CONGENITAL ROTATION OF THE SPINAL CORD
T. P. Morley, F.R.C.S.

University Department of Surgery, Toronto General Hospital, Toronto, Ontario

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A review of the medical literature disclosed no reference to an abnormality of the spinal cord of the type here described.

Case History No. D. 24296. J.G., male, aged 52, was admitted to the Neurosurgical Service on Oct. 27, 1952. Until 3 weeks previously he had been a fit man. On the day his illness began he was lifting a load of wood behind his house when he experienced a tight feeling amounting to a pain at the lower costal margin on each side of the trunk. This lasted for 20 minutes and he continued his work. A few hours later the pain recurred and, in addition, there was a pain in his back in the lower dorsal region. About an hour later he noticed numbness and weakness in both legs of which the right was almost paralyzed; he was unable to walk. The same evening retention of urine developed and he required catheterization. Thereafter his condition remained unchanged until his admission 3 weeks later.

Examination. The abnormal signs were referable to the spinal cord with the exception of a hypospadias at the penis-scrotum junction. (1) The right leg was paralyzed except for a flicker of movement in the hamstrings; the left leg was weak but all movements could be made against resistance. The lower abdominal muscles were weak. (2) The tone in the legs was increased, particularly on the left, and the left plantar response was extensor. (3) On the sensory side there was a complete absence of recognition of painful and thermal stimulation on both sides below and including the 9th thoracic dermatome. Immediately above this was a narrow band encircling the trunk where pin-prick could be recognized but it was not felt normally. Below this level there was an almost complete preservation of vibration and position sense, the only impairment being at the right ankle and foot. The sacral segments did not escape the general sensory involvement.

Investigation. (1) X-rays of the vertebral canal were normal. (2) Lumbar puncture: pressure 180 mm. CSF. Free rise and fall on jugular compression. Proteins 82 and 40 mg./100 ml. (3) Cisternal myelogram showed regular lateral indentations of the column of oil (Fig. 1) and in relation to these filling defects could be seen separate globules of oil of varying size. The significance of this appearance was not grasped before operation. There was no obstruction to the flow of oil up the spinal canal.

Operation. It was decided that exploration of the cord should be carried out, and since there was no myelographic localization the exposure was centred on the cord segments T6–9. The spines and laminae of vertebrae T6, 7, 8 and 9 were removed.

Figs. 2 and 3 illustrate the state of affairs that was discovered. The removal of the extradural fat revealed the spinal dura mater crossed on its dorsal

Fig. 1. Myelogram. Compare the appearances here with Fig. 2. Oil can be seen within the large cyst removed at operation, and the arrows indicate the regular indentations of the column caused by the constricting nerve roots.
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aspect at regular intervals by cord-like structures of 3 to 4 mm. diameter which could be identified as spinal nerves. Each nerve emerged, covered by its sheath of dura mater, from the left posterolateral aspect of the spinal dura mater and escaped from the vertebral canal through the right-hand intervertebral foramen at the same level. A probe could be passed between the nerve and the dural sac to show that it was a separate structure. Before the nerve escaped through the foramen it expanded into a cystic swelling, the size of which varied in different nerves from an insignificant bleb to a round cyst measuring 8 mm. in diameter. The wall of the largest of these cysts appeared to consist only of arachnoid for it was transparent and some nerve fibres could be seen glistening through it. The spinal dura mater was pulsating slightly but no pulsation could be made out in the cyst. Retraction of the dura mater towards the right exposed the left lateral and anterior walls of the vertebral canal. Here the intercostal nerves of the left side could be seen emerging from the intervertebral foramina, but each could be traced back into the vertebral canal as it ran at right angles to the long axis of the body closely applied to the wall of the canal and embedded in the extradural tissues until it disappeared beneath the spinal dura mater. By retracting the dura mater on the opposite side (the right) the emergence of these left-sided nerves could just be seen. No cysts were encountered on the left intercostal nerves in the operative field.

The largest cyst was excised between silver clips and the dura mater was opened as far as the nerves above and below would allow. The dorsum of the cord was not revealed, but instead nerve rootlets running diagonally downwards from right to left could be seen. These were the motor rootlets before they had joined the corresponding sensory ones to form the mixed nerve. The ligamentum denticulatum was identified clearly and could be seen attached to the dura mater a little to the left of the mid dorsal line; this was its true right-sided attachment. There was not any gross vascular abnormality. Nothing more was done and the wound was closed.

Progress. Four months after this operation he showed only slight improvement in the movement of the right leg. He was unable to walk and was committed to a wheel chair. The

Fig. 2. Appearance at operation (see text). The dura mater has been retracted to show a left nerve root lying on the wall of the vertebral canal.

Fig. 3. Diagrammatic cross-section of the anomaly (see text).
paraparesis was complicated by flexor spasms. The upper level of sensory impairment had risen to include the 4th thoracic dermatome.

Histology (Professor Eric A. Linell). The section contains a dorsal root ganglion and some dorsal nerve roots. Many of the ganglion cells have an irregularly shrunken cytoplasm in which the Nissl substance stains abnormally deeply. Nuclei are small, pyknotic and frequently eccentric. There is a marked increase in fibrous tissue within the ganglion. One of the nerve roots shows similar severe endoneural fibrosis with loss of myelin sheaths. A second bundle of nerves must have been twisted almost at right angles as part of it has been cut transversely and the remainder longitudinally. Its transverse portion is essentially normal. In the longitudinally cut portion the myelin sheaths are severely swollen and disrupted, their microscopic appearance suggesting operative trauma. The fibrous tissues surrounding both the ganglion and the nerve roots are thickened. Some normal extradural fat is adherent to the outer surface of this fibrous tissue along one edge of the section. A thin, moderately cellular fibrous membrane lies free on the opposite side of the section. This probably represents a portion of the wall of the cyst seen at operation.

DISCUSSION

Rotation of the spinal cord as described in this report has no embryological justification. It is curious that the dura mater and ligamentum denticulatum, both of mesodermal origin, should be involved as well as the ectodermal element, the cord itself. The only other congenital abnormality demonstrated was the hypospadias; radiographic investigation of the chest and abdominal viscera did not disclose further anomalies. It would have been interesting to know at what level the rotation had taken place. The myelograms of the cervical region were completely normal and the shadow of the nerve roots could be seen streaming out equally and at the correct angle from the cord. The lateral indentations formed in the thoracic area by the encircling peripheral nerves are absent from the cervical pictures. It can only be presumed that the rotation took place at the cervical-dorsal junction.

The sudden onset of symptoms may be explained on the basis of inflation of one of the cysts with cerebrospinal fluid at the time of a strain. On the other hand, the cord lesion may have been caused by a spinal vascular accident, although the bilateral girdle sensation of which he complained in the first place does not suggest that this mechanism was responsible. The rise of the level of sensory loss during the weeks succeeding the operation indicates either that a fresh compressive lesion arose in the postoperative period, or that the original exposure, although sited according to the neurological signs, did not coincide with the situation of cord damage. This would not be surprising since the myelogram gave no indication of the appropriate level.

SUMMARY

A case of rotation of the spinal cord and meninges is reported and illustrated. No previous published reference to this anomaly was discovered.

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