Solitary Plasmacytoma of Dorsal Spine

Ram Chaddha, Amit Kohli, Saurav Narayan Nanda, Sanjay Kumar Tripathi, Munjal S Shah, Prashant Pradhan, Shaikh Muzammil Shiraz

ABSTRACT

Solitary plasmacytoma (SP) of bone is rare presentation and only few cases are reported in scientific literature. We report a case of solitary plasmacytoma of bone treated at our tertiary care hospital in Mumbai.

Keywords: Solitary plasmacytoma, Plasma cell infiltration, Plasma cell tumor.


Source of support: Nil

Conflict of interest: None

INTRODUCTION

Solitary plasmacytoma (SP) of bone is a rare hematologic malignancy, with radiologic evidence of a solitary lesion in the absence of significant bone marrow plasma cell infiltration (5-10%). It leads to absent (or low) serum and urine levels of monoclonal protein. Solitary plasmacytoma is the most common form of plasmacytoma and accounts for 3 to 5% of all plasma cell malignancies. This isolated lesion is most commonly found in the spine, without evidence of multiple myeloma elsewhere. It most commonly affects the axial skeleton (25-60%), such as vertebra and skull, and has a predilection for the thoracic spine.

The median age of the patients is 55 years. The male to female ratio is 2:1. Incidence rate rises exponentially by advancing age; however, it is less common at older ages in comparison with multiple myeloma (MM). The incidence rate of SP in black race is more than white race about 30%. The mean survival of patients with skeletal SP is 75% at 5-year follow-up. Spinal stability is compromised due to this osteolytic lesion. Treatment includes radiotherapy, surgery, radiotherapy combined with surgery and vertebroplasty or kyphoplasty for local control of the disease and fixation of spine for spinal stability and correction of deformity which decrease pain and can improve or prevent neurological deterioration.

CASE REPORT

A 54 years old male presented with gait imbalance with the history of upper and mid back pain for the past 3 months that was worse in morning and after prolonged sitting. There was bilateral lower limb weakness and imbalance while walking for past 10 days and patient needed support during ambulation. The symptoms had aggravated for past 1 week prior to admission.

The patient was a businessman with mostly sitting job. He had been treated with nonsteroidal anti-inflammatory drugs and physiotherapy (electrotherapy, massage, McKenzie home exercise) prior to admission.

Examination revealed broad-based shuffling gait. Tenderness was present at mid dorsal and dorsolumbar junction. Trendelenburg test and cervical slump were negative. Straight leg raise test was equal at 70° b/l. Thomas test was normal and there was full range of motion of the hips. Bilateral lower limb motor power was grade 3 to 3+. Sensory hypoesthesia was present below D7 vertebral level. Bilateral knee and ankle jerks were brisk and plantar reflexes were up going. Upper limb reflexes were normal, and distal pulsation was palpable.

The patient was from outstation and referred to tertiary hospital for further management and admitted with a provisional diagnosis of infection/primary neoplasm/secondary metastasis for further management. Routine blood investigation was normal with an erythrocyte sedimentation rate (ESR) of 9 mm/hr and C-reactive protein (CRP) of 3. Serum protein electrophoresis showed no M band. Dorsal spine X-rays were otherwise normal except for slight irregularity of D6 vertebral body. Bilateral knee and ankle jerks were brisk and plantar reflexes were up going. Upper limb reflexes were normal, and distal pulsation was palpable.

The lesion most looked like neoplastic lesion and it was suggested to perform plain computer tomographic study to rule out possibility of hemangioma. Computed tomography (CT) scan was performed and the report of dorsal spine revealed homogenous parapheyngeal (Rt) paraspinal soft-tissue lesion with course calcification at D6 vertebral body with a large mass in the Rt>Lt prevertebral region as well as in the anterior epidural space causing significant cord compression (Figs 2 to 4). The lesion most looked like neoplastic lesion and it was suggested to perform plain computer tomographic study to rule out possibility of hemangioma. Computed tomography (CT) scan was performed and the report of dorsal spine revealed homogenous parapheyngeal (Rt) paraspinal soft-tissue lesion with course calcification at D6
level associated with coarse trabecular pattern D6 vertebral body, suggestive of hemangioma and plasmacytoma (Figs 5 and 6). The patient was posted for posterior thoracic decompression with stabilization to relieve significant neural compression at D6 vertebral lesion, and surgical biopsy was planned.

To decrease intraoperative blood loss (as investigation was suggestive of vascular tumor), selective catheterization and embolization were done at D6 level with polyvinyl alcohol mucoadhesive patch preoperatively (Fig. 7). Through a posterior approach, surgical incision from D3 to D9 was given. Posterior stabilization was achieved at D4, D5, D7 and D8 level using pedicle screws (Fig. 8). Posterior decompression was done at D6 level (Figs 9 and 10). Mass was sent for histopathological examination. According to the histopathological examination report of paraffin sections revealed soft-tissue bits showing diffuse infiltration by sheets of plasma cells exhibiting eccentrically placed dense nuclei with few binucleated plasma cells (Fig. 11). The immunohistochemical study shows CD 138 positive and CD 20 negative with kappa light chain restriction by plasma cells and bony sections demonstrated plasmacytoma.

Postoperatively, patient was able to ambulate without support with the brace. To exclude multiple myeloma, biopsies from multiple sites like iliac crest, sternum and femur were also performed but showed no evidence of tumor. Detail bone survey, serum protein electrophoresis, urinary protein electrophoresis and urinary Bence-Jones protein were performed and were negative.

A final diagnosis of SP was thus made. The patient has been receiving 10 doses of radiotherapy to 40 gray (Gy) and is free from pain 3 months postoperative.

