SPINAL EXTRADURAL CYST ASSOCIATED WITH KYPHOSIS DORSALIS JUVENILIS

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SPINAL extradural cysts are sufficiently rare and the results of their operative removal are so gratifying that further discussion of this interesting lesion seems warranted. Elsberg, Dyke, and Brewer[1] were the first to describe this lesion, reporting four cases, in 1934. Their search of the literature failed to reveal a single instance of a similar case on record, although Lehman,[2] one year later, added two additional cases and was able to find reports of three other instances of extradural cysts, only two of which probably fall into the group under discussion. Cloward and Bucy[3] were the first to recognize and point out the relationship of extradural cyst as an etiologic factor in the production of bony changes within the spine characteristic of and identical with those occurring in kyphosis dorsalis juvenilis (the rounded humpback of adolescence). In their article, which appeared in 1937, they cited nine other proved cases of spinal extradural cyst in addition to the one they reported, and found one other, unproved and unrecognized case, originally reported by Blum. Since the publication of their paper an additional case has been reported by Kelly.[5]

In the present communication we wish to report a case of spinal extradural cyst associated with changes within the spine identical with those described by Cloward and Bucy.[3]

Case Report.—H. H., colored, male, age 14, was referred from the Orthopedic Clinic of the James Walker Memorial Hospital, Wilmington, N. C., by Dr. Alonzo Myers of Charlotte, N. C. He was admitted to the James Walker Memorial Hospital, December 18, 1937.

Six months prior to admission the child first noticed weakness and stiffness of the right leg which gradually spread to involve the left leg and within two months had progressed into a complete spastic paraplegia. No history of trauma could be elicited. At no time had there been any pain, and the child was aware of no subjective change of sensation; although at the first examination a superficially infected, second degree burn was found over the left hip, which caused no pain at the time of its occurrence three weeks prior to admission. There had been no disturbance in the control of either the vesical or rectal sphincters. The general health of the child had been good and there was no history of recent illnesses or infections. He had received no medical attention prior to admission to the local hospital.

Examination.—The child was an illiterate but cheerful, well-nourished and well-developed Negro boy. The temperature, pulse, respiration, and blood pressure were all within normal limits. General physical examination showed the head, neck, lungs, heart, and abdominal viscera to be normal. The spine was in good alignment and no evidence of a rounded dorsal kyphosis could be demonstrated.

Neurologic examination revealed the cranial nerves to be intact. The musculature and strength of the upper extremities were normal and their deep tendon reflexes were

Submitted for publication September 29, 1938.

present and bilaterally equal. Hypesthesia was present below the level of the anterior iliac spines, the perception of light touch and pin-prick being diminished below this level. There was no "saddle area" of anesthesia. Kinesthetic proprioception and position-sense were completely absent in the lower extremities. There was motor paralysis of both lower extremities with marked spasticity. No muscle atrophy was discernible, but early contracture deformity was beginning to become evident. A mass reflex response could be elicited on painful stimulation. The deep tendon reflexes were markedly exaggerated and there was well-sustained clonus both at the ankle and patella. A positive Babinski sign was present bilaterally.

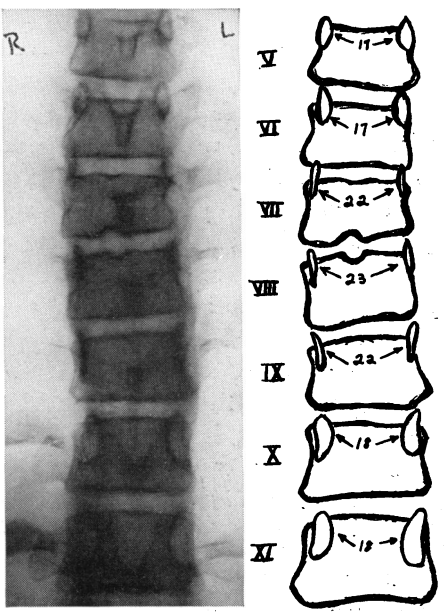

FIG. 1a.—Anteroposterior view of the dorsal spine. Note the thinning of the pedicles of the seventh, eighth, and ninth vertebrae, with a corresponding dilatation of the spinal canal. The disk between the seventh and eighth vertebrae has ruptured into their bodies.

