removal might not be attempted. We describe a cervical intramedullary schwannoma which was successfully excised, and discuss the origin of schwannoma within the spinal cord.

A 47-year-old man with progressive weakness and pain of the right arm and leg was admitted to the neurosurgical department. One year earlier he began to suffer from pain and paraesthesia around the right shoulder and in the lower cervical area which radiated to the right hand. For the last 4 weeks, weakness and paraesthesia in the right side limbs and urinary incontinence advanced progressively. Neurological examination revealed a severe weakness in right arm and leg with a mild spasticity. Muscles of the right shoulder girdle appeared slightly atrophic and deep tendon reflex was more brisk on the right side. There was hyperalgesia on the C3 to T8 dermatome in the right side and C7 to T8 dermatome in the left side and hypoalgesia below T9 level. Vibration and position sense in the lower extremities were impaired. Pantopaque myelography demonstrated an intramedullary space-occupying lesion with the lower level at C5. The cerebrospinal fluid contained 180 mg/dl protein. An emergency laminectomy from C3 to C6 was performed since quadriparesis developed suddenly following myelography. The dorsal column of the C4 appeared slightly expanded and was bluish in colour. A longitudinal incision at the right posterior column of C4 was performed. At a depth of 2·5 mm a purplish, firm, fairly well encapsulated tumour was encountered. It was possible to separate the tumour from the surrounding neural tissue. Total removal of the mass, which was in the posterolateral portion of the cord near the dorsal root entry zone was accomplished, and the tumour did not seem to have any continuity with the posterior root. Microscopic examination revealed a dense cellular mass of bipolar spindle-shaped cells arranged in palisades. Two weeks after operation, volitional movement of both legs was noted and superficial sensations were also improving. On examination six months after operation, he was able to walk unaided for a short distance and had good function of both hands with no sphincter disturbance.

There has been debate concerning the origin of the intramedullary spinal schwannoma, and various hypotheses have been advanced to explain the occurrence of this tumour. Schwann cells have been found along the endomedullary perivascular nervous plexuses,[7 8] or along some aberrant endomedullary peripheral fibres[9] in the spinal cord. Mason[4] outlined the role of the so called "critical area" as source of the tumour. This area corresponded to the point where the posterior roots lose their sheaths on penetrating the pia mater, and could be the origin of these tumours. They might arise from pial cells, considered to be of neuroectodermal origin, which might be transformed into schwann cells, or schwann cells may accompany the spinal root inside the spinal cord for a short distance. The possible differentiation of multipotential mesenchymal elements of central nervous system into schwann cells has also been hypothesised.[10]

It seems more likely that the neoplasm in our case began near the dorsal root entry zone with its sheath containing schwann cells and pierced the pia-arachnoid; the schwann cells subsequently proliferated within the spinal cord. Because this tumour is slow growing and benign, the functioning neural tissues surrounding it are relatively tolerant to chronic compression of mass, and functional recovery of the neural tissue after total removal of the tumour can be expected. Since it is a circumscribed, potentially enucleable and curable neoplasm, every effort should be made to remove the entire mass with minimum damage to the adjacent neural tissue. If the tumour is found to be large, it is preferable to remove it in sections with internal decompression of the tumour to avoid damage of nerve tissue. On the other hand, it should be considered that tiny vessels from the anterior spinal artery must be preserved to avoid ischaemic damage to the cord.

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## Cervical cord compression due to chondromatous change in a patient with metaphysial aclasis

Sir: Metaphysial aclasis affects only those bones or the portions of bones which develop both from cartilage and from membrane. The most characteristic feature is multiple outgrowth of exostoses from the surface of long bones.[1] Exostoses remain asymptomatic in most cases and rarely cause secondary neurological complications, in the form of compressive neuropathies and cord compression.[2]

A 25 years old Sikh male, 5 months before admission to the hospital noted slight weakness of the legs and difficulty in walking which did not interfere with daily activities. Three months later weakness of the arms was also noticed. Two months later there was dull aching pain and rapidly increasing weakness of both legs. At the time of admission he was able to walk about 50 metres with support. There was no history of root pains, girdle sensation or bladder disturbance. The patient was the youngest of six siblings. No one else in his family suffered from a similar illness. On examination there was a thoracic scoliosis with convexity to right and the lumbar lordosis was exaggerated. Height was 164 cm upper segment 84·5 cm, lower segment 79·6 cm and span 163·5 cm. The wrist was abruptly widened and a genu valgum deformity was present. There was a painless hard swelling of 8 cm × 5 cm on the dorsum of neck at the level of spinous process of fourth cervical vertebra. Wasting of the small muscles of right hand was present. Muscle tone in the arms was normal but there was spasticity of the legs. Power

