CASE REPORT

SOLITARY BONE PLASMACYTOMA: A CASE REPORT FROM A PRIVATE SPECIALIST REFERRAL CENTRE IN PORT HARCOURT

Aaron FE[1], Ijah RFOA[1*], Omunakwe HE[2]

[1]Department of Surgery, Rivers State University, Nigeria
[2]Department of Haematology and Blood Transfusion, Rivers State University, Nigeria

***Correspondence:** Ijah RFOA ; **Email:** rexijah@gmail.com

Abstract

Introduction: Multiple myeloma is uncommon in Nigeria with about 10% of cases presenting with paraplegia. The aim of this study was to provide a description of a case report of spinal myeloma seen and treated at a private specialist referral hospital in Port Harcourt from 2016 to 2023.

Case Presentation: A 56-year-old male professional diver presented in 2016 with complaint of progressive low back pain of two years duration of insidious-onset and associated with paraparesis. Physical examination and laboratory investigation findings were in-keeping with a diagnosis of solitary bone plasmacytoma of the spine. Patient responded well to novel systemic chemotherapy and has been on out-patient follow-up.

Conclusion: Plasmacytoma can occur in any part of the body, and spinal plasmacytoma affecting the spine can cause paraparesis. Prompt diagnosis and appropriate treatment can improve disease free survival, and use of novel systemic chemotherapy is a viable option.

Keywords: Low Back Pain, Multiple Myeloma, Solitary Bone Plasmacytoma, Spine, Port Harcourt, Nigeria

Cite as*: Aaron FE, Ijah RFOA, Omunakwe HE. Solitary Bone Plasmacytoma: A Case Report from a Private Specialist Referral Centre in Port Harcourt AJRMHS. 2024;2(1):26-32*

INTRODUCTION

Samuel Solly, a distinguished British surgeon and educator, is credited with the first well-documented clinical description of Multiple Myeloma in 1844,[1] and further historical records are highlighted in the works of other researchers.[2,3] Multiple Myeloma (MM), is the second commonest hematologic malignancy of plasma cell origin.[4,5] Its incidence is rising accounting for about 15% of all lymphohematopoietic cancers and 1-2% of all cancers globally.[6] Worldwide about 155,688 cases of Multiple Myeloma was reported in year 2019, with 54.3% male involvement and a current ranking as the most frequent hematologic cancer. The median age at diagnosis is 70 years, with high incidences seen in Australia, North America, Europe, and New Zealand, and low incidence in Asia.[7,8] Multiple myeloma is not so common a malignancy in Nigeria, accounting for 102 (0.8%) of newly diagnosed cancers, 5.6% and 12.9% of all hematologic malignancies.[9] Multiple myeloma presents commonly with weight loss, bone pain, fatigue, anemia, hypercalcemia, infection and renal dysfunction, and less commonly with hyper-viscosity, splenomegaly, hepatomegaly, and spinal cord compression.[10,11] Paraplegia however, is an uncommon feature seen 10% of cases.[12] In year 2013, 32 cases were reported in Enugu Nigeria in a retrospective study.[13] In that study, the median age of patients was 62 years, and late presentation was a feature. Modern care of multiple myeloma involves the use of induction chemotherapy, immunomodulatory drugs, proteasome inhibitors, and stem cell transplant, of which only 4% of these patients in Nigeria are able to afford the stem cell transplant outside our shores.[14,15] Newer approaches to management are being suggested,[16,17] and newer drugs have been approved for its care.[18,19,20]

In solitary bone plasmacytoma however, there is localized (e.g. bone) proliferation of neoplastic monoclonal plasma cells with no systemic features.[21,22] A recent publication reported primary bone tumors to be rare with Solitary Bone Plasmacytoma (SBP) accounting for 30% of all cases,[23] and 3%–5% of all plasma cell neoplasms.[24] The treatment for SBP reported globally has been by radiotherapy or in combination with chemotherapy.[20,25] Solitary Bone Plasmacytoma (SBP) is an infrequent occurrence, with few reports in the literature in Nigeria. Earlier reports of solitary bone plasmacytoma in Nigeria were treated with radiotherapy,[26] and surgery when amenable.[27] We therefore present our experience of a rare case of SBP of the lumbar spine diagnosed and treated with chemotherapy from 2016 to 2023 (7 years), as a case report.

