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Solitary plasmacytoma of the spine associated with neurological complications

Hisatoshi Baba[1], Yasuhisa Maezawa[1], Nobuaki Furusawa[1], Makoto Wada[1], Yasuo Kokubo[1], Shinichi Imura[1], Yoshiaki Imamura[2] and Yoshio Yamada[3]

[1]Department of Orthopaedic Surgery, Fukui Medical School, Shimoaizuki 23, Matsuoka, Fukui 910-11; [2]The First Department of Pathology, Fukui Medical School, Shimoaizuki 23, Matsuoka, Fukui 910-11; [3]Department of Orthopaedic Surgery, Tsuruga City General Hospital, Mishima 1-6-60, Tsuruga, Fukui 914, Japan

We report eight patients with a solitary plasmacytoma of the spine associated with neurological complications. The patients included five men and three women with an average age at presentation of 59 years (range, 47 to 73 years). The tumour was confined to the thoracic spine in six cases, cervical spine in one and lumbar spine in one. Duration of symptoms ranged from 2.5 to 22 months. Treatment consisted of a combination of radiotherapy, melphalan and surgery. One patient progressed to multiple myeloma 7 years after surgery. Surgical treatment (anterior surgery in three cases and posterior surgery in five) produced neurological improvement in all patients. We stress the importance of an early diagnosis followed by appropriate treatment including surgery for this clinical entity and long-term follow-up to detect a disseminated disease.

Keywords: solitary plasmacytoma; spine; neurological complications; multiple myeloma; melphalan

Introduction

Solitary plasmacytoma is a localised osseous tumour of a single focus of plasma cells.[1–3] The tumour may manifest clinical features different from those of the relatively more common disseminated form of the disease, multiple myeloma.[4–8] A firm diagnosis of solitary plasmacytoma is established after a thorough skeletal radiological survey or surgical treatment confirming a lack of additional bone lesions. Solitary plasmacytomas comprises $<3\%$ of all primary spinal neoplasms. The spine is a common site of involvement where it is possibly associated with neurological complications. Early diagnosis followed by irradiation and chemotherapy may reduce the frequency of such neurological complications.

The present communication provides a description of our experience and summarises the clinical presentation and treatment of a number of patients with solitary plasmacytoma of the spine complicated by neurological compromise.

Patients and methods

A total of 73 patients with plasma cell myeloma were seen in our Medical Centres between 1987 and 1995, for examination of spinal involvement as well as neurological assessment. After systemic skeletal survey and serological tests, 54 cases were confirmed to have multiple myeloma, while 19 had solitary plasmacytoma. The latter group (11 men and eight women; age, 42 to 73 years) showed radiological involvement of the spine in 16, ilium in two, and the ischium in one. Neurological complications were present in nine patients with spinal lesions. One of these patients, a 73 year-old man with Frankel C paresis pertaining to T10 involvement who was treated with total *en bloc* spondylectomy, was reported recently[9] and, thus, is excluded from the present study.

The clinical presentation, including preoperative and postoperative Frankel's grading, and radiological findings on presentation were reviewed in each patient. The level of vertebral involvement, results of histopathological examination, treatment modality including surgery and its results, and physical status at follow-up were studied. In these patients, no surgery-related complications occurred.

Results

Clinical presentation

Table 1 shows the demographic clinical data of our patients. One patient (case 7) with Frankel E grade, presented with impending paraesis with minimal numbness below T9 segment and a minimal urinary incontinence. All except two cases were classified as

Correspondence: H Baba

Table 1 Preoperative and follow-up clinical data

Case	*Gender/ age*	*Duration of symptoms (months)*	*Frankel's grade before*	*after*	*Follow-up (years)*	*Status*[a]
1	M/56	16	C	E	9.8	MM
2	F/58	6	D	E	9.0	CDF
3	F/47	5	D	E	8.2	NED
4	M/65	13	C	D	5.4	CDF
5	M/49	22	B	D	3.8	CDF
6	M/73	6	D	E	3.5	deceased[b]
7	F/57	4	E	E	2.5	CDF
8	M/66	2	D	E	2.0	CDF

[a]Physical status at follow-up; [b]died due to disseminated intravascular coagulopathy, unrelated to the disease. MM, multiple myeloma; CDF, continuously disease free; NED, no evidence of disease

