Vertebral plasmacytoma

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Spinal disease is observed in ~50% (range 34-72%) of cases.

The thoracic vertebrae are most commonly involved, followed by lumbar, sacral, and cervical vertebrae the rib, sternum, clavicle, or scapula is involved in 20% of cases.

see Plasmacytoma of the cervical spine.

The first case of solitary plasmacytoma of spine was reported by Shaw in 1923 .They are very rare precursor lesions for multiple myeloma. Solitary myeloma is a focal malignant proliferation of plasma cells with no evidence of diffuse marrow involvement.

Epidemiology

Peak incidence is in the 4th to 6th decades, and it is more common in males.

Clinical presentation

Physical findings are related to the site of involvement, presenting as a painful mass, pathologic fracture, or root or spinal cord compression syndrome.

The most common symptom of solitary bone plasmacytoma (SBP) is a pain at the site of the skeletal lesion due to bone destruction by the infiltrating plasma cell tumour.

Diagnosis

Patients with solitary plasmacytoma in the spine frequently require a biopsy for diagnosis of their condition.

Typically plasmacytoma presents as a single collapsed vertebra. On axial images classical mini-brain appearance may be seen due to replacement of cancellous bone with preserved cortical outline resulting in hollow vertebral body. Unlike metastatic lesions adjacent Intervertebral disc space are preserved in plasmacytoma.

Identification of a “mini brain” in an expansile lesion in the spine is characteristic of plasmacytoma. It is important that radiologists note this characteristic because biopsy can be avoided in patients with this appearance. Although biopsy might still be required at many institutions, at the Department of Radiology, Duke University Medical Center, Durham, USA., surgeons find this appearance sufficiently pathognomonic to bypass biopsy and start treatment [1)].

Diagnostic criteria for solitary bone plasmacytoma has been considered as follows:

single area of destruction due to clonal plasma cells

bone marrow plasma cell infiltration <5% of all nucleated cells

absence of osteolytic bone lesions or other tissue involvement (i.e. no evidence of myeloma)

absence of anaemia, hypercalcemia or renal impairment (usually attributable to myeloma)