CASE PRESENTATION

Clinical History*: A* 56-year-old male professional diver presented on the 21st of August 2016 with complaint of insidious-onset progressive low back pain of two years, and inability to walk for a year six and months duration. He had sought help from traditionalists and herbal medications were used without sustained improvement. There was no associated faecal or urinary incontinence, and no other significant information from the history. Patient was reviewed by orthopaedic surgeon, neurosurgeon, and haematologist.

Physical Examination*:* Examination findings were essentially a middle-aged man unable to stand and walk. Sensation was present in both limbs, and power was 3/5 in all lower limb muscle groups. Other systems were essentially normal.

Investigations*:* The *Full Blood Count (FBC)* revealed relatively elevated Red Cell Distribution (packed cell volume = 16.1%), white cell count was 12.20 x 10^9/L, and the eosinophil was 4.0%. The *Serum Electrolyte, Urea and Creatinine* was essentially normal except for marginally low potassium value (3.0mmol/L). *Liver Function Test* showed normal values. The serum calcium was 2.10 (2.25-2.75mmol/L). *Radiograph* of the lumbosacral spine showed non-visualization of the L1-L2 bony elements. *Magnetic Resonance Imaging* of the lumbosacral spine revealed a mass lesion at L1-L3 that completely enveloped the L1-L3 bony elements with thecal compression, but no intradural involvement. *Bone Marrow Aspiration* showed a normal marrow for the age with normal erythropoiesis, sequential myelopoiesis, normal lymphopoiesis. Megakaryocytes were normal, plasma cells made up about 3% of marrow nucleated cells, there were no abnormal cells in the marrow smear. Open (wedged) biopsy was done in the lumber spine and the histopathologic report showed a tumour composed of diffuse sheet of malignant plasma cells infiltrating the fibro-collagenous tissue, the tumour cells are fairly uniform with eccentric nucleus and eosinophilic cytoplasm, no neural tissue seen, indicative of Plasmacytoma. The *Immunohistochemistry profile* showed: CK 5/6 Cytokeratin marker – negative, EMA epithelial marker – negative, LCA lymphoid marker – positive, CD138 plasma cell marker – positive, and S100 Protein Neural Marker – negative. Protein electrophoresis done revealed: S-Total Protein 73 (66-83g/L), S-Albumin value of 27 was low (53-52 g/L), S-Alpha-1-Glob. 2.5 (2.1-3.5g/L), S-Alpha-2-Glob. 8.4 (5.1-8.5g/L), S-Beta-Glob. Value of 5.5 was low (6.0-9.4g/L), S-Gamma-Glob. Value of 26.3 was elevated (8.0-13.5g/L). Serum Paraprotein concentration was 24.0g/L, and the

Albumin-Sebia value of 30.4 was low (35.0 – 52.0 g/L). The electrophoretic pattern reveals a monoclonal peak (24g/L) in the GAMMA region. A diagnosis of Solitary Bone Plasmacytoma affecting the L 1- L3 region of the spine was made.

Treatment*:* Based on the above diagnosis, considerations for treatment were local radiotherapy and systemic chemotherapy. However, the use of local radiotherapy was not possible, because of lack of a functional radiotherapy service within our region of practice at the time, and the patient was too ill to travel over 900 kilometres from Port Harcourt to the closest centre where it could have been done. The decision was made to commence systemic chemotherapy with subcutaneous Bortezomib 2mg weekly, tablets Dexamethasone 40mg weekly for 4 weeks and Lenalidomide 25 mg daily for 21 days. The plan was to administer this for 6 – 8 cycles while observing the patient's response, the aim of which was palliation and improvement of the patient's quality of life. Additional treatment was the use of tablets Allopurinol 300mg daily and Zoledronic acid 4mg in 250mls of Normal Saline monthly. After the 3rd cycle, he achieved neurological recovery and was able to move his lower limbs, power increased to 4/5, there was complete remission of pain and physiotherapy was commenced. By the end of the 6th cycle, the radiological investigations and immunological investigation were repeated. The mass enveloping the L1-L3 was not visualizable on the repeat MRI and there was no abnormality in the Free Light Chains, no serum paraprotein detected. We continued the chemotherapy to the 8 cycles and supported with physiotherapy for the same period as well. Chemotherapy was continued to the 8th cycle, and he had complete neurological remission. He was able to walk with the aid of a Zimmer frame, and a repeat of baseline tests - EUC, FBC, SPE showed no abnormalities. A radiograph (x-ray) of the lumber spine was able to visualize L1 and L2, however there were areas of hypodensity, and MRI did not visualize any mass in L1-3. Chemotherapy was discontinued and he was placed on maintenance therapy with capsule lenalidomide 25mg daily for 21days and pulsed tablet dexamethasone 40mg monthly for a period of one year.