Table 2 Site of the lesion and treatment modalities

Case	*Affected level*	*Chemo-therapy*	*Radio-therapy*	*Surgical Treatment*
1	T4	Yes	Yes	Laminectomy, and CDI[a]
2	L4	Yes	Yes	Laminectomy alone
3	T11	Yes	Yes	Anterior vertebral resection, Kaneda instrumentation
4	T3	Yes	No	Laminectomy, CDI, and iliac bone grafting (posteriorly)
5	T5	Yes	Yes	Laminectomy, CDI, and ceramic spacer replacement
6	C4	No	Yes	Anterior vertebral resection, ceramic spacer replacement
7	T8	Yes	Yes	Anterior vertebral resection, ceramic spacer replacement
8	T4	Yes	Yes	Laminectomy, CDI, and iliac grafting (posteriorly)

[a]Cotrel-Dubousset instrumentation

Frankel E grade at follow-up, or at the time of death (case 6). In one case (case 1) osteolytic lesions in the skull, ribs, ilium and the proximal part of the humerus appeared approximately 7 years after surgery. The disease process remained quiescent in two patients (case 4 and case 5) at Frankel D grade but presented with persistent urinary problems (retardation in case 4, and pollakiuria in case 5), despite considerable improvement in leg movement.

Radiological findings and treatment

The level of vertebral involvement and treatment modality including surgery are summarised in Table 2. One patient (case 6) refused melphalan therapy while another patient (case 1) was treated with melphalan and interferon for 2.8 years after he was diagnosed with multiple myeloma. A variable dose and course of radiation therapy were used in seven cases after surgery.

Anterior surgery was performed in three cases and posterior surgery in five cases. At follow-up, spinal stability was maintained in all cases with fusion or incorporation of the bone graft or ceramic spacer with the affected vertebral bodies.

Illustrative case presentations

Case 2

A 58-year-old woman was admitted complaining of a progressive lower back and leg pain, weakness of legs and a mild degree of dysuria. Neurological examination showed a hypaesthesia in the L4 and L5 dermatomes bilaterally. The strength of the leg muscles was [4^{+}/5] bilaterally. Blood chemistry was normal except for a positive M-type protein. C-reactive protein was slightly elevated (1.40 mg/100 ml^{-1}), as was the erythrocyte sedimentation rate (ESR, 55 mm/h^{-1} and 115 mm/2 h^{-1}). Bence-Jones protein was positive.

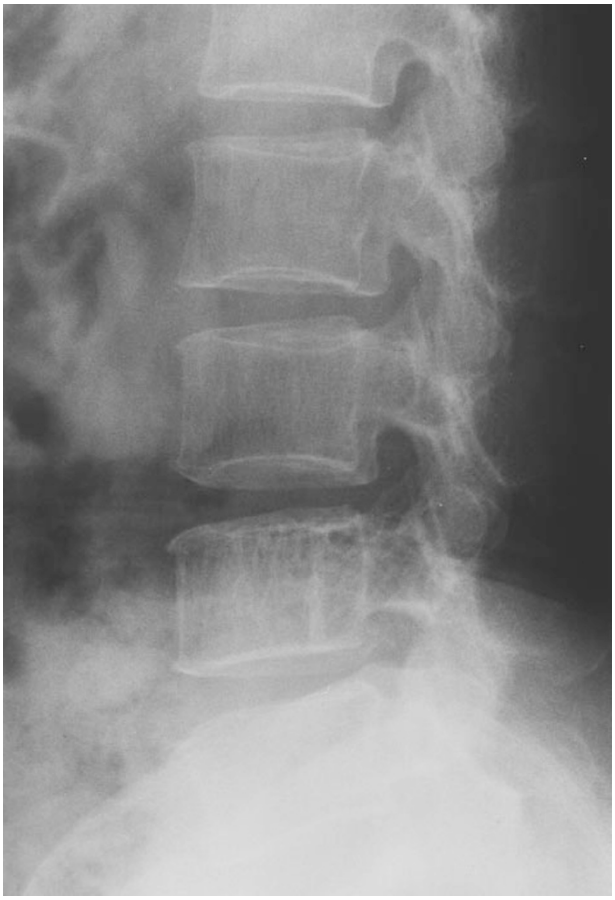

Figure 1 Lumbar spine radiograph of case 2 before surgery

Lumbar spine radiographs demonstrated 'corduroy cloth'-like appearance of the L4 vertebra (Figure 1), and computed tomography (CT) scan showed

'honeycomb'-like destruction (Figure 2). The L4 vertebra was negative on ^{99m}Technetium phosphate scintigraphy. Magnetic resonance imaging (MRI) scans showed isointensity-signals on a T2-weighted sequence and high-intensity signals on a gadolinium-diethylenetriaminepentaacetic acid contrast image. Transpedicular needle biopsy suggested the lesion to be a myeloma.

