Intraosseous neurilemmoma of the cervical spine causing paraparesis and treated by resection and grafting

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SYNOPSIS A neurilemmoma presented as an expanding lesion of the bodies of C6 and C7 vertebrae and caused local neurological signs together with a spastic paraparesis. This tumour was treated by preliminary posterior fusion, followed by its complete removal via an anterior approach and stabilization by anterior spinal fusion. Other cases in the literature are reviewed and discussed.

Solitary lesions affecting the vertebral bodies are uncommon and present problems in regard to both diagnosis and treatment. This case report describes how such a lesion was investigated and found at operation to be a neurilemmoma probably arising from within the vertebral body. The extirpation of this lesion is described and discussed.

CASE HISTORY

A housewife, aged 34 years, was admitted to the Neurosurgical Unit in February 1974. In January 1970, after falling in snow and injuring her back and neck, she experienced pain in the neck for a few days. In June 1970 after a fall she had further pain and a radiograph showed a lesion, assumed at that time to be a fracture, in the body of C6 vertebra (Fig. 1). In April 1973 she began to have pain in the greater trochanter of the left femur which woke her at night. By December 1973 she also had pain in the right buttock and both sacroiliac regions, and she became aware that she was dragging her left foot and scuffing the toes of her left shoe. She also noticed some weakness of the left hand and a patch of numbness around the left knee. During the week before admission she experienced a band-like feeling around her upper abdomen and tightness in both legs.

Examination showed a healthy woman with no abnormalities apart from the neurological signs described below; in particular, there were none of the

FIG. 1 Lateral radiograph of the cervical spine in June 1970 showing the pathological fracture in the body of C6 vertebra.
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FIG. 2 (left) Lateral radiograph of the cervical spine in January 1974 showing progression of the osteolytic lesion which has now expanded the body of C6 and can also be seen in the upper half of the body of C7 vertebra. FIG. 3 (right) Anteroposterior radiograph of the cervical spine in January 1974 showing the osteolytic lesion in the bodies of C6 and C7 vertebrae, the pedicle of C6 vertebra is also involved on the left side (arrow).

stigmata of von Recklinghausen's disease. The cranial nerves were normal apart from a minimal Horner's syndrome on the left. In both upper limbs there was weakness of extension and flexion of the wrist and fingers and slight weakness of the left interosseous muscles. There was bilateral weakness of hip flexion, left more than right, and weakness of dorsiflexion of the left ankle and toes. The biceps and supinator jerks were present on both sides but both triceps jerks were absent. Both knee and ankle jerks were increased and the plantar responses were bilaterally extensor. Sensory testing revealed a slight bilateral loss of pain and tactile sensation involving the ulnar two fingers of both hands and the ulnar borders of the forearms, and extending down to the umbilicus.

Below the umbilicus pinprick became normal but light touch remained impaired. Joint position sense was impaired in the toes of both feet and vibration sense was absent in the left leg. Thus there was clinical evidence of cord compression in the lower cervical region together with bilateral nerve root involvement at C7 level.

INVESTIGATIONS Radiographs of the neck in January 1974, including tomography, showed an osteolytic lesion expanding the body of C6 vertebra and the left half of the body of C7, with some destruction of the pedicle of C6 vertebra (Figs 2 and 3). A lumbar myelogram in February 1974 showed a complete block at the lower border of C7 vertebra, with the spinal cord pushed to the right. The lumbar CSF contained no cells, but the protein content was 1.3 g/l. A left vertebral arteriogram showed that there was no gross blood supply to the affected vertebrae from this vessel. A skeletal isotopic scan (99 m Tc polyphosphate) demonstrated that the lesion was solitary. Other investigations showed a normal blood picture with a haemoglobin level of 13.5 g/dl, a white cell count of 7 600/mm³ with a normal differential, and an ESR of 10 mm/h (Westergren). The serum
electrolytes and urea were normal and the blood WR, VDRL, and Reiter's protein tests were normal.

