SOLITARY MYELOMA OF THE SPINE

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Plasma cell myeloma of bone may present initially with multiple lesions or with an apparently solitary lesion. Multiple myeloma runs its natural course in a period varying from a few months to an average of two years after diagnosis (Christopherson and Miller 1950), with fatal termination regardless of treatment. Solitary myelomas are unpredictable in their behaviour; they may remain localised for several years or they may disseminate.

In 1923 Shaw reported the first acceptable case of solitary myeloma, and since then a number of cases have appeared in the medical literature, though the majority lack sufficient detail to support the diagnosis. As yet, there is no agreement among pathologists as to whether the so-called "solitary myeloma" should be considered as a form of myelomatosis or as an unrelated benign lesion (Johnson and Meador 1951).

Willis (1941) and Raven and Willis (1949) were satisfied that solitary myeloma of bone is an entity distinct from multiple myelomatosis, but they stressed that a myeloma can only be accepted as a truly solitary lesion if the radiographic examination of the skeleton fails to reveal other deposits for at least one year after the original diagnosis. Using this criterion they accepted only eighteen cases, including two of their own, from the literature.

Lichtenstein (1959) considered even a three year period of observation hardly adequate, and thought it likely that cases of solitary myeloma, if followed up for a sufficiently long time, would eventually show obvious dissemination.

Those who consider that solitary myeloma of bone exists as an entity distinct from myelomatosis stress that multiple myeloma appears to be a neoplasm with multiple foci of origin rather than primarily a metastatic tumour, and suggest that the dissemination which occurs in a number of cases of solitary myeloma may be metastatic (Aherne 1961).

The vertebral column is the most commonly affected site in multiple myelomatosis and according to Christopherson and Miller (1950), and Griffiths (1966), this applies to solitary myeloma also, but in Raven and Willis's (1949) review of the literature the lesion was localised in the spine in only four cases of the eighteen that they accepted.

In the twenty year period from 1942-1962, forty-seven patients with histologically proven myeloma of the spine were seen at the Radcliffe Infirmary or the Nuffield Orthopaedic Centre, Oxford. Thirty-three cases had demonstrable multiple lesions at the first examination and all patients died within four years of diagnosis (twenty-five within the first year). The remaining fourteen cases were, at the time of presentation, apparently solitary. In five of these dissemination occurred from two to fourteen years after the clinical diagnosis. Four cases in which there are grounds for believing the lesion remained solitary have been discarded because they lack either adequate follow-up or complete post-mortem examination. The remaining five patients are alive and well without any signs of dissemination four to fourteen years after diagnosis and with an average survival of ten years.

We have also studied two further cases of solitary myeloma which were localised in the greater trochanter, and in these cases there were survivals of fourteen and twenty years without signs of dissemination.

In forty-seven cases reported in this paper histological confirmation of the diagnosis was obtained. In the fourteen cases of solitary myeloma the following criteria were considered necessary to exclude dissemination. 1) Normal cell pattern of the sternal marrow.
2) Normal electrophoretic pattern of the plasma proteins. 3) Normal peripheral blood picture and erythrocyte sedimentation rate. 4) Absence of Bence-Jones protein in the urine. 5) Absence of other skeletal lesions on radiographic examination.

CASE REPORTS

Case 1—A man of forty-eight complained in 1957 of pain in the upper chest and across the shoulders which was treated as osteoarthritis. In early 1958 he developed numbness and weakness of the lower limbs. Radiographs of the spine showed marked destruction of the first thoracic vertebra which was suspected to be due to a secondary neoplasm (Fig. 1).

On admission to hospital he was found to have severe spastic paraparesis, more marked on the left side than on the right. Myelography showed complete obstruction at the level of the first thoracic vertebra.

Investigations—The erythrocyte sedimentation rate was 4 millimetres in the first hour; haemoglobin 94 per cent; white blood cells 9,000; blood serum proteins 6-2 grams per 100 millilitres; albumin 4-4 grams per 100 millilitres; the urine was normal and no Bence-Jones protein was detected. Other investigations, including a radiographic skeletal survey, were negative. Sternal marrow biopsy showed no evidence of myelomatosis.