The patient refused antero-posterior combined surgery, and thus a bilateral laminectomy of L4 was performed, decompressing the L4 and L5 nerve roots as well as the cauda equina. Histology of the resected sample again showed a myelomatous lesion with irregularly clumped chromatin pattern and cluster of bizarre-type plasma cells (Figure 3). The postoperative course was uneventful. The patient then underwent bone marrow examination, but the results were all negative. Radiation therapy was then instituted (dose, 1200 Gy). A follow-up visit 9-year postoperatively showed that the patient was still healthy with no evidence of recurrence or dissemination.

Case 6

A 73-year-old man was referred to our clinic complaining of intractable arm pain and progressive muscle weakness of the extremities. A sensory deficit was detected below the C5 level and the paraesis was classified as Frankel D grade. There was a slight bladder dysfunction. Laboratory tests showed an elevated ESR (23 mm/h^{-1} and 38 mm/2 h^{-1}), and serum electrophoresis showed increased levels of M-protein.

A cervical spine radiograph showed lucent C4 vertebral collapse (Figure 4) and a CT scan showed invasion of the posterior vertebral cortex, while MRI showed the lesion compressed to the cord (Figure 5). A ^{99m}Technetium phosphate scintigraphy showed slightly abnormal uptake at C4 level, and vertebral arteriography demonstrated minimal tumour stains. Bone marrow examination was negative (plasma cells: 7% in the serum, and 6% in the iliac bone), and thus a provisional diagnosis of solitary myeloma was made.

The lesion was explored through a left-sided anterolateral approach followed by subtotal resection

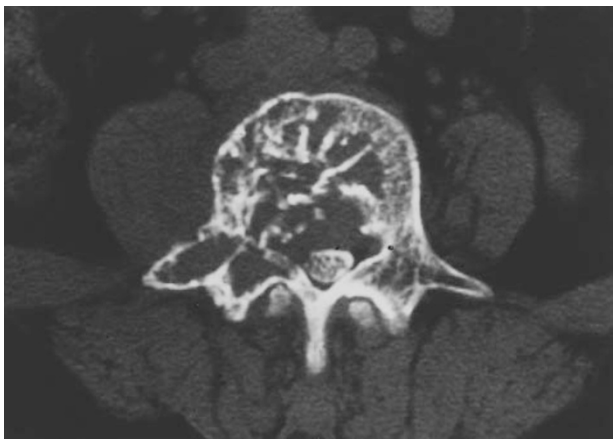

Figure 2 Computed tomography scan of case 2 showing tumourous involvement of the vertebral body, facet joint and the lamina at L4 level

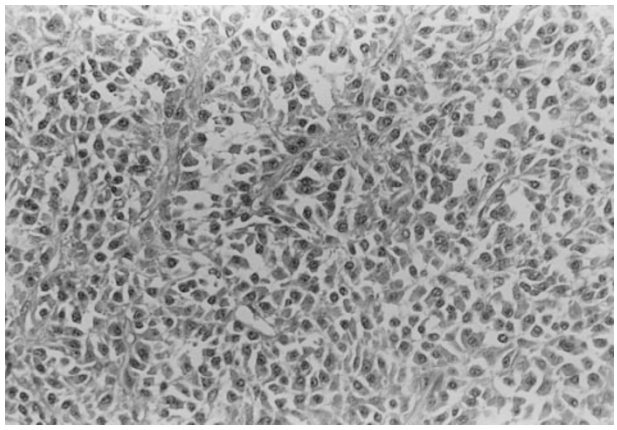

Figure 3 Photomicrograph showing irregularly clumped chromatin pattern and cluster of bizarre-type plasma cells (haematoxylin and eosin, oil immersion, $\times 100$ objective)

