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PAGET'S SARCOMA OF THE VERTEBRAL COLUMN WITH NEUROLOGICAL COMPLICATIONS

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A patient with Paget's sarcoma of the vertebral column who developed cauda equina compression is reported. This is the first report of this condition in an orthopaedic journal. The literature is reviewed. Palliative treatment is recommended.

Key words: complications; osteitis deformans

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Spinal cord and cauda equina compression is a recognised complication of Paget's disease of the vertebral column (Hartman & Dohn 1966, Miller 1967, Siegelman et al. 1968). One hundred and four such cases have been reported in the English literature. Six of these had histological proof of sarcomatous change. We report details of a further patient with sarcoma in Paget's disease of the vertebral column who developed paraparesis.

CASE REPORT

In November 1973, E.H., a 58-year-old woman, first complained of low back pain radiating to both iliac fossae. In January 1974, she was admitted to the general medical ward because of vomiting and abdominal pains. These complaints changed within 2 weeks to bilateral buttock pain, at which time it was first noted that she had Paget's disease of her lumbar spine and left hemipelvis; she was advised bed rest at home and a surgical corset support. In February 1974, she complained of pain in her right groin radiating to all dermatomes of the right lower limb, with similar less severe pain in the left side.

In March 1974, her pain worsened and she rapidly developed bladder and bowel paresis leading to her admission to Harlow Wood Orthopaedic Hospital.

On examination there was evidence of an incomplete paraplegia with an approximate sensory level at T_{12}. There were no external features of Paget's disease. X-rays again showed Paget's disease of L_2 and sclerosis of L_1. It now seemed that there was possibly some destruction of the right pedicle of L_1. Chest X-ray was normal. Myelography showed a complete block at L_1 level (Figure 1) with features of an extra dural compressive lesion on the right side. CSF protein was 200 mg per cent.

It was thought that the neurological condition might be due to structural narrowing of the spinal canal because of Paget's disease of L_1 and L_2, though tumour was considered.

On the 20th of March 1974, a decompressive laminectomy from T_{12} to L_5 inclusive was performed. The findings were of softened thickened vertebral arches at L_2 and L_3 levels, narrow root canals at L_1 level, thickened dura in all upper lumbar levels, but normal dura at L_4 and L_5 levels. There was extensive haemorrhage from the bone during operation, the blood loss totalling 2,100 ml. No certain tumour tissue was seen, but specimens were taken from all levels. Histology confirmed Paget's disease affecting all the arches from T_{12} to L_3. The specimens from the L_1 nerve root canals (the level of myodil block) revealed a cellular osteosarcoma on the right side with some infiltration at the edge on the left side (Figure 2).

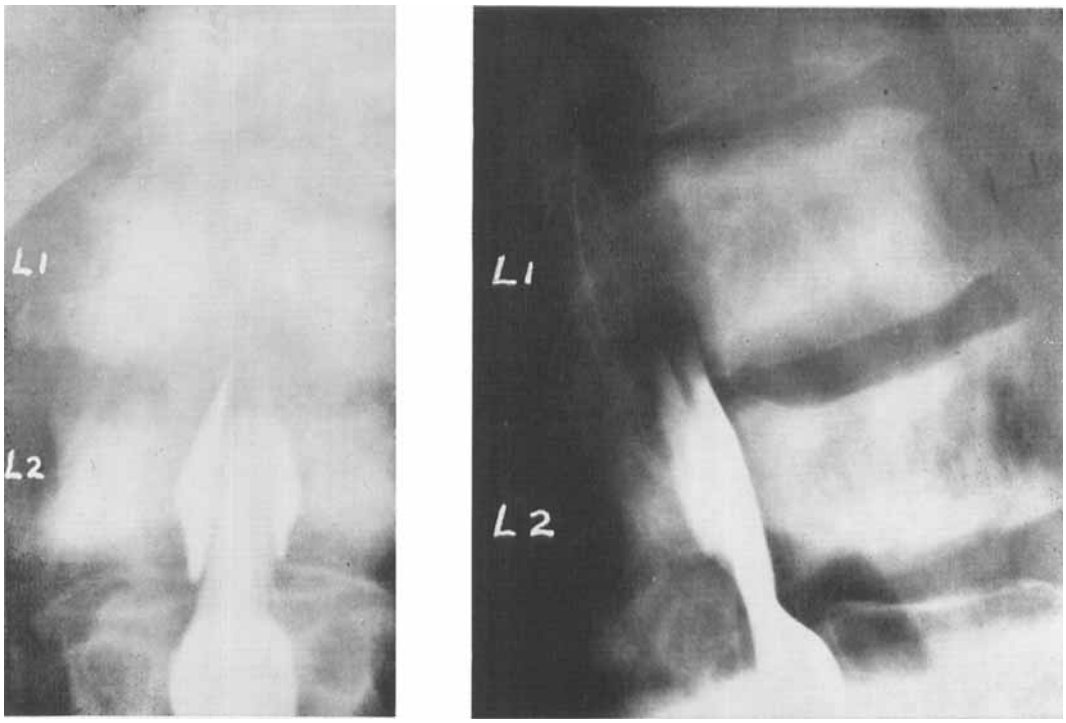

Figure 1. Sclerosis of the bodies of L_1 and L_2 with possible destruction of right pedicle at L_1. A complete block to the flow of myodil at L_1 level.