Treatment and progress—Cervico-thoracic laminectomy was performed and subtotal removal of a tumour, which had destroyed the left side of the first thoracic vertebra and formed a collar around the spinal cord from the fifth cervical vertebra to the third thoracic vertebra, was carried out.

There was an immediate improvement with restoration of almost full power and sensation in the lower limbs two weeks after operation. Radiotherapy was given (2,400 r. tumour dose to the cervico-thoracic spine in fifteen sessions for twenty days with a maximum skin dose of 3,800 r. posteriorly). Paper electrophoretic analysis of the serum protein done after operation indicated an increased gamma globulin fraction. Smears from the excised lamina of the first thoracic vertebra and sections of the neoplasm showed myeloma tissue.

One year later the patient was back at work. The only residual neurological abnormality was an increase of tone in the left lower limb and a bilateral extensor planter response which persisted for the following two years. Successive blood and urine tests, including electrophoresis of the plasma proteins, showed no abnormality.

In 1965, when he was again examined, the patient was doing full-time work as a farmer and had no complaints about his health. He had an even cervico-thoracic kyphosis without any point
of tenderness and full neck movements except for the extremes of flexion and extension. Muscle power, deep tendon reflexes and sensation were normal, the only neurological abnormality found being an equivocal right plantar reflex. Sternal marrow biopsy showed no signs of myelomatosis. Plasma proteins were 6-5 grams per 100 millilitres and the serum electrophoresis showed a normal globulin pattern. No Bence-Jones protein was detected in the urine. Haemoglobin, erythrocyte sedimentation rate, white blood cells, blood smear, serum calcium and alkaline phosphate showed no abnormality. Radiographs of chest, skull, pelvis, upper femora and the rest of the spine showed no lesions. Localised views of the cervico-thoracic junction (Fig. 2) showed complete destruction of the first thoracic vertebra with marked angulation, but no appreciable change since 1959.

Case 2—A woman of fifty-five was admitted to hospital in 1955. Twenty months before she had had an episode of pain in the thoraco-lumbar region which lasted for three weeks. Three months later, while carrying a light weight, she felt something slip in her back, and since then had been unable to walk properly because of weakness of the right leg. She had no pain until six to eight months later when she began to have severe low back pain with radiation to the right thigh.

Clinical examination demonstrated marked tenderness and a little prominence of the spine at the level of the fourth thoracic vertebra, flexion contractures of both knees, mainly on the right, and slight wasting of the right calf. Deep tendon reflexes were slightly increased and the plantar responses were extensor on both sides. There was no detectable weakness in the lower limbs, and she had normal sensation of the trunk and limbs except for some hyperaesthesia on the right at about the level of the fourth and fifth thoracic vertebrae.

Investigations—The haemoglobin was 82 per cent; white blood cells 6,800; erythrocyte sedimentation rate 31 millimetres in one hour (Westergren); blood plasma proteins 6-5 grams per 100 millilitres; albumin 3-7 grams per 100 millilitres. There was no Bence-Jones protein in her urine and a sternal marrow biopsy showed a normal distribution in erythroid and myeloid series. The cerebro-spinal fluid had 80 milligrammes per cent of protein with no cells.
Radiographs of the spinal column showed a complete collapse of the fourth thoracic vertebra with a small paravertebral swelling and partial destruction of the pedicles. Myelography showed an almost complete anterior extrathecal block at the level of the collapsed vertebra. No secondary deposits could be found in radiographs of the chest, skull and pelvis.

Treatment and progress—A thoracic laminectomy of the fourth and fifth thoracic vertebrae was performed with removal of a soft, pinkish-white tumour lying anterior to the cord and on either side of it. The cord did not look compressed but was displaced backwards. The right knee was manipulated. Histologically, the tumour was composed of myeloma cells showing considerable variation in size (Fig. 3). She was treated with radiotherapy, a total dose of 3,400 r. being given. She has never had any pain since operation and in 1957 neurological examination revealed no abnormality. She was able then to walk without aid and there was no sign of dissemination by radiographic or laboratory investigations.