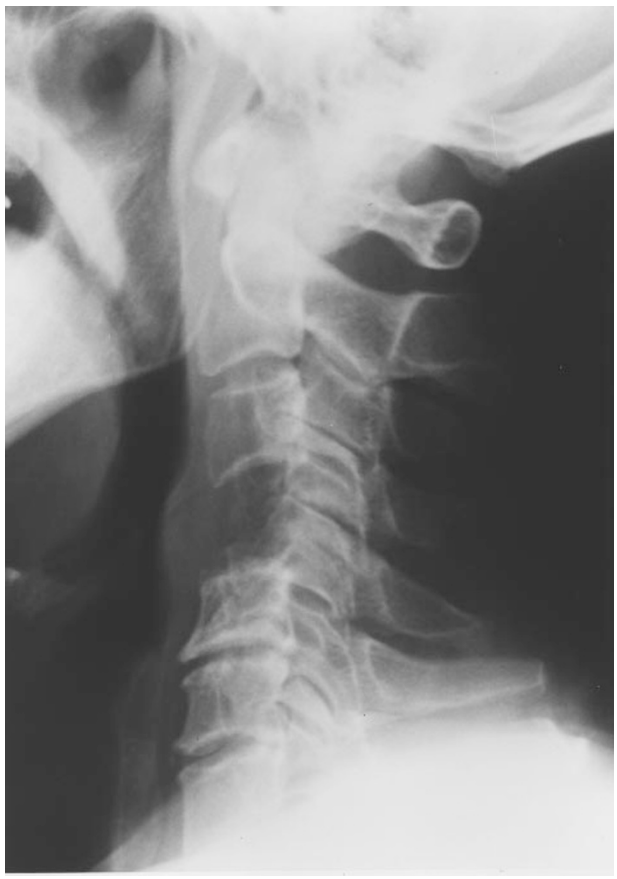

Figure 4 Cervical spine film of case 6 at presentation

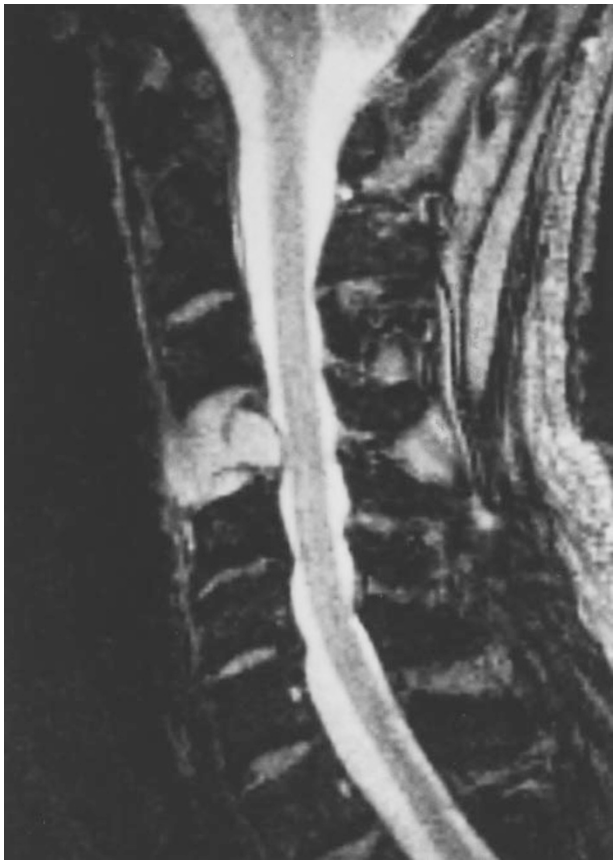

Figure 5 T2-weighted (TR, 2000 ms, TE, 100 ms) MR image of the cervical spine in case 6