Thus, the investigations showed an osteolytic lesion in the bodies of C6 and C7 vertebrae, crossing the C6/7 intervertebral disc space and causing an extrathecal block. The slow progression and lack of other constitutional disturbance suggested that it was probably benign and the preoperative diagnosis was an aneurysmal bone cyst.

**TREATMENT**  As excision of the two affected vertebral bodies carried a risk of collapse of the spinal column a posterior fusion to stabilize the cervical spine was carried out on 1 March 1974. Two bone grafts (each 8 cm x 1 cm) were taken from the right iliac crest and wired into place on either side of the spines and laminae of C5 to T1 vertebrae. Three weeks later the patient was allowed home to wait until this fusion had become solid. She was readmitted on 8 May, nine weeks after the posterior fusion and she was neurologically unchanged. On 16 May, under general anaesthesia, using controlled respiration and with the patient in the supine position, a pair of Crutchfield callipers was placed on the skull for skeletal traction. A left supraclavicular incision was made in a skin crease, as for an anterior interbody fusion, the anterior aspects of the C6 and C7 vertebrae were exposed and the site of the C6/7 disc space was confirmed by a lateral radiograph taken after a needle had been put into it. As the paravertebral muscles were dissected off the anterior aspect of the bodies of C6 and C7 vertebrae an avascular, greyish-brown, nodular tumour (2.5 cm x 2.0 cm) was seen protruding in places through the bone. The demarca-
mobilized using a Plastazote collar and, in late June, a soft collar was substituted to restrict flexion of her neck. It then became possible to see the graft in both oblique and lateral radiographic projections and these showed it to be in good position (Fig. 6). Six weeks after the definitive operation, examination revealed some weakness of the left triceps with slight weakness of extension of the left wrist and fingers, possibly due to damage to the C7 anterior nerve root at operation. However, this was recovering. The left triceps jerk remained absent. There was also a small area of impairment of pinprick in the left C8 dermatome. The only evidence of her paraparesis was bilateral extensor plantar responses. Twelve weeks after her operation her walking was completely normal and her other signs continued to improve.

DISCUSSION

In a series of 34 950 neurosurgical admissions, in Bucharest (Arseni et al., 1959) only 350 (1%) were patients with tumours affecting the vertebral column, which is an index of the rarity of these lesions. Also the majority affect the lower dorsal and lumbar spine and, among this group of 350, only 28 (8%) were in the cervical spine. Likewise, in the series of 413 apparently solitary lesions of the vertebral column described from the Mayo Clinic by Cohen et al. (1964), 45 (11%) were in the cervical spine. Metastatic lesions formed a sizeable proportion of these tumours. In the series of Arseni et al. (1959) they formed 66% of all tumours affecting the vertebral column. In the series of Cohen et al. (1964) of apparently solitary lesions affecting vertebrae, metastatic deposits formed 29.2% of the total, and 17.7% of the tumours in the cervical vertebrae.

Because of their rarity (less than 0.1% of all bone tumours in the series of Arseni et al., 1959), these cervical vertebral lesions pose a considerable problem in differential diagnosis. If the common tumours found by Cohen and colleagues are considered, we would need to distinguish between a metastatic deposit, a chordoma, a solitary myeloma, a chondrosarcoma, an osteoid osteoma and an aneurysmal bone cyst, with the addition (from the 103 primary tumours in vertebrae in the series of Arseni et al.) of giant cell tumour and angioma.

Several of these possibilities can be easily excluded; angioma, myeloma and chondrosarcoma have typical radiographic appearances.
which were not seen in the present case. Giant cell tumour (osteoblastoma) rarely occurs above the sacrum; 14 of 16 cases reported by Cohen et al. (1964) were in the sacrum. A solitary myeloma deposit is more difficult since this may occur in a vertebra without the disturbances of plasma proteins found in multiple myelomatosis. This was the case in 12 of the 20 patients described by Griffith (1966).