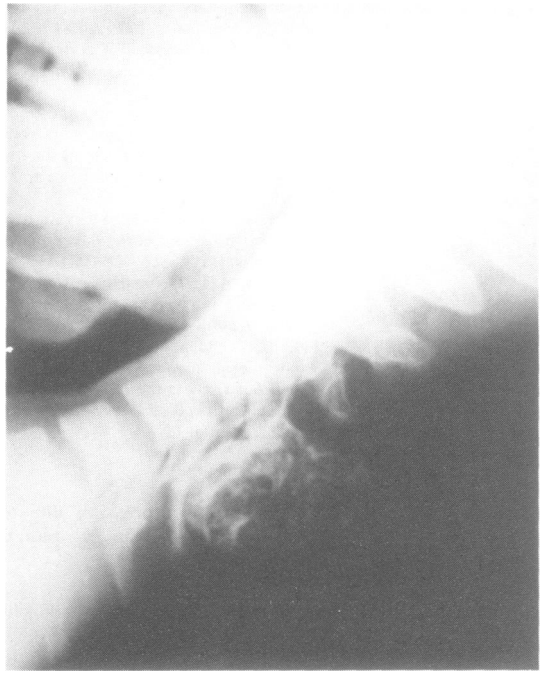

Fig 1 *Radiograph of cervical spine, lateral view showing a growth arising from the spine of fourth cervical vertebra.*

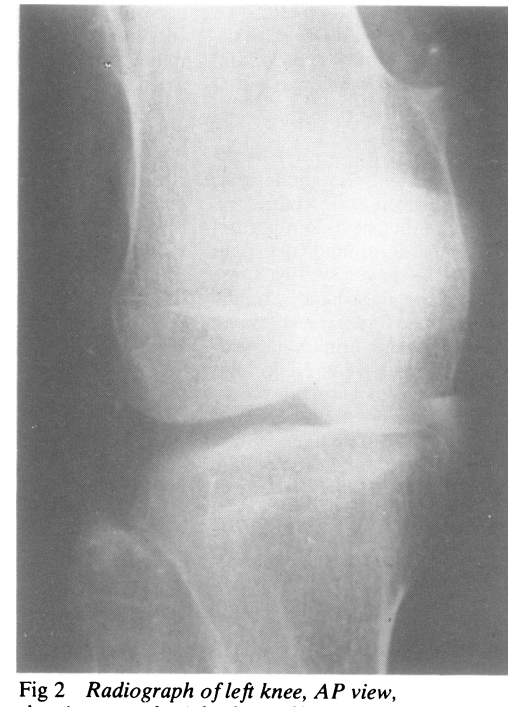

Fig 2 *Radiograph of left knee, AP view, showing metaphysial aclasis of lower end of femur and upper end of fibula.*

was grade IV in all the extremities (right side was weaker than the left). Abdominal and cremastric reflexes were absent. Deep tendon jerks-biceps, triceps, brachioradialis, knee and ankle were exaggerated and the plantar responses were extensor. Pain, touch and temperature were reduced below the level of C8. Sense of vibration and joint movement were absent in the left leg. The haemoglobin was 12·5gm%; total leucocyte count 8300/cu. mm, with 66% polymorphs, 32% lymphocytes and 2% eosinophils. Serum calcium was 8·4mg%, serum phosphorus 5·6 mg% and alkaline phosphatase 13 KA units. Blood WR and VDRL were negative. Radiographs of the cervical spine in the lateral view revealed a circular heterogenously calcified growth arising from the spine of fourth cervical vertebra; the growth displaced the spine of third and fifth cervical vertebrae (fig 1). Exostoses were present at the lower end of radius, ulna, femur, upper end of tibia and fibula (fig 2) bilaterally. Radiographs of the left arm showed a calcified shadow in the soft tissue, 4 cm × 3 cm with irregular margins. Lumbar puncture revealed clear cerebrospinal fluid under normal pressure, protein content 0·6 g/l, cells less than 5/cu mm and WR negative. Lumbar myelography showed total block with irregular margins at the level of fifth cervical vertebra (fig 3). Posterior midline exploration of the lesion was done. On retracting the paravertebral muscles, a large, irregular hard mass 8 cm × 5 cm of moderate vascularity was seen arising from the spine and lamina of fourth cervical vertebra. The spine and laminae of third and fifth cervical vertebrae were splayed. The dura was indented but not adherent. This growth was removed through a laminectomy from C3 to C6. Histopathological examination of the growth revealed it to be chondroma. Postoperatively he showed gradual improvement in motor power.