Outcome: The patient continued to improve clinically as he was followed up every 6 months over the past 7 years. He is currently bearing weight, walking unaided and able to drive. A recent review of his FBC, EUC, MRI at the 5th year of his follow-up showed significant improvement.

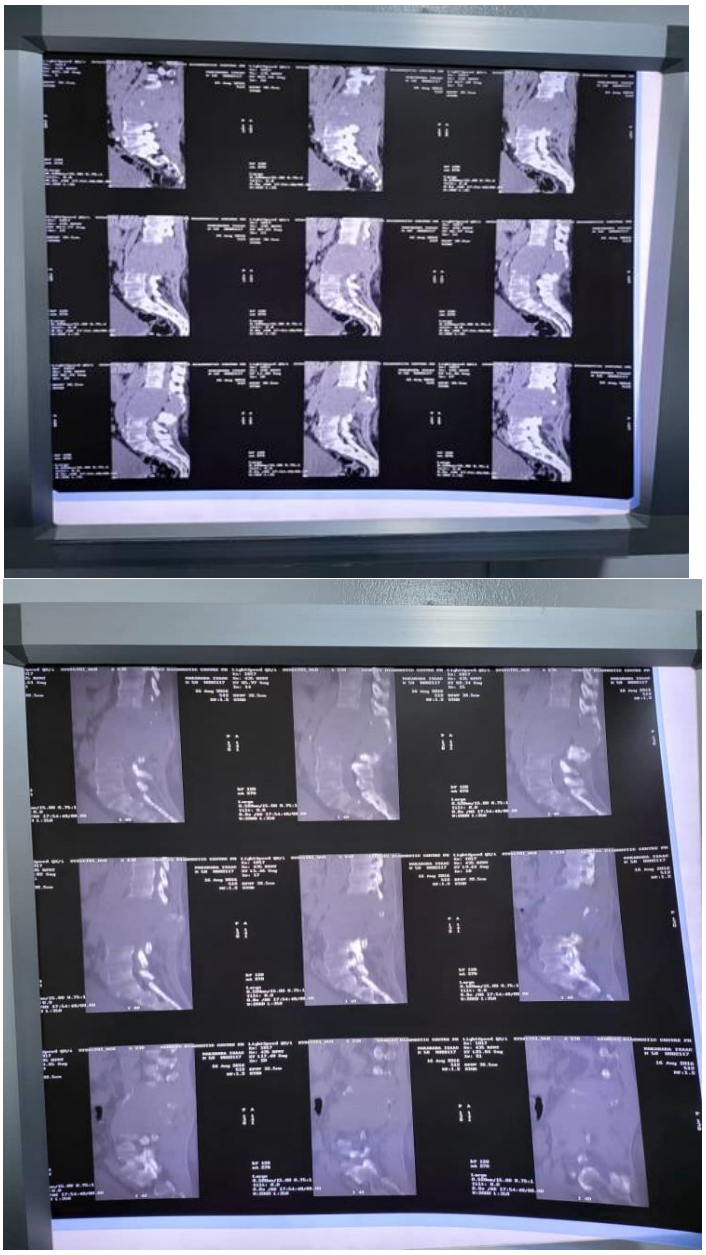

Figure 1: MRI before treatment (Sagittal Section)

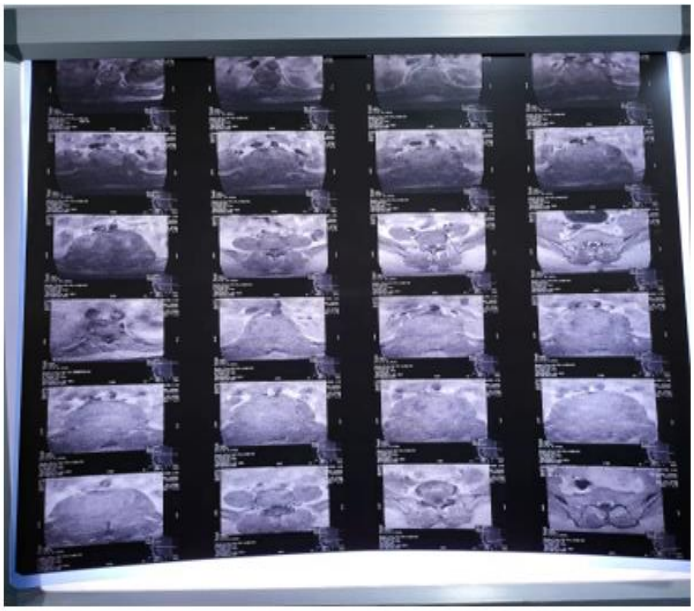

Figure 2: MRI before treatment (Transverse Sections)

