Plasmacytoma

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61.1 Definition

There is a wide range of clinical highlights, from monoclonal gammopathy of obscure importance to multiple myeloma (MM) to plasma cell leukemia. Most of the plasma cell tumors are analyzed as MM and will in general influence a more established grown-up population. In any case, a minority (<5%) of patients with plasma cell malignancies present with either a solitary bone mass called solitary bone plasmacytoma (SBP) or less ordinarily a soft tissue mass of monoclonal plasma cells called solitary extramedullary plasmacytoma (SEP).

SBP is a plasma cell problem described by a localized collection of neoplastic monoclonal plasma cells in the bone. SBP has a male/female ratio of 2:1, with an average age of 55 years. Solitary type essentially involves axial skeleton, particularly the vertebrae.

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Malignant bone tumors of the spine are extremely uncommon. SBP is the most common separate tumor inside this gathering, representing around 30% of the aggregate. These tumors place in the spinal bony structures twice as frequently as other bones.

61.2 Natural History

SBP has a high incidence of progression to MM, and on the magnetic resonance imaging (MRI) assessment, at any rate, 25% of patients with an evident solitary mass have proof of disorder somewhere else. Interestingly, SEP is almost in every case genuinely restricted and has a high healing rate with local treatment options. Most of the patients with obvious SBP progress to myeloma, with a middle opportunity to the progression of 2 to 4 years. The middle by and large endurance in various studies fluctuates from 7.5 to 12 years.

If a MM patient has features including low levels of uninvolved immunoglobulins, axial disease, older age, lesion size >5 cm, and persistence of the M protein after treatment, it will result in disease progression. The presence of M protein has been accounted for in 24% to 72% of patients in various studies. The recurrence presumably relies upon the degree of affectability of the tests utilized.

61.3 Physical Examination

The most common manifestation is the pain at rest (Chap. 41); however, it can likewise present spinal cord or root compression findings (Video 61.4). Inclusion of the base of the skull can give cranial nerve compromise (paralysis).

61.4 Imaging

Just as the proper blood and urine tests, imaging of the spine is required and best accomplished by MRI and computed tomography (CT) scan. A mini-brain appearance has also been described in MRI (Fig. 61.1). It is seen as curvilinear low-signal-intensity areas within the lesion, giving an appearance of sulci in the brain [1]. Indeed, this appearance is a trademark to such an extent that it might block the requirement for a symptomatic biopsy. The biopsy is generally conceivable percutaneously, guided either by fluoroscopy or CT (Video 61.8). Positron emission tomography (PET) has as of late been assessed in the organizing of patients with myeloma and plasmacytoma. PET, similar to MRI, seems to be valuable in identifying occult disease in patients with SBP.

There are no detailed rules to characterize contribution on MRI scans with regard to evident SBP. Be that as it may, the MRI appearances in MM have been very much portrayed. The presence of at least one foci of anomalous sign force, for example,

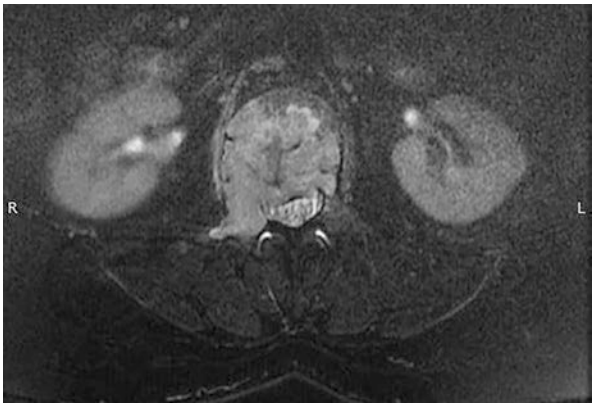

Fig. 61.1 A case with "mini-brain" appearance in axial MRI (L2 vertebrae)

low on T1-weighted imaging and high on T2-weighted or STIR (short TI inversion recovery) images, which upgrade after the organization of paramagnetic differentiation without realized late pressure cracks, other essential danger, or average attributes of generous or harmful essential bone tumors, is viewed as proof of removed association in patients with obvious SBP.

61.5 Differential Diagnosis

The accompanying measures are suggested:

- Single region of bone destruction because of plasma cells.
- Histologically <5% plasma cells in bone marrow suction.
- Totally normal outcomes on skeletal overview including radiology of long bones.
- No hypercalcemia or renal debilitation due to plasma cell dyscrasia.
- Missing or low serum or urinary degree of monoclonal immunoglobulin (level of >20 g/l) dubious of MM.
- No extra lesion on MRI of the spine.

SBP is for the most part diagnosed by biopsy or fine-needle aspiration. Percutaneously guided biopsy of the spine is typically conceivable either by fluoroscopy or CT (Videos 61.7 and 61.8). As these tumors are uncommon, pathology survey by a pathologist with an exceptional interest in either bone tumors or lymphoproliferative issues is firmly suggested.

61.6 Treatment Options

Although the mainstay treatment for MM is systemic chemotherapy, radiotherapy often has an important supportive role, offering very effective symptom relief for plasmacytomas in bone or soft tissues. Radical radiotherapy is the preferred

treatment for SBP. As with other rare tumors, the evidence base for treatment consists largely of retrospective studies of a small number of patients. Progression to MM is common, despite the high local control rates of 83% to 96% achieved with medium-dose radiotherapy. Based on the results of retrospective case studies, a dose of 40 Gy in 20 fractions is recommended for SBP. For SBP larger than 5 cm in size, a higher dose up to 50 Gy in 25 fractions should be considered [2]. An alternative strategy for tumors larger than 5 cm is chemotherapy followed by radiotherapy. This is theoretically attractive, but there is little published evidence to support it.

Radiotherapy remains the treatment of choice of the primary pathology, and surgery is contraindicated in the absence of structural instability or neurological compromise. However, early diagnosis and referral for a neurosurgeon/orthopedic surgeon opinion is advised in most cases with spinal involvement. Due to the development of modern spinal fixation systems over the last decade, surgical treatment is now a viable and successful option for patients who develop pain caused by structural compromise within the vertebra, vertebral instability, neurological compromise, or a combination of these.

Close contact between hematologist, radiotherapist, and neurosurgeon/orthopedic surgeon is along these lines essential in planning ideal treatment options for patients.

61.7 Expected Outcomes

As a radiotherapy and/or chemotherapy-sensitive neoplasm, the prognosis of plasmacytoma with conservative treatment is satisfactory in general.

61.8 Potential Complications

Spinal instability and neurological compromise are extremely rare.

61.9 What Should Patient and Family Know?

SBP is a rare disease in the spectrum of plasma cell dyscrasias. Solitary lesions have a high potential to transform into MM.

Further Readings

1. Ferreira-Filho LA, Pedroso JL, Sato EA, et al. Teaching neuroimages: "mini brain" sign: a radiologic marker for vertebral solitary plasmacytoma. Neurology. 2014;82(23):e210–1.
2. Soutar R, Lucraft H, Jackson G, et al. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. Br J Haematol. 2004;124:717–26.