SOLITARY MYELOMA

Pages with reference to book, From 116 To 118

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Although multiple myeloma is generally considered to be in the realm of internal medicine, it is important for the neurosurgeon to be familiar with the problem since, occasionally, the patient presents with paraplegia of rapid onset from extradural spinal compression. This disease was first described in 1848 by Henry Bence Jones.[1] According to World Health Organization definition. myeloma is a malignant tumour, usually showing multiple or diffuse bone involvement, and characterized by round cells related to plasma cells but showing varying degrees of immaturity, including atypical form[2]. The disease is most common between the ages of 50 and 70 years with peak incidence between 50 and 60. Males are more commonly affected than females. The lesions are associated with widespread osteolytic bone destruction and presence of abnormal proteins in the blood and urine. However, uncommonly it presents in the form of the so called ‘solitary myeloma. The criteria for acceptance as given by Bichel and Kirketer[3], are:

1. Histologic proof of the lesion.
2. Complete skeletal survey ruling out the presence of other lesions.
3. Negative bone marrow examination.
4. Neither dysproteinemia nor Benes Jones Proteinuria.

Valderrama and Bullough,[4] in a review of 47 patients with myeloma of the spine, found apparently solitary lesions in 14. Patients presenting with a solitary lesion in the spine or elsewhere have a longevity which is greater than when the disease presents in multiple areas. Pain is the first symptom and is of rather characteristic nature, being deep and persistent. The spine, including the cervical, thoracic and lumbar areas, was the site of the first symptom in 25 of 70 eatients studied.[5] Radiologically, the lesions are purely lytic with bone destruction and no evidence of regeneration. Typically the vertebra collapses and the disc spaces are preserved, a feature that helps to differentiate myeloma from spinal tuberculosis. Differential diagnosis is from metastatic disease; but spinal tuberculosis, especially the atypical forms, pose serious diagnostic problems in the geographical regions where tuberculosis is common[6]. When a solitary myeloma presents with evidence of spinal compression as did our case, the best treatment appears to be urgent surgical decompression followed by radiotherapy to the symptomatic lesion. In a previous study of solitary myeloma with long term survival of 12 patients, six of the twelve patients survived from three to twelve years before the disease became disseminated.[7]

CASE REPORT

A 61 year old Yemeni male labourer was brought to the Emergency Room of King Khalid University Hospital, Riyadh, with a two-week history of severe backache in the mid-dorsal region with a girdle like radiation around the lower chest. This pain was abrupt in onset and was initiated by lifting a heavy stone at work two weeks prior to admission. While he was lifting the weight he had a sudden “click” in th back accompanied by sudden back and girdle pain around the trunk. For five days preceding admission he noted rapidly increasing weakness and numbness of both his legs as well as frequency and difficulty of passing urine. Two days before admission he was totally bed-ridden and unable to stand. On admission he had dense paraplegia with only grade 1—2 power in most groups of muscles of leg. There was a crisp sensory level at xiphisternum with a loss of all modalities of sensation below this

level. Just before operation a catheter had to be inserted as voluntary emptying of bladder became difficult. Local tenderness and mild kyphosis were noted in mid-dorsal region of spine. Urgent X-rays of dorsal spine revealed a collapse of the seventh dorsal vertebra with preservation of the intervertebral discs on both sides (Figure 1).

