd and 3rd lumbar vertebrae was palpated beneath the posterior ligament compressing the caudal roots. These were separated and an incision was made in the posterior ligament at the site of greatest swelling. Numerous fragments of nucleus tissue extruded themselves and were removed with a pituitary rongeur. The dura was not closed because of the swollen roots and arachnoiditis. No fusion was done.

Course. The patient had an uneventful convalescence with complete relief of pain (Fig. 1) and was discharged from the hospital on Feb. 19, 1951, wearing a back brace. He returned to his former work after 6 months and had no complaints when last seen 1 year after surgery. He had regained strength in his legs and was free of numbness. His deep reflexes were equal and physiological bilaterally.

Case 2. W. A., a 31-year-old colored female, was asymptomatic until October 1951, when she had vague bilateral hip pain radiating down both legs, especially the right. This was accompanied by numbness of her legs. She was 7 months pregnant at this time and was not too concerned with these symptoms. After she was delivered by cesarean section in mid December 1951, the pain and numbness persisted. On Jan. 19, 1952, she lost bladder sensation and was unable to void. When she tried to walk, her right leg was paralyzed and she experienced tingling sensations in her back and traveling down her lower extremities. She was admitted to the neurosurgical service on the same day.

Examination. The patient was a thin achondroplastic dwarf in acute distress. Positive findings were: limited straight leg raising on the right, saddle anesthesia, anesthesia of the lateral aspect of the right foot, absent right knee and ankle jerks and absent left ankle jerk. She was unable to stand or walk.

On Jan. 23, 1952, lumbar puncture and myelography were done. CSF pressure was low. The fluid was xanthochromic; total protein was 2550 mg per cent. Myelograms revealed a complete block at the junction of the 2nd and 3rd lumbar vertebrae.

Operation. On Jan. 25, 1952, an exploratory laminectomy at L2 and L3 was performed. Spines and laminae were removed, exposing a spinal canal which was narrower than normal. The dura was opened revealing a markedly congested cauda equina, the matted roots being tightly squeezed together. At the level between L2 and L3 on the right a large single herniated piece of nucleus pulposus material could be seen within the dura itself, pushing the conus and cauda from posteriorly (Fig. 2). The piece of nucleus pulposus had ruptured through the posterior ligament and dura and was found lying partially in the canal and partially in the intervertebral space. It was extracted in one piece, which measured 1 1/2 inches in diameter and 1 1/2 inch in thickness. Two smaller pieces were removed, and then a catheter was passed up and down the canal without resistance. The dura was left open and the incision closed. No spinal fusion was performed.

The pathological report was “nucleus pulposus.”

Course. The patient gradually regained strength in her legs, to the point where she got along well on crutches at time of discharge, Feb. 17, 1952. Her bladder sensation and function returned 1 week after operation.

She was seen 1 month after discharge. At that time her knees were wobbly but getting stronger, as were her legs. She continued to have good bladder function. However, there was still some numbness of the right foot and right saddle area. Two months after surgery she was walking without crutches and the numbness was subsiding.
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COMMENT

Both of our patients displayed signs and symptoms referable to a partial compression of the caudal roots. This was confirmed during laminectomy when herniated pieces of nucleus pulposus tissue were found between the L2 and L3 level impinging upon and causing congestion, edema and adhesions of the cauda equina. In the male the history suggests that his lifting and sudden turning were the factors precipitating the ruptured disc. In the case of the female, the strain of her pregnancy probably threw a greater burden on her spine with resulting rupture of her lumbar disc. After laminectomy and removal of the herniated pieces of nucleus pulposus material both dwarfs regained most of their lost motor and sensory functions and became symptom free.

In contrast to our findings of ruptured nucleus pulposus there are reported cases of compressive myelitis at higher levels (D10 or D11), caused by hypertrophic osteophytes. We found no osteophytes but did confirm previously reported findings of a narrowed spinal canal in these patients.

In both of our cases, the spinal column was normal in curvature by x-ray. Many of the previously reported cases revealed exaggerated lumbar lordosis of varying degree resulting from gibbous formation at the dorsolumbar area. This inherent spinal column weakness with wedging of the dorsolumbar vertebrae in some achondroplastic dwarfs may manifest itself in others by eventually herniating a nucleus pulposus under undue stress and strain.

SUMMARY

1. We have reported 2 very similar cases of achondroplastic dwarfs with partial paralysis of the legs and pain caused by a ruptured lumbar intervertebral disc. After laminectomy and removal of the herniated nucleus pulposus both dwarfs regained lost motor and sensory function.

2. In reviewing the literature there were found reported cases of achondroplastic dwarfs with varying degrees of compressive myelitis resulting from impingement of exostoses on the spinal canal.

3. Whereas some achondroplastic dwarfs form gibbi because of wedging of the dorsolumbar vertebral bodies, others may have a predisposition toward ruptured intervertebral discs. This is probably attributable to an inherent weakness in the spinal column of the achondroplastic dwarf.

REFERENCES