low or absent serum/urine monoclonal protein

preserved levels of uninvolved immunoglobulins

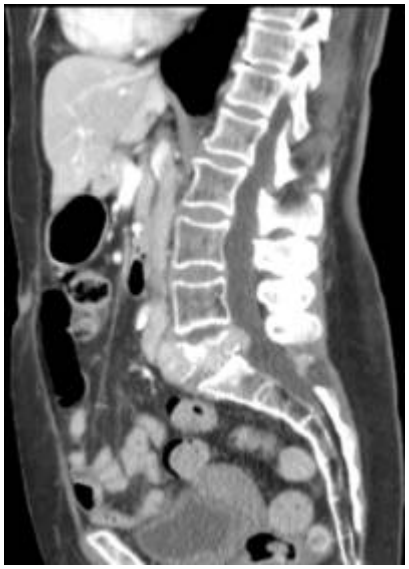
Radiographic features

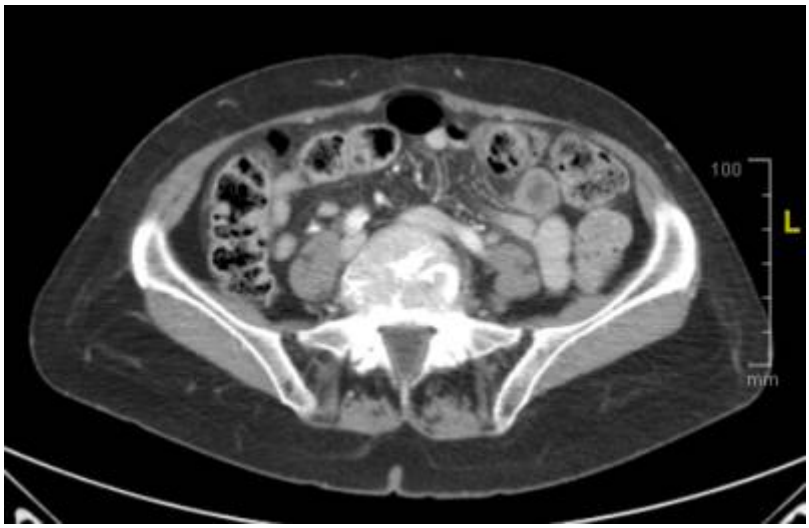
Plain radiograph

Solitary expansile lytic lesion with thinning and destruction of the cortex, and bubbly/trabeculated appearance. Characteristicaly, the absence of sclerotic reaction is seen.

CT

Expansile lytic lesion with thinned out cortex and characteristic 'mini-brain' appearance has been described in solitary vertebral lesions.





MRI

A mini-brain appearance has also been described on MRI

It is seen as curvilinear low signal intensity areas within the lesion, giving an appearance of sulci in the brain. In fact, this appearance is so characteristic that it may obviate the need for a diagnostic biopsy.

Signal characteristics include:

T1: hypo- to isointense

T2: iso- to hyperintense to muscle

T1 C+ (Gd): variable enhancement

It urges pathologists to exercise caution in cases where strong ERG-positivity implicates the presence of a prostatic neoplasia and illustrates the need for further immunohistochemical examination [2].

Differential diagnosis

The imaging mimics, acute osteoporotic compression fractures, metastasis and malignant melanoma or plasmacytoma pathological fractures are the important clinical problems in geriatric age group that need to be differentiated due to their grossly differing prognostic and therapeutic implications. There are few suggestive features on magnetic resonance imaging (MRI) that help differentiate between these entities. Hemangiomas are very common benign spinal tumors that have characteristic features on MRI. In the setting of multiple vertebral hemangiomas causing cord compression in elderly patients, the scenario is even more complex with four different entities with different prognostic profiles [3].

General imaging differential considerations include:

skeletal metastasis

chronic osteomyelitis

osteolytic metastasis

osteoid osteoma

osteoblastoma

inclusion cyst

Brown tumour of hyperparathyroidism

aneurysmal bone cyst

benign giant cell tumour

Treatment and prognosis

Excision of the tumour and its extent is done, with chemo-radiotherapy as an adjuvant. Local recurrence is less than 5% and dissemination is seen in 35-70% of patients. The disease may progress to multiple myeloma, with a bad prognosis.

Case series

Three male and four female patients were aged 40-85 years. The location was cervical spine in one patient, cervicothoracic in one, thoracic in two, thoracolumbar in one, lumbar in one, and extensive involvement in one. Progressive paraparesis and sensory disturbance were the predominant symptoms. Neuroimaging showed a compressive extradural mass lesion in the dorsal spinal canal without findings of local bone destructive changes in all cases. Four of five patients who underwent decompressive surgical maneuver and tumor resection showed neurological improvement. Immunoglobulin (IgG) kappa subtype was the most predominant histological type, followed by IgD lambda and IgA kappa subtypes. SSEP should be included in the differential diagnosis of an extradural tumor located in the dorsal spinal canal without associated bony changes. Surgery may be effective for symptomatic relief [4)].

Case series

2016

Four patients (one female, three males), mean age 58 years. There was one lesion of C1 and three of C2. Two patients with neck pain received vertebroplasty (C1 and C2, respectively) and RT as primary management. Both developed secondary instability of the CCJ after 12 and 5 months, respectively, and required occipitocervical stabilization (OCS). The other two patients underwent OCS and required no additional surgery and no signs of instability at follow-up. Forty-nine cases of OCS were published previously. Spinal stability was achieved significantly more frequently by OCS than by less invasive or medical interventional treatment options (p=.001; two-sided Fisher exact test).

Plasma cell neoplasms are highly radiosensitive. However, at the CCJ, a life-threatening instability may occur early and require surgical treatment. Based on personal experience, we favor OCS in this location. A systematic review of the literature supports this approach [5)].