On the day after operation the patient noted relief of pain and some recovery of power in the right leg. By the eleventh postoperative day she had regained bladder and bowel control and began exercising out of bed. One month postoperatively a right pleural effusion was noted. This was aspirated but soon recurred and a week later one and a half litres of haemorrhagic fluid was withdrawn. Subsequently lung metastases were noted in the left mid zone. In spite of teletherapy her condition deteriorated rapidly, and she died on the 13th of May 1974. Permission for post-mortem examination was denied.

DISCUSSION

It is now 100 years since Sir James Paget of St. Bartholomew's Hospital, London, presented his original paper describing the condition he called osteitis deformans (Paget 1877). This disease is said to occur in 3.7 per cent of the population over 40 years of age (Collins 1956). The majority are subclinical cases, the pathology commonly involving the silent areas with lumbo-sacral spinal involvment in 76 per cent of cases (Dickson et al. 1945, Collins 1956).

Vertebral Paget's disease causing neurological complications is rare—only 104 cases have been reported. Wyllie (1923) was the first to describe this problem. Turner (1940) reviewed 34 cases and suggested the pathogenesis of neurological signs to be due to:

a) progressive bony expansion causing spinal compression,
b) cord ischaemia without spinal obstruction,
c) vertebral displacement.

The typical patient is said to be a middle-aged male with a history of 12 months insidious progressive paraparesis, X-ray signs of Paget's disease in the thoracic spine and a block in the myodil

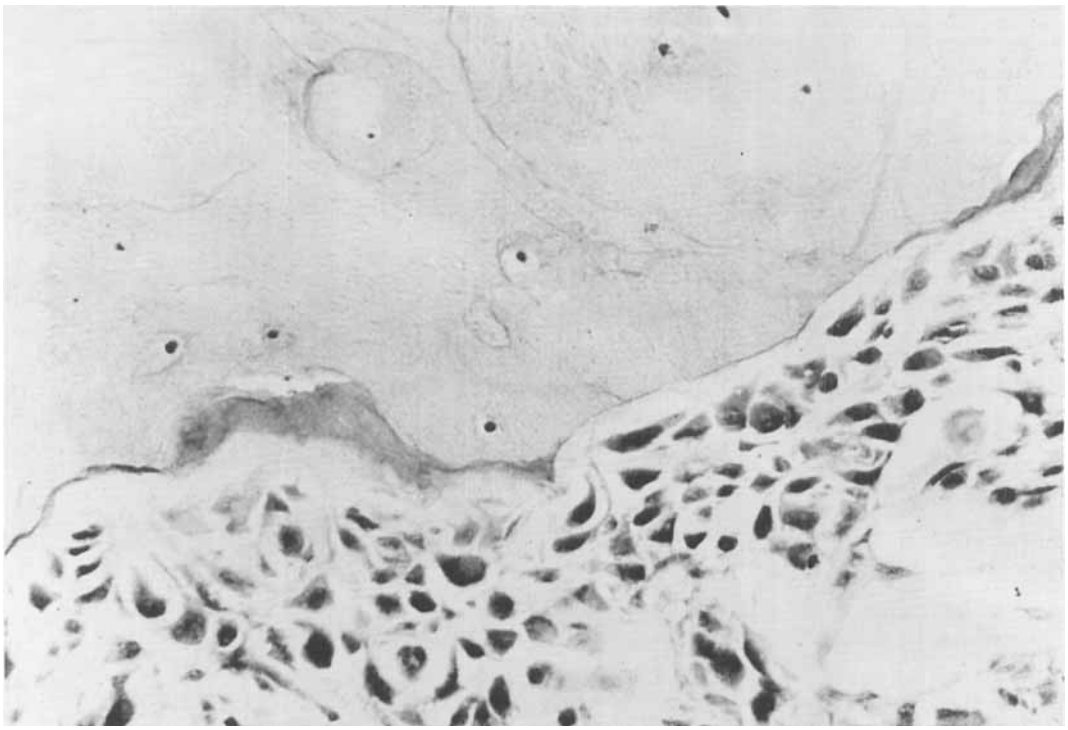

Figure 2. Paget's disease of bone with cement mosaic rimmed by malignant osteoblasts.

column. When treated by decompressive laminectomy a gratifying result may be expected (Hartman & Dohn 1966, Klenerman 1966, Miller 1967, Siegelman et al. 1968, Direkse & Milnes 1970, Cartlidge et al. 1972). Of the 104 cases so far reported, 69 had operative decompression and 58 had definite, though variable, relief of pain and neurological signs (Feldman & Seaman 1969, De Rose et al. 1971, Cartlidge et al. 1972, Sadar et al. 1972, Melick et al. 1976).

