Case Report

Solitary Plasmacytoma of Bone Involving Spine in a 12-year-old Boy: Report of a Rare Case and Review of Literature

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Introduction

Plasma cell neoplasms are malignant tumors characterized by uncontrolled proliferation of plasma cells. It generally presents as generalized disease known as multiple myeloma (MM) at diagnosis. However, <5% patients present with solitary lesions known as solitary plasmacytoma of bone (SPB).[1] SPB presents more commonly in elderly individuals with a median age of 55 years and predominantly affects the axial skeleton, especially the vertebrae, ribs, and pelvis.[2] SPB in young individuals has been very rarely reported. Here, we present a 12-year-old boy who presented with backache and lytic lesion in L5 vertebra which was initially misdiagnosed as large cell lymphoma but finally turned out to be solitary plasmacytoma.

Case Report

A 12-year-old boy presented with lower back pain, radiating to the left lower limb for 1 month. He was initially investigated with plain radiograph of the dorsolumbar spine which was normal and was treated with analgesics but with no relief. Hence, magnetic resonance imaging (MRI) of the lumbosacral spine (LS) was done which showed lytic lesion in L5 vertebral body and left pedicle, transverse process, and superior articular facet [Figure 1a]. Hence, the patient was referred to our hospital for further investigations and treatment. Computerized tomography (CT)-guided biopsy from the vertebral lesion was suggestive of malignant round cell tumor, morphologically large cell lymphoma [Figure 2a]. Immunohistochemistry (IHC) study revealed leukocyte common antigen, CD79a and multiple myeloma oncogene-1 (MUM1) positive with CD 20, AE 1, and Bortezomib, chemotherapy, lenalidomide, recurrent, solitary bone plasmacytoma, spine, youngest

Keywords: Bortezomib, chemotherapy, lenalidomide, recurrent, solitary bone plasmacytoma, spine, youngest

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CD 99 negative and MIBI of 20% [Figure 2b-g]. Bone marrow aspiration and trephine bone biopsy showed normocellular marrow uninvolved by any malignancy. Cerebrospinal fluid examination and cytology was within normal limits. Complete blood counts, liver and renal function tests, serum protein electrophoresis and immunofixation, beta 2 microglobulin, urine BJ proteins, serum free light chain (FLC) assay, and skeletal survey were all within normal limits. At this stage, considering the clinical presentation and young age, diagnosis of CD 20 negative large cell lymphoma (Stage IIE) was favored based on morphology and IHC, and the patient was treated with three cycles of cyclophosphamide, adriamycin, vincristine, and prednisolone with intrathecal methotrexate. After three cycles, MRI was suggestive of residual lytic lesion and involving L5 vertebra. The patient was given curative radiotherapy (RT; 40 Gy divided in 20 fractions). MRI after RT was suggestive of post-RT changes and hence the patient was kept on observation [Figure 1b]. After 9 months, the patient again presented with backache and sudden onset paraparesis. Repeat MRI of the LS and pelvis with whole spine screening was suggestive of recurrent lesion with large paraspinal soft-tissue component at L5 vertebral level [Figure 1c]. The patient underwent emergency decompressive laminectomy and partial excision of paraspinal mass. Histopathological examination of laminectomy specimen showed proliferation of mature binucleated plasma cells (plasmablasts) [Figure 3a]. Further IHC confirmed MUM1 strong positive, epithelial membrane antigen, and CD 138 positive in mature plasma cells [Figure 3b-d] with CD 20, CD 99, and CD 1a negative. Repeat bone marrow examination and myeloma workup including FLC ratio were negative. Previous pathological slides and biopsy blocks were again reviewed with additional IHC markers and revealed CD138 positive and kappa light chain-restricted plasma cells, thus favoring plasmacytoma. Thus, the diagnosis of recurrent solitary plasmacytoma of the spine was confirmed. Postlaminectomy MRI revealed residual paraspinal lesion extending to L4–L5 level compressing the exiting nerve roots [Figure 1d]. In view of prior curative RT received 9 months back, repeat RT or boost was not feasible. Hence, the only option left was to treat with systemic antimyeloma chemotherapy. The patient was started on bortezomib, lenalidomide, and dexamethasone (BLD) regimen. Repeat MRI after four courses showed significant regression of paraspinal mass with complete neurological recovery [Figure 1e]. Hence, BLD regimen was continued. After completion of nine courses, contrast-enhanced CT scan of the LS spine was performed which showed complete resolution of paraspinal mass and no evidence of any abnormal lytic lesion at the operated site [Figure 1f]. Since there is paucity of data regarding treatment of myeloma in children and use of lenalidomide and bortezomib, the patient has been started on bortezomib maintenance and is under close follow-up for any postchemotherapy complication and toxicities.