In 1965, she was feeling well and was able to walk three miles without discomfort. No neurological abnormality was detected. The haemoglobin was 89 per cent; white blood cells 6,300 with normal blood film; erythrocyte sedimentation rate 19 millimetres in one hour (Westergren); no Bence-Jones protein was detected in the urine; blood plasma proteins 6.6 grams per 100 millilitres; and plasma electrophoresis showed a normal protein pattern. Sternal marrow biopsy showed a normal erythromyelopoiesis and thrombopoiesis, with no increase in plasma cells.

Radiographs showed a collapsed and dense fourth thoracic vertebra (Fig. 4) without evidence of progression of the lesion, and no deposits could be detected in the rest of the spine, skull, chest and pelvis.

Case 3—A man of thirty-nine complained in 1951 of low back ache following a fall and was treated by his local doctor for fibrositis. In 1952 he was admitted to hospital and treated for a prolapsed disc. Because of continuing pain he was readmitted to hospital in 1954, when a large osteolytic lesion was found in the sacrum. A review of the films taken in 1952 also showed the lesion though it was smaller. Two attempts at biopsy failed to reveal the nature of the tumour. He was followed up at regular intervals and the area of osteolysis got larger (Fig. 5). In 1964 a further biopsy proved the lesion to be a plasma cell myeloma. A course of radiotherapy with a total dose of 2,400 r. was given. When last seen in 1966, a radiograph of the pelvis showed sclerosis at the margins of the

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FIG. 5
Case 3—Radiograph of the pelvis showing a large destructive lytic lesion of the sacrum.
tumour. A skeletal survey did not show any other deposits. A sternal marrow puncture was normal. The haemoglobin was 13.8 grams per cent; white blood cells 4,600; erythrocyte sedimentation rate 18 millimetres in one hour (Westergren); total protein 6 grams per 100 millilitres, and serum electrophoresis revealed a slight rise in the gamma globulin fraction. The urine showed no Bence-Jones protein.

Case 4—A man of forty-five complained in 1956 of severe thoraco-lumbar pain after a fall a month before. On examination there was rigidity of the spine and tenderness from the tenth thoracic to the second lumbar vertebrae. Radiographs revealed a destructive lesion in the body and pedicles of the eleventh thoracic vertebra. Drill biopsy showed a plasma cell myeloma. Investigations—Sternal marrow puncture was normal; total proteins were 7.9 grams per 100 millilitres; and albumin 4.0 grams per 100 millilitres. Radiographs of the chest, skull and pelvis did not show any tumours. He was treated by 2,500 r. to the body of the eleventh thoracic vertebra.

In 1957 radiographs of the spine (Fig. 6) showed extension of the tumour into the tenth and twelfth thoracic vertebrae with fracture dislocation of the tenth and twelfth thoracic vertebrae. In 1964 no other deposits could be found on radiographic survey. In 1966 the erythrocyte sedimentation rate was 7 millimetres in one hour (Westergren); haemoglobin 11.6 per cent; total proteins 7.5 grams per 100 millilitres; and albumin 3.8 grams per 100 millilitres.

Case 5—A woman of eighty-four was admitted to hospital in 1962 with paraplegia and a six month history of progressive leg weakness. Radiographs of the spine showed collapse of the sixth thoracic vertebra and myelography showed complete obstruction at this level. Sternal marrow was normal; the urine had no Bence-Jones protein and serum electrophoresis was normal. At operation an extrathecal tumour was found and partly removed. Histological examination confirmed a plasma cell myeloma. Radiotherapy was given with a total dose of 2,200 r. By 1963 she had made a good recovery and when last seen in 1966 there were no radiological signs of spread. At this time the haemoglobin was 13.6 grams per cent; and serum electrophoresis showed a normal pattern. There was no Bence-Jones protein in the urine.