2015

Zadnik et al. present the epidemiology, surgical indications, and outcome data of a series of consecutive cases involving 31 surgically treated patients with diagnoses of multiple myeloma and plasmacytoma of the spine (the largest such series reported to date).

Surgical instability was the criterion for operative intervention in this patient cohort. The Spinal Instability Neoplastic Score (SINS) was used to make this assessment of instability. The cases were analyzed using location of the lesion, spinal levels involved, Frankel score, adjuvant therapy, functional outcome, and patient survival.

All patients undergoing surgical intervention were determined to have indeterminate or gross spinal column instability according to SINS criteria. The median survival was 78.9 months. No significant difference in survival was seen for patients with higher SINS scores or for older patients (> 55 years). There was a statistically significant difference in survival benefit observed for patients receiving chemotherapy and radiation versus radiation alone as an adjuvant to surgery (p = 0.02).

In this 10-year analysis, the authors report outcomes of surgical intervention for patients with

indeterminate or gross spinal instability due to multiple myeloma and plasmacytoma of the spine with improved neurological function following surgery and low rates of instrumentation failure [6].

1998

Baba et al. 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 [7].

1983

Six patients with isolated plasmacytoma of the low thoracic or lumbar region, all of whom presented with pain and minimal neurological deficits. The approach to such cases included a complete medical work-up and radiographic definition of the lesion with a bone scan, a skeletal survey, myelography, computed tomography, and, in some instances, spinal angiography. All of these patients underwent laminectomy and spinal fusion with autologous bone and Harrington rods. There was no morbidity associated with the procedure, which was designed to prevent possible collapse during subsequent radiotherapy or chemotherapy, with its attendant potential for neurological catastrophe. Several patients have developed systemic involvement necessitating chemotherapy, but in follow-up extending now to 4 years all patients remain ambulatory and pain-free. Loftus et al. are encouraged by the potential for symptomatic relief and neurological improvement of patients with isolated spinal plasmacytomas who are treated with an aggressive medical and surgical approach [8].

Case reports

A 57-year-old man with IgG multiple myeloma and medullary plasmocytoma C7-T3, was to undergo decompressive spinal laminectomy and vertebral fixation leading to a wound dehiscence with exposed instrumentation. Autologous micrografts were obtained by Rigenera protocol and directly applied to the dehisced wound. After 60 days of negative pressure wound therapy, we observed reduction of the diameter and depth of wound dehiscence, with a coverage of instrumentation, without complete re-epithelialization, that instead was reached by application of autologous micrografts after 70 days.

The Rigenera protocol may be the solution for complex wounds in oncological and immune-compromised patients where other treatments are contraindicated [9].

A patient with plasmacytoma of the axis vertebra who underwent decompressive surgery with reconstruction via a posterior approach. The patient was referred because of quadriparesis with severe neck pain. Magnetic resonance imaging revealed a relatively demarcated, highly enhanced

mass lesion in a destructed axis, with spinal cord compression. Computed tomography revealed a 5.6×4.3 cm adrenal mass at the left retroperitoneal space.

Park et al. suspected the axis lesion to be a metastatic paraganglioma from the adrenal mass. The patient underwent total excision of the tumor under an operative microscope with occipitocervical fixation. Histopathologically, the tumor was shown to be a plasmacytoma. Following the operation, the patient recovered without significant complications. This was a rare case of plasmacytoma in the axis, mimicking metastatic paraganglioma [10].

2013

Yang et al describe one case of solitary plasmacytoma of the lumbar vertebra that was treated with surgical decompression with stabilization and additional radiotherapy. The patient had no factors associated with rapid progression to multiple myeloma such as age, size, immunologic results, pathological findings, and serum free light chain ratio at the time of diagnosis. However, his condition progressed to multiple myeloma less than two months after the initial diagnosis of solitary plasmacytoma. They suggest that surgeons should be vigilant in watching for rapid progression to multiple myeloma even in case that the patient with solitary plasmacytoma has no risk factors for rapid progression to multiple myeloma [11].

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