**Discussion**

SP is defined as biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cells.[3] SP has been classified into two types: (1) osseous (2) nonosseous primary lesions. SPB is a localized plasma cell neoplasm that most commonly presents as an expansile lytic mass mainly localized within the axial skeleton. Thoracic spine is the most common site for SPB while the upper respiratory tract is the most common location for

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**Figure 1:** (a) Magnetic resonance imaging showing lytic lesion involving L5 with epidural soft tissue. (b) Postradiotherapy changes after completion of curative radiotherapy. (c) Recurrence in L5 with large epidural soft-tissue component. (d) Residual lesion after decompressive laminectomy and excision. (e and f) Magnetic resonance imaging and computer tomography scan showing complete resolution of lesion after nine courses of bortezomib, lenalidomide, and dexamethasone regimen

**Figure 2:** (a) Computed tomography biopsy showing malignant round cell tumor morphologically large cell lymphoma (b) leukocyte common antigen positive (c) CD 79a positive (d) multiple myeloma oncogene-1 positive (e) CD 20 negative (f) AE 1 negative (g) CD 99 negative
extramedullary lesions.\textsuperscript{[3]} SPB constitutes 70\% of SP and occurs primarily in marrow-containing bones. It has a male/female ratio of 2:1, median age of 55 years (about 10 years younger than patients with MM).\textsuperscript{[2]}

Pediatric spinal malignant tumors are very rare and SPB of the spine in pediatric population being even rarer. Most common tumors seen in the spine in children are Ewing’s sarcoma, lymphoma, neuroblastoma, osteosarcoma, or very rarely metastasis of primary distant malignancies.\textsuperscript{[3]} SPB involving spine is extremely rare in young individuals with only few cases reported in literature below 20 years of age. Extensive literature search revealed that the youngest cases reported till now of SPB are of 14 years of age [Table 1].\textsuperscript{[2,4‑9]} Extramedullary plasmacytoma and multiple plasmacytomas have been reported at very young age as low as 8 years.\textsuperscript{[10,11]} However, to the best of our knowledge, this is probably the youngest case of “solitary” osseous plasmacytoma reported in literature.

The exact pathogenesis of pediatric SPB is unknown. An association of preceding trauma leading to increased release of cytokines such as interleukin 6 causing increased proliferation of plasma cells and stromal cells in the bone has been mentioned in one of the studies.\textsuperscript{[3]} However, there was no history of any antecedent trauma in our case.

Kumar \textit{et al.} have shown that high-grade angiogenesis seen in SPB correlated strongly with progression to MM.\textsuperscript{[12]} Thus, angiogenesis plays an important role, and targeting angiogenesis could be a novel therapeutic approach in the treatment of SPB. Therefore, antiangiogenic compounds such as thalidomide, vascular endothelial growth factor inhibitors, and proteasome inhibitors have been proposed to be useful in this disease.\textsuperscript{[2]}

The diagnosis of SPB is based on recognition of monoclonal plasma cells and correlation with clinicoradiological findings to exclude systemic involvement. The International Myeloma Working Group has laid down diagnostic criteria for SP and requires all the following to be fulfilled:\textsuperscript{[13]} (1) Biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cells, (2) Normal bone marrow with no evidence of clonal plasma cells, (3) Normal skeletal survey and MRI (or CT) of the spine and pelvis (except for the primary solitary lesion), and (4) Absence of end-organ damage such as hypercalcemia, renal insufficiency, anemia, or bone lesions (CRAB) that can be attributed to a lymphoplasma cell proliferative disorder.

The distinction between neoplastic cells with plasmacytic differentiation in mature B-cell lymphomas versus neoplastic plasma cells in plasma cell neoplasms is difficult. Plasmacytoid cells in lymphoma are more likely to express CD19, CD45, and surface immunoglobulin and less likely to express CD 56 than those in myeloma. Myeloma cells express CD79a, CD138, CD38, and MUM1 which was seen in our case.\textsuperscript{[10]}