Case 6—A man of forty-eight was seen in 1956 with low back pain and right sciatic radiation. On examination flexion of the lumbar spine was limited by pain but no neurological abnormality was found. Radiographs showed partial collapse of the second lumbar vertebra (Fig. 7). A provisional
diagnosis of haemangiomata was made and a surgical belt provided. In 1959 he again complained of pain in his back which radiated to both legs, mainly the left, and he experienced some difficulty in walking. In 1960 radiographs of the lumbar spine (Fig. 8) showed complete disappearance of the body of the second lumbar vertebra. He was able to walk without discomfort, and had mild paralumbar muscle spasm and stiffness, with loss of lumbar lordosis. The second lumbar spinous process was slightly prominent but not tender. Straight leg raising was 80 degrees on both sides. There was no appreciable muscle weakness. The deep tendon reflexes were increased in the left lower limb but without clonus. The left plantar reflex was equivocal. Abdominal reflexes were

FIG. 7
Case 6. Figure 7—Radiograph of the lumbar spine in 1956.

FIG. 8
Figure 8—Radiograph of the lumbar spine in 1960.

FIG. 9
Case 6. Photomicrograph of tumour tissue obtained at biopsy. The cells are easily recognised as plasma cells; there is some variation in cell morphology. (Haematoxylin and eosin, x 100.)

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absent with inconstant hypoaesthesia of the lower abdomen to pin prick. Tactile, thermal sensation and joint position sense were not impaired.

Investigations—The haemoglobin was 101 per cent; white blood cells 6,700 with normal films; erythrocyte sedimentation rate 7 millimetres in one hour (Westergren); total plasma proteins 6.7 grams per 100 millilitres; albumin 4.6 grams per 100 millilitres, and a normal electrophoretic pattern; alkaline phosphatase 2 units; no Bence-Jones protein could be detected in his urine; and sternal marrow biopsy did not show any evidence of myelomatosis. The skeletal radiographic survey was negative. A needle biopsy of the "missing" second lumbar vertebra was done. Histological examination showed a well differentiated plasma cell myeloma (Fig. 9).

He was given radiation therapy (cobalt), with a total dose of 3,000 r. One month later he was discharged, wearing a surgical corset and walking.

When examined again in 1963, at which time the prominence of the second lumbar spinous process had increased slightly, he was able to bend to his toes despite a rigid mid-lumbar spine. There were no abnormal neurological signs. He was doing a full days work as a storekeeper and only wearing the corset occasionally.

In 1965 he suffered a pathological fracture of the left upper humerus, which was treated by radiotherapy. Serum protein and electrophoresis were normal. In 1966 radiographs revealed scattered osteolytic lesions in the ribs and pelvis and at this time the haemoglobin was 92 per cent; white blood cells 3,500; erythrocyte sedimentation rate 21 millimetres in the first hour (Westergren); total protein 7.5 grams per cent; electrophoretic pattern normal. The urine contained no Bence-Jones protein. The sternal marrow showed a heavy infiltration with sheets of myeloma cells.

DISCUSSION

In the cases of multiple myeloma reported here the commonest clinical presentation was backache with or without root irritation; in those of solitary myeloma of the spine the commonest clinical presentation was cord compression with paraplegia. Ten of the fourteen patients (71 per cent) with solitary myeloma had paraplegia in contrast to only six of the thirty-three (16 per cent) with multiple myeloma. This difference in clinical presentation is clearly related to size of the local lesion which is itself most probably time-dependent. The majority of cases of multiple myeloma terminate fatally, usually from renal complications within a year of diagnosis, and so do not grow as big as the solitary tumours which, as has been shown, have a very much longer survival time.

Cord compression in Cases 1, 2 and 5, and probably also in Case 6 was produced by the tumour mass originating in the adjacent vertebra and invading the extradural space; the rapidity of onset of signs of spinal cord compression can certainly be associated with the speed and extent of the vertebral collapse or the size of the extradural mass. Because of the extent of the extradural tumour and the mildness of the bony changes in some patients with solitary spinal myeloma, it has been suggested that possibly the lesion has arisen primarily in the soft tissue (Aherne 1961).

The difference in the radiographic appearances between solitary and multiple myeloma may be explained in a similar way to the differences in the clinical behaviour. No single description is generally applicable to the radiographic picture in cases of myelomatosis, and the classic characteristic of lack of osteoblastic reaction in the myeloma lesion, though common, is by no means diagnostic. Rypins (1933), Krainin, D'Angio and Smelin (1949), Sharnoff, Belsky and Melton (1954) and Engles, Smith and Krantz (1960) reported cases of bony sclerosis in myelomatosis, occurring sometimes in the presence of other purely osteolytic foci, and in rare cases it seems that osteosclerosis may even be the only demonstrable radiographic lesion (Odelberg-Johnson 1959).

