PAGET'S DISEASE OF THE AXIS CAUSING QUADRIPLEGIA

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Paget's disease of the axis is an uncommon condition. Only one case, reported by Whalley, was found in a review of the medical literature. In Whalley's case the atlas and the axis were both affected as well as the skull and there was an atlanto-occipital dislocation with paraplegia. In the case reported below the axis alone was affected and caused quadriplegia.

CASE REPORT

M. NS. 2635. A male aged 60 was admitted to the General Hospital on Oct. 10, 1933. For 15 years he had suffered pains all over the body, and during the last 5 years stiffness of the neck had developed with pain in the back of the neck. Three years prior to admission he noted weakness of the right lower extremity. Gradually this weakness involved the left lower limb also. During the last 6 months weakness spread to both upper limbs.

Examination. The patient was an ill-nourished old man. He was not able to walk or use his hands properly for eating or dressing. Intelligence and memory were good. The optic discs and all cranial nerves were normal. The size of the head was normal. The neck was stiff and the patient was not able to move the head freely in all directions. There was fullness in the upper part of the back of the neck. There was no tenderness anywhere on the spine. Muscle power in all four limbs was diminished and the tone was spastic. There was loss of appreciation for all modalities of sensation up to the level of the nipple. Deep reflexes in all extremities were exaggerated. Abdominal and cremasteric reflexes were lost. Plantar responses were extensor. The patient had retention of urine with overflow.

A diagnosis of cord compression at the cervical level was made. Roentgenograms of the cervical spine showed sclerosis of the 2nd cervical vertebra (Figs. 1 and 2).

The following diagnoses were entertained: (a) localized fluorosis, (b) localized spondylitis of the cervical vertebrae, and (c) secondary malignant bone deposit.

The blood Wassermann reaction was normal.

Lumbar puncture was done. There was a complete block on Queckenstedt's test. The cerebrospinal fluid proteins were 80 mg. per cent; there were no cells; and Wassermann reaction was negative. A myelogram done after cisternal puncture showed a block at the level of the 2nd cervical vertebra.

Operation. Anaesthesia was induced by intravenous Pentothal. An endotracheal catheter was introduced and anaesthesia was maintained with gas and oxygen. An incision was made in the mid line in the back of the neck. After separation of the muscles, the ligamentum nuchae was split and the spine of atlas and the axis were visualized. At this stage the patient was not breathing properly and respiration was maintained artificially by compressing the bag. The laminae of the axis were thickened, soft and very vascular. These were removed piecemeal and the cervical dura mater was exposed. The spinal cord was found to be kinked at the level of the body of the axis. The arch of the atlas was also removed to provide a good decompression and the wound was closed.

Course. At the end of the operation the patient still had difficulty in respiration, which had to be assisted. An hour later the patient regained consciousness. But still he could not breathe by himself. He was kept in this condition for 72 hours by artificial respiration. The trachea was kept clear by repeated aspiration. He was fed by a Ryles tube. On the 4th day the patient had cardiac failure and expired.
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Fig. 1 and 2. Roentgenograms of cervical spine, showing increased density of the axis. (Left) Lateral view. (Right) Anteroposterior view (patient’s mouth open).

Autopsy. The axis was enormously thickened, and there was a well marked hump at the level of the axis. The cord showed evidence of intramedullary haemorrhages.

Microscopical Examination. Irregular segments of the lamellae of bone are separated by short irregularly toothed cement lines. These appear fitted into one another and form an irregular mosaic. There is no attempt to form Haversian systems. The marrow between the bony trabeculae is fibro-fatty and in a few places consists of myeloid cells. There are also small isolated bits of deeply staining irregular trabeculae in the midst of the marrow. Of the lamellar pieces forming the mosaic a few stain differently from the others with haematoxylin, from light bluish- to purplish-red. There are also areas of reconstructed bone staining deep purplish-brown, irregular in their outline and extent, which in some places are alongside the normal trabeculae.

Discussion

Paget’s disease, or osteitis deformans, is not a common condition in India, although a few cases have been encountered. The spine often is involved as well as the other bones. The commoner sites of affection of the spine are the lumbar and the thoracic regions, the cervical region being less frequently involved.

Neurological complications of a milder type have been recorded in a number of cases. But paraplegia is not common. We found a total of 48 published cases of cord compression caused by osteitis deformans. Wyllie,8 Kay et al.,2 Schwarz and Reback,4 Turner,6 Whalley,7 Teng et al.,2 and Robinson5 have reported cases. The commonest site for compression paraplegia in Paget’s disease is the thoracic region. The interpeduncular spaces are the narrowest in the thoracic region between the 3rd and 10th dorsal vertebrae, and hence the cord is most likely to be compressed in the upper thoracic region. Paget’s disease of the cervical spine with cord compression is very uncommon. Only 3 cases were found in the literature. Garcin et al.1 presented a case of Paget’s disease with involvement of C4 vertebra. The patient had sudden development of quadriplegia and later partly recovered. Schwarz and Reback4 reported cervical cord compression at C6–C7 level in a patient with advanced Paget’s disease. Whalley’s7 case has already been referred to.