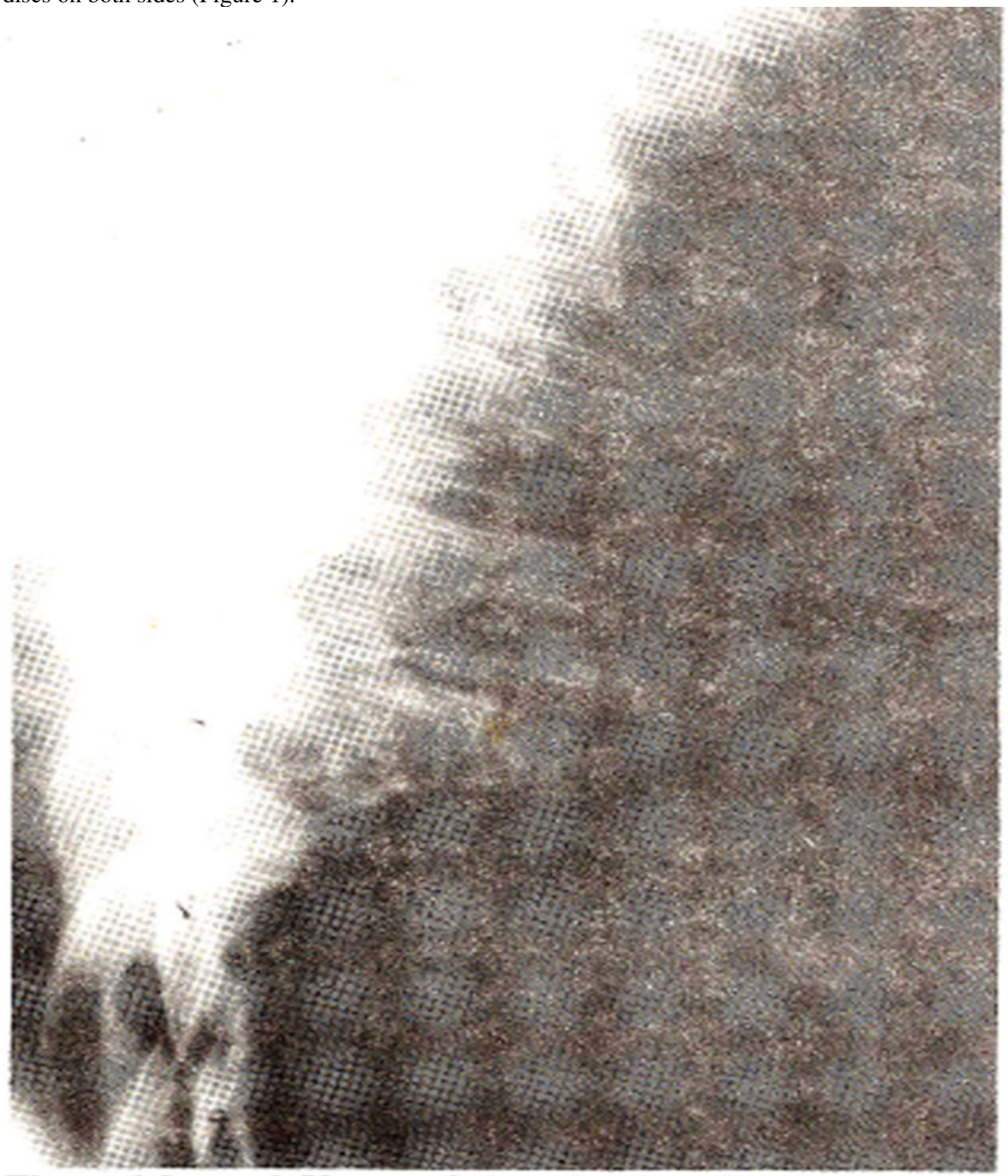

Figure 1 Lateral X-ray of dorsal spine showing collapse and ground glass appearance of 7th vertebra.

A paravertebral soft tissue shadow surrounded the compressed vertebra. Emergency myelogram was carried out and revealed a total block opposite the 7th dorsal vertebra from extradural compression (Figure 2).

Figure 2. Dorsal myelogram showing a total block at the level of 7th dorsal vertebra due to extradural compression.

The possibility of a metastasis was considered but there was no evidence of primary tumour.

Prevalence of spinal tuberculosis in the Arabian peninsula as well as a para vertebral shadow made spinal tuberculosis a likely diagnosis especially in the light of our previous experience with atypical forms of spinal tuberculosis with complete sparing of intervertebral discs.[6] Emergency laminectomy was carried out 12 hours after admission with a view to establishing the diagnosis and to decompress the spinal cord. At operation the lamina of the 7th dorsal vertebra was found to be partially destroyed and replaced by grey friable tumour-like tissue. The same tumour tissue was found to be completely filling the extradural space and surrounding the spinal cord in a cuff-like manner, causing constriction and almost strangulation of the cord. Most of the extradural compressing tumour was removed and at the end of operation the dural sheath had expanded to normal diameter and was pulsating normally. The histological examination showed a tumour composed of closely arranged round cells many of which had the features of typical myelorna cells, namely eccentric nucleus, widespread nuclear chromatin, and a perinuclear halo. Pleomorphism and multinucleation were also seen. Scattered foci of bony spicules were present. The so-called Russell bodies were not seen (Figure 3).