More than 2000 cases of metaphysial aclasis have been reported.[3] Originally named as exostosis it was renamed as diaphysial aclasis[4] but later a defect in the remodelling of metaphysis was pointed out and the term metaphysial aclasis was suggested. Metaphysial aclasis has two principal features, exostoses and unmodelling of metaphysis.[1] Exostoses are typically limited to the bones that develop in cartilage and come to be surrounded by sheath of subperiosteal bone. The long bones, iliac crest and vertebral border of scapula are the commonly involved sites.[5] Unmodelling of the metaphysis is characterised by abrupt enlargement of the metaphysis which is made up of poorly trabeculated bone.[1] Although metaphysial aclasis is an inherited disease, and is present at birth, its clinical manifestations are only observed after 7–8 years when exostoses become apparent owing to acceleration of growth.[1] New exostoses rarely appear after the completion of normal growth.[3] The present patient noticed the swelling at the back of neck at the age of 25 years. He had genu valgum, widening of the lower end of radius and scoliosis which are reported with metaphysial aclasis.[2,6] Other abnormalities reported in its association are growth retardation, bowing of radius due to shortening of the lower end of ulna, short arm span[7] and fusion of cervical vertebrae.[6,8]

Vertebral exostoses are reported in 7% cases of metaphysial aclasis.[9] They arise from the neural arch and are small in size.[10] Probably this is the reason that the spinal canal is spared and cord compression in metaphysial aclasis is so rare. Cord com-

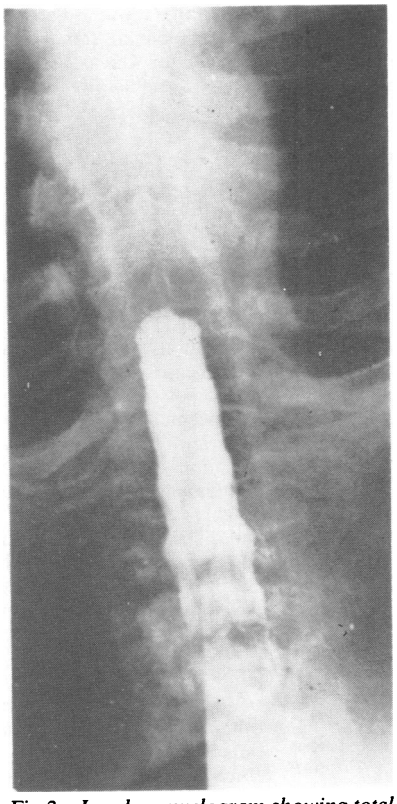

Fig 3 *Lumbar myelogram showing total block at the level of C5 (in Trendelenberg position).*

pression in association with metaphysial aclasis has been reported in 20 cases (1 in lumbar, nine in thoracic and 10 in cervical region[2 8]). Of the 10 cases of cervical cord compression associated with metaphysial aclasis, in seven cases the compression was at the level of second cervical vertebra.[2] In the present case the growth responsible for cord compression was at the level of fourth and fifth cervical vertebrae. It was probably due to chondromatous change in a vertebral exostosis. The rare association of metaphysial aclasis and chondroma has been explained on a developmental basis. In most cases when the growth of the skeleton ceases, the adjacent epiphysis fuses with the shaft, exostosis ceases to grow and the cartilagenous cap becomes ossified. Occasionally the cartilage on one of the exostoses may continue to proliferate and becomes so active to as form a rapidly growing chondromatous tumour.[10]

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