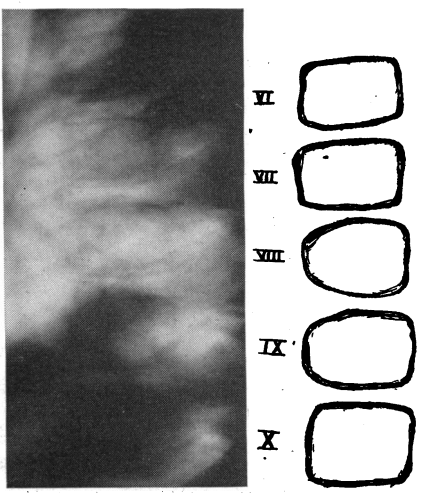

FIG. 1b.—Lateral view of the dorsal spine. The reproduction is not as clear as the original film, but a rounding off of the anterior superior and anterior inferior borders of the eighth and ninth vertebral bodies can be seen.

Spinal puncture between the third and fourth lumbar vertebrae revealed clear, colorless fluid under an initial pressure of 70 Mm. of water. There was no manometric rise on jugular compression, but a sharp response was elicited on firm abdominal pressure. The spinal fluid contained 15 lymphocytes and assayed a total protein content of 200 Mg. per 100 cc. of fluid.

Roentgenograms of the spine showed a slight, right lateral scoliosis in the mid-dorsal region (Fig. 1*a*). The laminae of the seventh, eighth, and ninth dorsal vertebrae were thin and their corresponding pedicles were markedly eroded and flattened on their medial surfaces. The spinal canal was somewhat dilated between the levels of the seventh and ninth dorsal vertebrae, inclusive. The transverse diameter, as measured by the interpedicular space, reached a maximum of 23 Mm., whereas the space immediately above and below this dilatation measured only 17 Mm. and 18 Mm., respectively. The intervertebral space between the seventh and eighth dorsal vertebrae was somewhat narrowed and there was evidence of rupture of this disk into the contiguous vertebral bodies. In the lateral view (Fig. 1*b*) the contour of the spine was normal. The anterior superior

and anterior inferior borders of the bodies of the eighth and ninth dorsal vertebrae were rounded and indistinct, but definite wedging of the bodies could not be demonstrated.

Diagnosis.—It was plainly evident that we were dealing with cord compression, most likely due to an expanding neoplasm in the mid thoracic region.

Operation.—December 21, 1937: Under avertin and ether anesthesia, the laminae of D IX, D X, and D XI were exposed and removed. This brought into view a thin-walled cyst which bulged down from under the eighth neural arch, filling the spinal canal entirely from side to side. The cyst was easily pulled down from under the arch and was found to be attached to the dura by a single narrow pedicle arising from the dorsal surface of the dura near the exit of one of the posterior roots. The pedicle was cut between silk ligatures and the cyst removed. This left a greatly dilated spinal canal and the laminae over this area were seen to be very thin. The cyst was roughly the shape