**DISCUSSION**

Solitary plasmacytoma of bone is a localized plasma-cell tumor that comprises 5% of all malignant plasma-cell tumors. It can be classified into two groups regarding to
Solitary Plasmacytoma of Dorsal Spine

Fig. 5: Computed tomography scan of dorsal spine lateral views showing course calcification with trabecular pattern of D6 vertebral body

Fig. 6: Computed tomography scan of dorsal spine axial view showing course calcification with trabecular pattern of D6 vertebral body

Fig. 7: Postembolization X-ray of dorsal spine lateral view showing polyvinyl alcohol patch at D6 vertebra level

Fig. 8: Intraoperative picture showing posterior decompression of D6 vertebra with stabilization

Solitary plasmacytoma of the bone, particularly in spine, has a better prognosis and tends to affect elderly people. Most authors consider radiation therapy, the best treatment +/- chemotherapy depending on the systemic diffusion of the disease.14,15 Half of SP turn into multiple myeloma 2 to 10 years from diagnosis (average 3.5 years).16-18 Modern therapy of multiple myeloma relies on chemotherapy. The median survival of these patients has increased to 2-3 years. Particularly, contributed to these results high dose chemotherapy (melphalan 140mg/m²).19,20 Followed by bone marrow transplantation and new drugs specifically active in the marrow microenvironment.20 The role of surgery in the treatment of plasmacytoma is to decompress cord, reduce pain and stabilize spine in order to allow weight bearing without external orthosis. From an oncological point of view, surgery can be employed to reduce or remove a tumor mass if it is low sensitive to radiation therapy and chemotherapy. Hence, improve the quality of life of patients also by a local control of the tumor mass. In the treatment of spine metastasis, in

location as SP of the bone and extramedullary plasmacytoma (EMP).12,14 Of these, 69% are osseous and the remaining 31% are extramedullary, involving soft tissue. In SP of bone, the most frequent location is the spine (68.5%). The EMP is generally observed in the head and neck and most frequently in the nasal cavity and nasopharynx.6-8 The mean survival of patients with skeletal solitary plasmacytoma is 75% at 5-year follow-up.2 Solitary bone plasmacytoma of the spine is predominantly lytic; typically involves the vertebral body and the posterior elements with compression of the cord; but, on occasion, may present as an expansile lesion with a soft-tissue mass, fractures, or, rarely, osteosclerosis.2-4

The chief complaint upon presentation is pain, which is secondary to neoplastic infiltration and/or structural loss and instability. In the Delauche-Cavallier series, 71% of patients had symptoms secondary to neurological compression at initial diagnosis. Pain and/or paralysis impair the ability of these patients to ambulate, and also impair their quality of life.
very selected cases, the best local control can be achieved by an ‘en bloc’ excision with adequate surgical margins.

Diagnosis is made by multiple investigations. Histological demonstration of plasma-cell tumor by bone biopsy. To rule out systemic tumor involvement, a bone marrow aspirate must be performed in both sternal and iliac locations. Routine investigation, i.e. white blood cell count and differential count, ESR, serum calcium level, serum alkaline phosphatase isoenzymes, serum creatinine, serum protein electrophoresis and urinalysis with Bence-Jones protein inspection. Imaging studies should include CT scan of the chest, abdomen and pelvis for primary or secondary tumor location. An MRI of the affected vertebra is important to determine the degree of tumoral compromise of the vertebra, and to quantify vertebral canal occupancy and spinal cord compression by neoplastic tissue. A CT scan to demonstrate the degree of architectural compromise as well as the presence of any bone fragments occupying the spinal canal. Technetium-99 bone scanning is also useful, but this may be positive only after a pathological fracture. The mean delay for diagnosis is one year. A prospective study by Schirrmeister et al assessed the accuracy of positron emission tomography (PET) scanning, which has the advantage of whole body imaging, in staging patients with presumed solitary plasmacytoma. Positron emission tomography is a potentially useful tool in the staging of such patients but cannot, as yet, be substituted for conventional imaging methods and currently remains an investigational tool rather than recommended for routine use. Monoclonality and/or an aberrant plasma cell phenotype should be demonstrated with useful markers being CD19, CD56, CD27, CD117 and cyclin D1.

Treatment includes: radiotherapy, surgery, radiotherapy combined with surgery and vertebroplasty or kyphoplasty. The goals of treatment for solitary plasmacytoma of the spine are local control of the disease and preservation or re-establishment of spinal stability, these help with pain management and can improve or prevent neurological deterioration.

As per recommendation, plasmacytoma should be treated by radical radiotherapy encompassing the primary tumor with a margin of at least 2 cm. The cervical nodes should be included if involved. For plasmacytoma up to 5 cm, a radiotherapy dose of 40 Gy in 20 fractions is recommended. For plasmacytoma > 5 cm, a higher dose of up to 50 Gy in 25 fractions is recommended. Knobel et al confirmed excellent local disease control with radiotherapy alone in their review of 206 patients with SBP.

Patients are re-evaluated with the measurements of M-protein and complete blood counts for progression and development of multiple myeloma. It should be repeated at 6-week intervals for the first 6 months and then with prolongation of clinic visits. If a new bone pain takes place, additional workup like appropriate imaging will be needed.
Systemic myeloma progression, being the main problem for the prognosis of the disease. The diagnosis and staging of plasmacytoma need an evaluation with more specific histological, phenotypic and radiographic methods in order to exclude occult multiple myeloma and other plasma cell neoplasm. Close cooperative work done by a team of a hematologist, a radiotherapist, and a surgeon will ensure the best results after the treatment.

REFERENCES