Curative RT with doses up to 40–50 Gy has been the standard of care in SPB providing a high local control rate (83\%–96\%). The target volume defined on MRI should include a margin of at least 2 cm.\textsuperscript{[11]} For patients

\begin{figure}
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\includegraphics[width=\textwidth]{figure3.png}
\caption{(a) Laminectomy specimen showing mature binucleate plasma cells (plasmablasts) (b) epithelial membrane antigen positive (c) CD 138 positive (d) multiple myeloma oncogene-1 positive suggestive of plasmacytoma}
\end{figure}

\begin{table}
\centering
\caption{Previously reported cases of solitary plasmacytoma of the bone younger than 20 years of age}
\begin{tabular}{|l|l|l|l|l|l|}
\hline
Reports published & Age & Sex & Site & Treatment given & Outcome \\
\hline
Pavithran \textit{et al.}, 1997\textsuperscript{[3]} & 16 & Female & Head of humerus & RT (42 Gy) & CR (10 years F/U) \\
Bertoni-Salateo \textit{et al.}, 1998\textsuperscript{[6]} & 17 & Female & Tibia & - & - \\
Boos \textit{et al.}, 1997\textsuperscript{[7]} & 16 & Female & Lumbar spine (L2) & Surgery + RT (36 Gy) & CR (18 months F/U) \\
Gossios \textit{et al.}, 2002\textsuperscript{[8]} & 18 & Male & Thoracic spine & - & - \\
Panteli \textit{et al.}, 2002\textsuperscript{[9]} & 18 & Male & Thoracic spine & - & - \\
Dumesnil \textit{et al.}, 2006\textsuperscript{[4]} & 14 & Female & Lumbar spine (L2) & RT (30.6 Gy) + CT (VAD) + ASCT & CR (10 years F/U) \\
Kumar \textit{et al.}, 2011\textsuperscript{[2]} & 14 & Female & Tibia & RT (45 Gy) & CR (34 months F/U) \\
Our case & 12 & Male & Lumbar spine (L5) & RT + surgery + CT (BLD) & CR (on maintenance CT) \\
\hline
\end{tabular}
\end{table}

RT: Radiotherapy, CT: Computerized tomography, VAD: Vincristine-adriamycin-dexamethasone, BLD: Bortezomib, lenalidomide, and dexamethasone, F/U: Follow-up, CR: Complete remission, ASCT: Autologous stem cell transplant
with a vertebral involvement, the irradiation should be extended to the two adjacent vertebrae with moderate doses. With this radiation dose, 30% of patients with solitary bone lesions achieve long disease-free survival with local tumor recurrence rate <10%.\[11\] Emergency surgical intervention like decompressive laminectomy may be required in situations such as spinal cord compression or neurological compromise.\[4,14\] Role of adjuvant chemotherapy in SPB is still controversial with some reports suggesting that adjuvant chemotherapy after definitive surgery/RT may delay progression to MM with prolongation of overall survival but others observed no benefit.\[15\] Only one randomized controlled study suggested benefit on disease-free survival from prolonged adjuvant melphalan and prednisolone chemotherapy given for 3 years after RT.\[16\] Dumesnil et al. have used adjuvant chemotherapy (vincristine, adriamycin, and dexamethasone) followed by autologous stem cell transplant (ASCT).\[4\] Recurrent solitary plasmacytoma at different sites may be treated with additional RT, but patients with more extensive disease or early relapse may benefit from systemic therapy and/or ASCT, as indicated for myeloma.\[17\] Newer agents including thalidomide and bortezomib have also been used successfully, before transplantation, in small numbers of patients with relapsed plasmacytoma.\[18,19\]

In our patient, systemic antimyeloma therapy was preferred in view of early recurrence at same site. Extrapolating the data from adult MM, the patient was started on BLD regimen since it is the current first-line standard regimen for adult MM.\[13\] The patient tolerated chemotherapy very well. To the best of our knowledge, this is the first case report using novel agents such as bortezomib and lenalidomide in pediatric patient with excellent outcome. Since there is a lack of long-term safety data of bortezomib and lenalidomide in pediatric population, the patient is currently being followed up closely for any sign of toxicity or progression to systemic myeloma.

The prognosis is usually poor for SPB as compared to extramedullary plasmacytomas. However, SPB in younger patients is more indolent, and progression to MM has been reported to be less frequent in younger patients, whose survival is also better.\[5,4\]

The failure pattern in most patients with SPB is either local recurrence or progression to MM. More than 75% patients with apparent SBP progress to MM, with a median duration of 2–3 years, and this proportion increases with passage of time. MM may even develop as late as 15 years after RT.\[2\]

**Conclusion**

SPB though very rare can occur in young age. Age alone should not defer the clinician or the pathologist from diagnosing SPB. High index of clinical suspicion is required for early diagnosis of pediatric SPB. If strongly suspected, appropriate and expanded IHC panel for confirming the presence of clonal plasma cells should be ordered to avoid misdiagnosis. Novel agents in myeloma treatment such as bortezomib and lenalidomide can be used with good results like in our patient in case of post-RT recurrence though further studies are warranted for long-term side effects and outcome of these agents in pediatric population.

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**Conflicts of interest**

There are no conflicts of interest.

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