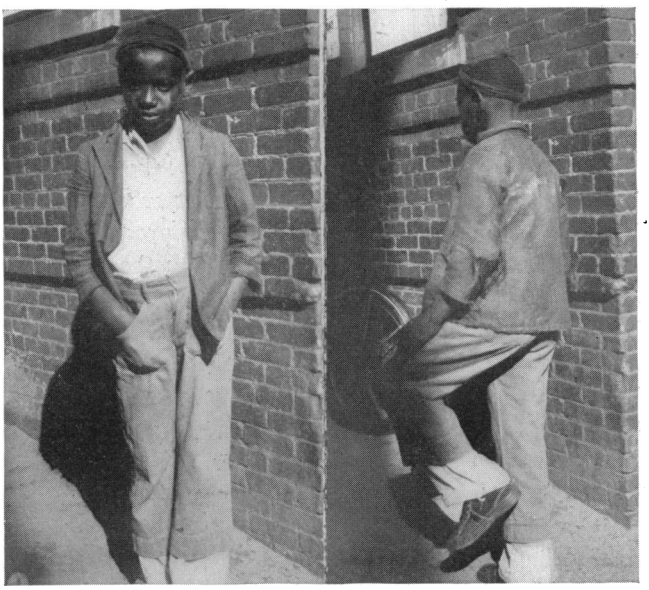

FIG. 2.—The patient three and one-half months after operation. Note the ability to stand on one leg unsupported, and the erectness of the spine.

of an egg and measured approximately 3x4 cm. Its wall was thin and translucent and contained clear, colorless fluid. Microscopically, the cyst wall was composed of thin avascular fibrous tissue lined with a single layer of flattened epithelium similar to that of the arachnoid membrane.

Postoperative Course.—Convalescence was smooth, and on the third postoperative day function began to appear in both legs, and thereafter strength and motion rapidly returned. A body encasement was applied with the spine in hyperextension and the patient was discharged, January 12, 1938, 22 days after operation. At this time the child was able to stand and walk with assistance.

Subsequent Examinations.—April 5, 1938: "The child walks with a normal gait. Muscle tone normal. Position-sense good. Sensation normal. Tendon reflexes hyperactive, and there is still unsustained ankle clonus. The plantar response is down."

June 25, 1938: "Normal muscular strength. Romberg negative. Knee jerks normal. No clonus. Abdominal and cremasteric reflexes active. Carriage is erect and the spine is of normal contour."

Discussion.—The clinical picture of spinal extradural cyst is remarkably constant and this feature has been emphasized in every article that has appeared on the subject. The lesion usually appears in adolescent boys between the ages of 12 and 16, but it is not unknown in girls. The cysts seem to have a predilection for the dorsal region, the majority arising from the dura in the midthoracic area usually between the sixth and ninth dorsal vertebrae. Just why they should arise in this region is not entirely clear, and we have no plausible explanation to offer. Elsberg, Dyke, and Brewer[1] advance the hypothesis that the cysts may arise either as a congenital diverticulum from the dura mater or as a herniation of the arachnoid through a defect in the dura. Cloward and Bucy[3] conclude from their microscopic examination of the cyst wall that there is more evidence to support the diverticular origin, although Kelly[5] favors the herniation theory. Direct communication between the cyst and the subarachnoid space has been demonstrated.[2, 6] Unfortunately, the pedicle in the present case was ligated before its connection with the dura was severed and a communication with the subarachnoid space was not shown. Microscopically, the cyst wall is composed of avascular collagenous fibrous tissue and its inner surface is lined by a single layer of flattened epithelium similar to the arachnoid membrane. It thus seems to contain the elements common to both the dura and arachnoid.

The symptoms of spinal extradural cyst are largely those of any other tumor encroaching upon the cord and giving rise to signs of cord compression. Weakness and spasticity of one or both legs are usually the first symptoms to appear, which gradually progress into a severe spastic paraplegia. Pain is usually strikingly absent. This seems strange since most of the cysts arise from or near the exit of one of the posterior nerve roots. This seeming paradox is probably best explained by the nature of the tumor itself; the soft, easily adaptable encysted fluid causes very little irritation and hence gives rise to no irritative phenomena. The tracts lying in the dorsal column of the cord are usually severely affected. Loss of position and kinesthetic sense is the usual rule, whereas the epicritic sense is usually less seriously involved. Loss of sphincter control is rare, as one would expect from the location of the tumor in the thoracic region. Sphincter disturbances are late manifestations and usually indicate severe cord damage.