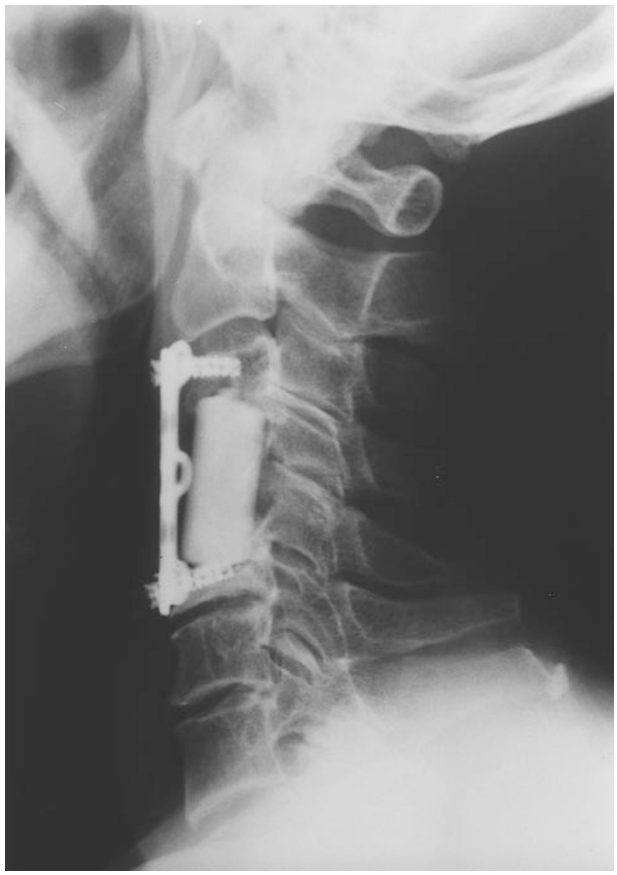

Figure 6 Cervical spine film of case 6 at 3-year follow-up

of the C4 vertebra. A reddish-purple soft tumour occupied approximately 80% of the anterior part of the C4 vertebral body and, posterior to it, a yellowish-white necrotic corticocancerous bone was present. Following resection of the tumour, a bioactive hydroxyapatite ceramic prosthesis (Pentax, Tokyo) was anchored to C3 and C5 vertebrae, and fixed using a titanium cervical plate and screws (Mathys, Bettlach, Switzerland). The histological findings were consistent with those of a relatively well differentiated myeloma and, the tumour was finally diagnosed as a solitary plasmacytoma.

Postoperatively, a total of 600 Gy (50 Gy $\times$ 12) was administered over a 4 month period, but the patient refused chemotherapy. A 3-year follow-up radiograph showed incorporation of the ceramics (Figure 6) and the patient had no recurrence or dissemination. However, the patient died suddenly 3.5 years after surgery due to disseminated intravascular coagulopathy with unknown cause.

Discussion

Solitary plasmacytoma may potentially progress to a disseminated form of multiple myeloma.[10–13] The age, gender and survival rate of patients with solitary plasmacytoma differ from those with multiple myeloma.[2,3] Though the course of multiple myeloma is usually rapid and fatal, patients with solitary plasmacytoma may live longer despite possible progression of the disease. There are no histopathological differences between the two tumours,[1] but treatment and prognosis are different. A diagnostic work-up should follow carefully and promptly to contain the malignant growth as well as preserve spinal function.

A firm diagnosis of a solitary plasmacytoma could be made in the presence of four features,[2] including (i) histological evidence of the lesion; (ii) absence of other lesions as confirmed by a complete skeletal survey; (iii) negative bone marrow examination, and (iv) absence of dysproteinemia and Bence-Jones proteinaemia. Since the solitary lesion may often disseminate, a complete skeletal survey and immunological as well as biochemical studies during follow-up (extending at least for 10 years) are important.[2] In addition, McLain and Weinstein[3] stressed the need for documenting a lack of progression or dissemination during the first 2 postoperative years following establishment of a firm diagnosis.

Solitary plasmacytoma comprises 24% of cases of plasma cell myeloma.[2] The tumour commonly occurs in the vertebra of elderly male patients (>50 years, male:female ratio = 3 : 1), but the initial diagnosis may infrequently be difficult to differentiate from aneurysmal bone cyst, giant-cell tumour, haemangioma, or metastatic spinal lesions. Myelomatous tumours, including solitary plasmacytomas, have a predilection for the vertebral body, frequently involving both the anterior and middle columns. A review of 84 cases of solitary spinal plasmacytoma by McLain and Weinstein[3] showed that 61% of the tumours occurred in the thoracic vertebrae while 18% were detected in the lumbar spine. Cervical spine involvement was only 8%.