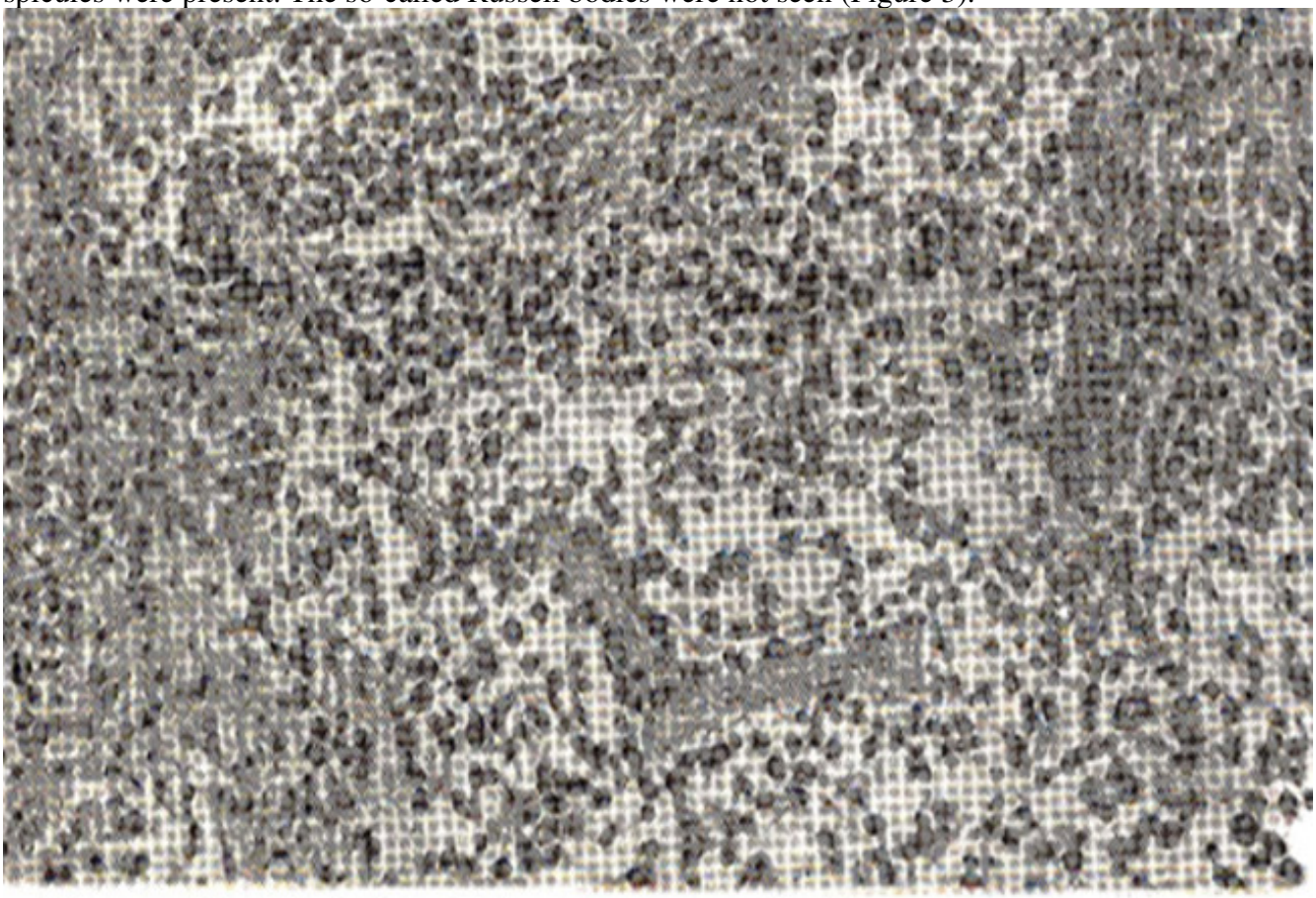

Figure 3. Histological section of the tumour showing closely arranged myeloma cells with eccentric nucleus, widespread nuclear chromatin and a perinuclear halo. Haematoxylin and Eosin x 250.

Post-operatively the neurological deficit rapidly disappeared. Motor power of grade 4-5 was noticed in all the groups of muscles in both legs on the 2nd post-operative day. Bladder continence returned so

that patient was catheter free from 3rd post-operative day. Except for poor return of position sense, all the' other modalities of sensation returned to normal range. Postoperative radiotherapy resulted in remarkable relief of local pain.
Results of further investigation were as follows:
- Bone scan showed no other bone to be involved.
- Serum protein electrophoresis was done which was normal.
- Sternal negative from the urine. bone marrow examination was Bence-Jones proteins were absent.

DISCUSSION

"Solitary" myeloma is an uncommon form of the neoplastic proliferation of plasma cells known as myeloma. A single clear-cut definition of solitary myeloma has been largely disputed. The criteria for acceptance laid down by Bichel and Kirketerp[3] have eliminated many of the purported cases. Some others restrict the acceptability of solitary myeloma to those cases that fail to show radiographic evidence of additional lesions for at least one year.[8] The most common sites of single bone involvement are the vertebral bodies, the bones of the pelvic and shoulder girdles, and, less frequently the long tubular bones of the lower extremities[9]. The long evolution and unpredictable behaviour of this tumour is at present generally accepted, in contrast to the relatively short and invariably fatal course of multiple myeloma. A single focus may remain localized for several years and finally it may disseminate[10]. Out of the 14 cases of solitary myelomas described by Valderrarna and Bullough[4] 5 of the patients were alive and well without signs of dissemination 4—14 years later, with an average survival of 10 years. In 5 cases, however, dissemination occurred 1 — 14 years after the clinical diagnosis. The prognosis is much better than in multiple myeloma. Valderrama and Bullough[4] emphasized the fact that even in the event of further dissemination the prognosis of these patients remains definitely better than that of those with initially multiple lesions. These tumours should be treated by a wide excision or block dissection with or without previous radiation. When there is some doubt about possible dissemination, or a second focus appears in a different bone, chemotherapy associated with irradiation is indicated.

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