The true incidence of sarcomatous change in Paget's disease remains uncertain. When it occurs the sites commonly involved are the femur, pelvis, humerus and skull in that order (Porretta et al. 1957, Schajowicz & Slullitel 1966, Barry 1969).

Vertebral Paget's disease with sarcomatous change is exceedingly rare (Porretta et al. 1957, Price & Goldie 1969). Including our patient there are now seven reported cases of histologically proven Paget's sarcoma of the vertebral column causing neurological complications.

The seven case reports are summarized in Table 1. Symptoms of backache or sciatica had been present for an average of 13 months (range, 2 weeks to 4 years). Neurological signs were present on average two and a half months (range, 1 week to 12 months). At least two patients (nos. 5 and 7) showed no external evidence of Paget's disease. Paraparesis of varying degree was present in all patients on admission to hospital. A correct radiological diagnosis was made in the first three cases only. Polyostotic Paget's disease was present as a rule and a block was seen on the myodil column whenever myelography was performed.

Temporary (approximately 1 month) relief of pain and neurological signs was gained by all the five patients who had

Table 1. Summary of the seven case reports

Cases reported	History	Examination
1. 42-year-old male (Davie & Cooke 1937)	4 years leg pains 3 months weak left leg	Paraplegia
2. 52-year-old male (Campbell & Whitfield 1943)	4 months right sciatica 1 month frequent micturition	Cauda equina signs
3. 53-year-old male (Campbell & Whitfield 1943)	1 month back pain 1 week leg weakness	Spastic paresis. Distended bladder
4. 64-year-old female (Finneson et al. 1958)	4 years low back pain 1 month bilateral sciatica + leg weakness	Flaccid paraplegia with incontinence
5. 64-year-old male (Barry 1969)	12 months difficulty walking	Spastic paraparesis T_3 level
6. 53-year-old male (Sadar et al. 1972)	2 weeks bladder paresis	Flaccid paraparesis T_{11} level + sphincter loss
7. 58-year-old female (Shannon & Hopkins 1977)	4 months low back pain 2 weeks paraparesis	Flaccid paraparesis + sphincter loss

operative decompression performed. All patients died within 6 months of admission (average 4.2 months). Widespread metastases were seen in the four patients on whom post-mortem examinations were recorded.

Conclusions

It is important to recognise benign lesions causing spinal cord and root compression. Paget's disease of the vertebral column, often seen in orthopaedic clinics, sometimes causes backache, but seldom causes neurological complications. Cord and root compression in Paget's disease of the spine is rarely due to malignant change. The vast majority of patients with spinal compression who develop slowly progressive paraparesis (12 months) have simple Paget's disease (Turner 1940, Sadar et al. 1972). For these patients operative decompression should be advised since this frequently provides prolonged relief (Miller 1967, Siegelman et al. 1968, Direkse & Milnes 1970, Cartlidge et al. 1972).

A rapidly developing paraparesis (2 to 3 months) in a patient with Paget's disease of the spine suggests sarcomatous change. This possibility should be confirmed histologically.

In view of the inevitably rapid fatal outcome in the patient with sarcoma, the value of an extensive spinal decompression must be questioned. The histological diagnosis may well be established by a simpler procedure such as a needle or limited open biopsy, allowing palliative treatment of a non-surgical nature to be undertaken.

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of Paget's sarcoma of the vertebral column.

X-ray	Progress	Histopathology
Paget's subluxation L_1 and multicentric sarcoma	Death within 3 months No operation	Sarcoma L_1, L_2, L_3
Polyostotic Paget's and filling defects L_4, L_5, S_1 levels	Home 1 month Death 5½ monts post op. + DXT	Chondrosarcoma
Vertebral Paget's Myodil block T_{5-6}	Home 1 month Death 5 months post-op.	Osteosarcoma
Polyostotic Paget's Myodil block L_2	Slight improvement Death 6 weeks post-op.	Osteosarcoma
(Polyostotic Paget's) Not x-rayed	Death 'shortly' No operation	Osteosarcoma
Polyostotic Paget's Myodil block T_{12} + L_2	Urine control 3 days Death 5 months post-op.	Osteosarcoma
Polyostotic Paget's Myodil block L_1	Visceral control 10 days Walking 2 weeks Death 7 weeks post-op. + DXT	Osteosarcoma L_1

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