A common clinical feature of solitary spinal plasmacytomas is pain. It is usually of a radiating type increasing in intensity with time. This is usually associated with sensory disturbances such as dysesthesia and hypaesthesia. Weakness of muscles in this condition usually indicates a spinal cord or cauda equina compromise. In this regard, Bullough and Boachie-Adjei[14] suggested that a solitary spinal plasmacytoma is more likely to cause spinal cord complications than multiple lesions. McLain and Weinstein[3] have described that in eight of 12 cases of their own series an objective neurological deficit was found: however, the authors did not describe the frequency of neurological complications in the other 72 literature-reviewed cases. Radiologically, a solitary plasmacytoma may appear as an osteolytic defect in the vertebral body without significant new bone formation, or as a sclerotic reaction, as was observed in case 2. However, as seen in this patient, the affected vertebra may infrequently show coarse trabeculation typical of a haemangioma. Additional CT and MRI studies are usually necessary to determine the exact structures affected by the pathological process and to confirm the solitary nature of the lesion.[15] To confirm the diagnosis, a biopsy and histopathological examination are mandatory, followed by bone marrow examination.

Most solitary spinal plasmacytomas can be treated by local irradiation often in conjunction with chemotherapy.[16–18] The effects of localised radiation therapy in alleviating spinal pain, spinal cord compression, and limiting extramedullary invasion have been well-established. A radiation dose less than 4000 Gy may increase the chance of local recurrence,[2] although a dose of 3000–4000 Gy has also been described to be adequate for solitary plasmacytoma.[3] During radiotherapy, however, blood tests should be performed regularly to monitor any unfavourable reaction.[19] In the present series, high-dose radiation therapy could not be applied in all patients because of dysphagia in case 2 and a rapid development of leukocytopenia in case 6. In five of the other six cases, a relatively small radiation dose (800 to 1400 Gy) was given and, though generalisation must be avoided due to the small sample size, the radiation dose could be reduced when chemotherapy and surgery are instituted. Chemotherapy is also offered as an alternative treatment. This is usually in the form of melphalan monotherapy, similar to its use in multiple myeloma. The MB2 protocol chemotherapy of Memorial Sloan-Kettering Cancer Center[2] and a combination of melphalan and corticosteroid may be reserved for patients with a disseminated disease.[20] Unfortunately, in the review of McLain and Weinstein[3] of 84 cases with the disease, however, neither the effect of chemotherapy alone on solitary plasmacytoma nor prognosis of patients treated with a combination of radiation and chemotherapy, were clearly described.

Patients presenting with neurological complications should be considered candidates for elective surgery. A combination of radio- and chemotherapy may temporarily alleviate pain caused by a collapsing vertebra destroyed by the tumour. However, surgical decompression followed by stabilisation usually produce a rapid neurological improvement. The conventional anterior route surgery, combined with irradiation and chemotherapy, is the recommended therapy. On the other hand, a tumour involving both the anterior and posterior vertebral columns can be resected in one-session posterior surgery. Tomita and his co-workers[9] reported a patient who underwent one-session posterior route total *en bloc* spondylectomy for complete resection of a solitary plasmacytoma, subsequently resulting in complete neurological recovery. From an oncological point of view, this approach is more useful and radical compared with the 'curettage of a lesion' by the anterior or anteroposterior route. A similar technique was used in the present three cases (cases 4, 5 and 8) to produce posterior decompression and instrumentation as well as resection of the affected vertebral body followed by anterior spacer (iliac bone or bioactive ceramics) grafting, and the results were successful. In the series (twelve cases) of McLain and Weinstein,[3] incomplete excision or biopsy only was performed but it appears that prognosis could be improved by increasing the dosage of irradiation. We are by no means opposing the treatment strategy recommended by that group, but as shown in case 6 in the present study, resection of the tumour as much as possible may lessen the need for chemotherapy and reduce the radiation dose.

The survival rate in solitary spinal plasmacytoma is favourable. McLain and Weinstein[3] reported a 5-year disease-free rate of 60%, although 44% of patients developed disseminated disease, 2 to 13 years after the initial diagnosis. Although these authors did not provide firm statistical comparisons, they suggested that the use of radiation therapy may influence prognosis. On the other hand, Fechner and Mills[1] showed that between 36 and 54% of patients develop multiple myeloma. They also suggested that a poorly differentiated tumour is a high risk for subsequent dissemination. While we cannot draw any conclusion from our study on survival rate and prognosis due to the small sample number, we suggest that a proper

management including early diagnosis using multimodal diagnostic imaging as well as appropriate surgery, followed by irradiation and chemotherapy may be necessary in patients with solitary spinal plasmacytoma. Irradiation of the tumour should suppress the lesion biologically, however, in the presence of a serious vertebral collapse, surgical stabilisation becomes necessary.