In the vertebral column solitary myeloma is most commonly encountered as an osteolytic lesion, in contrast to the varying degrees of osteoporosis and wedging with ballooning of the discs often found in multiple myelomatosis. As Paul and Pohle (1940) described, osteolysis is in most instances limited to a single vertebra, as in Cases 1 and 2, but in others it has a tendency to extend across the intervertebral discs and invade the next vertebrae by direct extension, as in Cases 4 and 6. The lesion in Case 6 had other notable radiographic
appearances; in a very extensive review of the available literature no similar picture was found. The anterior osteophytic outgrowth bridging the bodies of the first and third lumbar vertebrae with complete disappearance of the whole body and pedicles of the second lumbar vertebra was most extraordinary; it was already present in 1956 (Fig. 7) and in 1960 it probably fractured (Fig. 8), producing a sudden small collapse which caused the symptoms of cord compression before his admission to hospital. Resorption of the whole of the affected first thoracic vertebra also occurred in Case 1, producing complete listhesis of the seventh cervical over the second thoracic vertebra (Fig. 2). In both of these cases, after treatment with radiotherapy, a certain degree of bone formation occurred, solidly bridging the adjacent vertebrae. Increased density of the affected vertebral bodies was seen in some patients after radiotherapy.

Christopherson and Miller (1950) did not find the variegated histological picture, which some cases of multiple myeloma present, in any of their cases of solitary myeloma. However, in common with other authors, we have not found it possible to distinguish by cytological examination between cases of solitary myeloma, solitary myeloma with subsequent dissemination, and multiple myeloma. In Case 2 of the present report there was considerable variation in cell size (Fig. 3) and yet the lesion has not disseminated, whereas in Case 6, in which dissemination occurred, the cells were remarkably regular (Fig. 9).

Johnson and Meador (1951) suggested that the benign form of solitary myeloma, though cytologically resembling a neoplasm, is in reality a focal hyperplasia of bone marrow histiocytes, possibly induced by some mild inflammatory agent. Nevertheless, true myelomas may be histologically distinguishable from non-neoplastic plasma cell masses, and replacement of local tissue by broad sheets of plasma cells oriented on a delicate capillary stroma is indicative of plasma cell tumour (Rawson, Eyler and Horn 1950). Furthermore, the local invasive properties demonstrated by all the solitary lesions reported here supports the view that they are truly neoplastic.

We suggest that there is no essential pathological difference between these two presentations of myeloma except for the individual host-tumour response. However, it is just this host-tumour response, at the present time poorly understood, which determines early death (multiple syndrome) or long survival (solitary syndrome). An analogy may be drawn between solitary myeloma and those rare cases of reticulum cell sarcoma and other malignant lymphomas which, instead of showing the usual pattern of widespread involvement of the lympho-reticular system, present with a solitary lesion. Early treatment in these cases may effect a lasting cure; on the other hand, either with or without treatment the classical widespread involvement of malignant lymphoma may develop.

Solitary myeloma should not be regarded as just an early stage of the disseminated syndrome but should be treated by excision or radiotherapy. Even in the event of subsequent dissemination, which may occur after many years, the prognosis is decidedly better than in cases of multiple myeloma. Histological examination will not distinguish the solitary from the multiple lesion and all stages of cytological differentiation may be observed in both types.

In solitary myeloma of the spine, paraplegia is a common presentation, but much less so than in multiple myelomatisis.

**SUMMARY**

1. Of forty-seven patients with histologically proven myeloma of the spine, thirty-three had multiple lesions at the time of the first examination and fourteen were solitary.
2. Five of the solitary cases, in which the patients are alive and well without signs of dissemination four to fourteen years after diagnosis, are considered in detail and the differences in clinical presentation and prognosis are discussed.
3. A sixth case, described in detail, showed scattered osteolytic lesions after ten years.
REFERENCES