The striking features on examination are the signs that usually accompany cord compression and pyramidal tract involvement, *i.e.*, spastic paraplegia, hyperactive knee and ankle jerks, sustained clonus, and a positive Babinski sign. The deep sensibilities are severely impaired and the position-sense is usually absent. Cutaneous sensation to light touch, pin-prick, heat and cold is usually diminished below the segmental level of the lesion, but the changes are usually minimal unless the lesion is of long duration.

Lumbar puncture may or may not show a complete block. Even if the subarachnoid space is completely occluded, as pointed out by Cloward and Bucy,[3] there may be a manometric rise on jugular compression due to the

pressure transmission quality of the fluid tumor. In these instances, however, lipiodol injection will demonstrate the block. The spinal fluid may be clear or xanthochromic, depending upon the completion and duration of the block. The total protein content of the spinal fluid is usually increased and in some instances may be quite high (200 mg. per cent in the case herein reported).

In the roentgenograms the most striking changes are demonstrated. Changes within the neural arch are the earliest to appear and later manifest themselves in the bodies of the vertebrae. Elsberg, Dyke, and Brewer[1] focused attention on the broadening of the spinal canal at the site of the intraspinal lesion as measured by the transverse interpedicular diameter. The medial surfaces of the pedicles are flattened or concave and the thickness of the pedicles is greatly reduced. The overlying laminae are likewise thin and eroded. The bodies of the vertebrae in the lateral views, as was first demonstrated by Cloward,[4] may show concavity of the posterior surfaces with broadening of the spinal canal. Other changes within the vertebral body characteristic of those seen in kyphosis dorsalis juvenilis are usually an accompaniment of these extradural cysts. The earliest change demonstrable is an erosion or rounding off of the anterior superior and anterior inferior corners of the vertebral bodies, as described by Scheurmann.[6] As the process progresses the intervertebral disk may rupture into the bodies and the involved vertebrae may collapse anteriorly giving rise to a rounded kyphosis. Cloward and Bucy[3] were the first to recognize that the changes occurring within the vertebral bodies in cases of extradural spinal cyst were identical with those occurring in kyphosis dorsalis juvenilis. They surmised that these vertebral changes were secondary to the presence of the intraspinal cyst, and advanced the hypothesis that the destruction of the vertebral body was the result of venous stasis caused by compression and occlusion of the venous channels draining these bodies. If this theory is correct, then it is reasonable to assume that this destructive process ceases to exist upon the removal of the cyst and that kyphosis if not present should not develop; and if it already exists its progress should be arrested provided adequate protection is given the spine during the period of reconstructive healing, either in the form of plaster jacket or spinal fusion. Further reports and observation of cases will prove or disprove this theory.

SUMMARY.—A case of spinal extradural cyst, occurring in a 14-year-old Negro boy, is presented. The cyst, containing clear fluid and lined by flattened epithelium, arose from the dura by a thin pedicle near a posterior nerve root in the midthoracic region. It was accompanied by spastic paraplegia and was associated with erosion of the neural arches, broadening of the spinal canal, and changes within the vertebral bodies identical with those occurring in kyphosis dorsalis juvenilis. Complete recovery resulted from its operative removal. A brief discussion of the symptomatology and pathology is presented.

CONCLUSIONS

(1) Paraplegia in an adolescent, associated with broadening of the neural canal and erosion of the vertebral bodies in the midthoracic region, as described by Elsberg, Dyke, and Brewer,[1] and Cloward and Bucy,[3] is pathognomonic of spinal extradural cyst.

(2) There is more evidence that spinal extradural cyst arises as a true meningeal diverticulum rather than as a herniation of the arachnoid through a defect in the dura.

(3) The theory, advanced by Cloward and Bucy,[3] that the destruction of the vertebral body results from venous stasis secondary to the presence of the intraspinal cyst appears to be tenable.

(4) Excellent results can be expected from the early recognition and prompt operative removal of the cyst.

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