References

1 Fechner RE, Mills SE. Solitary (plasma cell) myeloma. In: *Atlas of Tumor Pathology: Tumors of the Bones and Joints*. Armed Force Institute of Pathology: Washington DC, 1992, pp 218–219.
2 Huvos AG. Multiple myeloma, including solitary osseous myeloma. In: *Bone Tumors: Diagnosis, Treatment, and Prognosis*. 2nd ed, WB Saunders: Philadelphia, 1991, pp 653–676.
3 McLain RF, Weinstein JN. Solitary plasmacytomas of the spine: a review of 84 cases. *J Spinal Disord* 1989; **2:** 69–74.
4 Delauche-Cavalier MC, Laredo JD, Wybier M. Solitary plasmacytoma of the spine. *Cancer* 1988; **61:** 1707–1714.
5 Di Lorenzo N *et al*. Primary tumors of the cervical spine: surgical experience with 38 cases. *Surg Neurol* 1992; **38:** 12–18.
6 Jackson A, Scarffe JH. Prognostic significance of osteopenia and immunoparesis at presentation in patients with solitary myeloma of bone. *Eur J Cancer* 1990; **26:** 363–371.
7 Meier C *et al*. Polyneuropathie, Organomegalie, Endoklinopathie und Hauptveränderungen bei einem Fall mit solitarem Myelom. *Schweiz Med Wochenschr* 1986; **116:** 1326–1331.
8 Weinstein JN. Differential diagnosis and surgical treatment of primary benign and malignant neoplasms. In: Frymoyer JW, Ducker TB, Hadler JM, Kostuik JP, Weinstein JN, Whitecloud TS III (eds). *The Adult Spine: Principles and Practice*. Raven Press: New York, 1991, pp 829–860.
9 Tomita K *et al*. Total en bloc spondylectomy: a new surgical technique for primary malignant vertebral tumors. *Spine* 1997; **22:** 324–333.
10 Corwin J, Lindberg RD. Solitary plasmacytoma of bone versus extramedullary plasmacytoma and their relationship to multiple myeloma. *Cancer* 1979; **43:** 1007–1013.
11 Knowling MA, Harwood AR, Bergsagel DE. Complication of extramedullary plasmacytomas with multiple and solitary plasma cell tumors of bone. *J Clin Oncol* 1983; **1:** 255–262.
12 Meis JM, Butler JJ, Osborne BM, Ordonez NG. Solitary plasmacytoma of bone and extramedullary plasmacytoma. A clinicopathologic and immunohistochemical study. *Cancer* 1987; **59:** 1475–1485.
13 Wiltshaw E. The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of the bone and myelomatosis. *Medicine* 1976; **55:** 217–238.
14 Bullough PG, Boachie-Adjei O. Solitary myeloma. In: *Atlas of Spinal Disease*. JB Lippincott: Philadelphia, 1988, pp 192.
15 Pozzi-Mucelli RS *et al*. La tomographia computerizzata del rachide nella valutazinone del mieloma multipo. *Radiol Med Torino* 1988; **75:** 291–296.
16 Kaplan GA, Bennet J. Solitary myeloma of the lumbar spine successfully treated with radiation: report of a case. *Radiology* 1968; **91:** 1017–1018.
17 Loftus CM, Michelsen CB, Rapoport F, Antunes JL. Management of plasmacytomas of the spine. *Neurosurgery* 1983; **13:** 30–36.
18 Valderrama JA, Bullough RG. Solitary myeloma of the spine. *J Bone Joint Surg Br* 1968; **50:** 82–90.
19 Jackson A, Scarffe JH. Upper humeral cortical thickness as an indicator of osteopenia: diagnostic significance in solitary myeloma of bone. *Skeletal Radiol* 1991; **20:** 363–367.
20 Kyle RA. Treatment of multiple myeloma: a small step forward. *New Engl J Med* 1984; **310:** 1382–1383.