This, therefore, leaves the aneurysmal bone cyst, the osteoid osteoma, and the chordoma, with the very remote possibility, because of the long history and lack of constitutional disturbance, of a metastatic deposit. Both the osteoid osteoma and the aneurysmal bone cyst are more common in men than women and tend to appear before the age of 30 years (MacCarty et al., 1961). The chordoma, also more common in men, tends to present at an older age, between 30 and 60 years. Among the remaining lesions which occurred singly and in pairs in the cervical spine in the series of Cohen et al. (1964) was Ewing's tumour (which has a characteristic radiographic appearance) lymphoma and fibrous dysplasia of bone. A number of other lesions, including the eventual diagnosis of neurilemmoma, appeared in other parts of the spinal column. In this case the most likely preoperative diagnosis was thought to be an aneurysmal bone cyst. A neurilemmoma arising within bone is rare. In his monograph on bone tumours Jaffe (1958) describes only seven cases, five from the world literature and two of his own. Among a series of 3,987 primary bone tumours of all parts of the skeleton described from the Mayo Clinic by Fawcett and Dahlin (1967) there were only seven neurilemmomas. These authors excluded the six neurilemmomas described by Cohen et al. (1964) from their solitary vertebral lesions because they felt that the tumours had not arisen from within the vertebral body, although in two cases the radiological description suggests the contrary. In the most recent review by Spjut et al. (1970) of neurilemmoma arising within bone, 40 cases from the world literature are mentioned but only one of these was in the spine. Jaffe (1958) describing the osseous abnormalities associated with neurofibromatosis makes no mention of neurilemmoma, and in the 40 cases reviewed by Spjut et al. (1970) only one neurilemmoma is in a patient with neurofibromatosis. The cystic bone changes of neurofibromatosis are usually either multiple or subperiosteal (Friedmann, 1944; Hensley, 1953; Hart and Basom, 1958).

Neurilemmoma must arise from the nerve sheaths and over half these tumours in man occur in the mandible (Jaffe, 1958; Owen, 1969; Spjut et al., 1970). This has been attributed to the long intraosseous course of the mental branch of the inferior dental nerve. Small nerves have been described in the vertebral bodies of man and animals which could give rise to neurilemmoma (Pedersen et al., 1956; Stilwell, 1956; Sherman, 1963).

A careful search of recent literature, together with the present case, revealed nine cases of neurilemmoma possibly arising within a vertebral body. Of these, six cases were described by Cohen et al. (1964) and one each by Orf (1970) and Dickson et al. (1971). The present case was the only one in the cervical spine. Of the other eight, one was in a thoracic vertebra, four in lumbar vertebrae, and the remaining three in the sacrococcygeal vertebrae.

They had several clinical features of interest. They all had long histories, extending over several years. Seven were women, and two were men. Seven presented with pain, the other two presented with neurological symptoms. Five had neurological signs when they were first seen, four did not. One had the features of von Recklinghausen's disease (neurofibromatosis). The radiological appearances in these cases were variable. In our case and that of Orf (1970) the lesion spanned a disc space involving two vertebral bodies. In the others only one body was involved. The lesion in our case was thought to be an aneurysmal bone cyst. The radiological features of these cases, as described, seem to fall into two groups perhaps related to their point of origin. In one group there is gross destruction of the vertebral body, lamina and pedicle from an extrinsic expanding lesion, as in four cases described by Cohen et al. (1964) and in Orf's case where L1 and L2 vertebrae were involved. In the remaining group of three cases there was an erosive expanding lesion within the vertebral body, with sclerotic margins.

This is seen in the present case and corresponds nicely with the microscopical appearance of the decalcified specimen (Fig. 5). This also resembles the radiographic appearance of neurin-
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I wish to thank Mr Murray Falconer whose case this was, Dr Anne Marshall of the Department of Neuropathology of the Institute of Psychiatry, who prepared the histological description and photomicrographs of the tumour, and Professor H. Sissons, who confirmed her histological diagnosis. Mr J. S. Batchelor assisted with the positioning of the graft, and Mr R. C. F. Catterall also helped us. The prints of the radiographs were prepared in the Department of Medical Photography at Guy’s Hospital.

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*J Neurol Neurosurg Psychiatry* 1975 38: 776-781
doi: 10.1136/jnnp.38.8.776

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