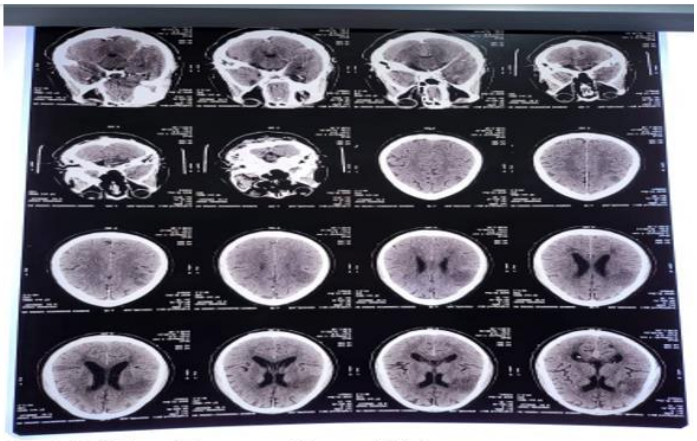

Figure 3: Head CT scan before treatment (Transverse Slides)

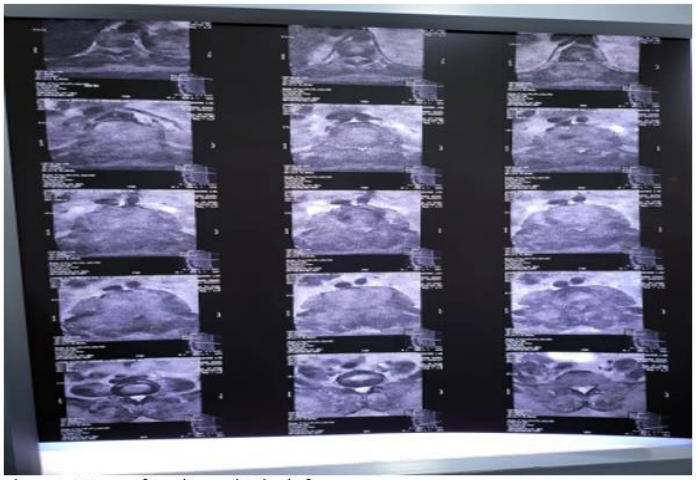

Figure 4: CT scan of Lumbosacral Spine before treatment

Figure 5: MRI of Lumbosacral spine after treatment (Transverse View)

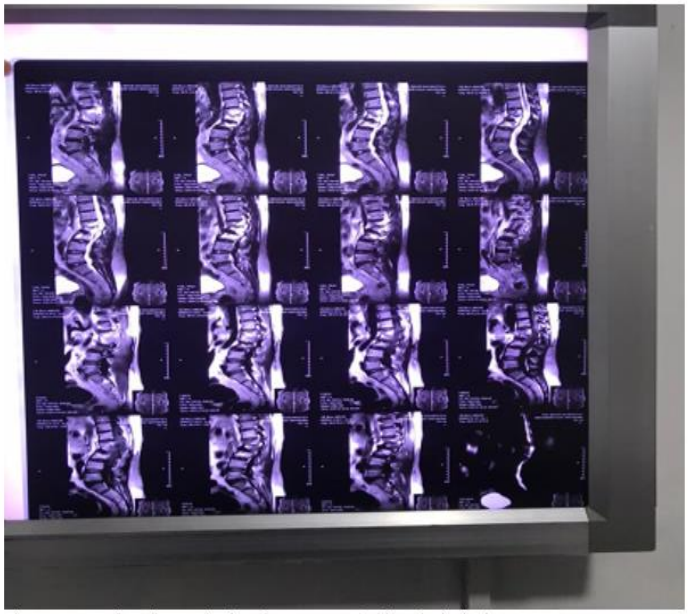

Figure 6: MRI of Lumbosacral spine after treatment (Mid-Sagittal View)

DISCUSSION

The patient discussed in this report had a 2-years history of low back pain and progressive paraplegia. Low back pain (LBP) is a frequent clinical concern in the Nigerian setting. Nottidge and his colleagues reported a point-prevalence of 42.1% in a tertiary hospital in Southern Nigeria.[28] Birabi and colleagues also reported a prevalence of 67.1% among healthy peasant farmers in South-South Nigeria.[29] LBP is commonly associated with type of occupation, previous back injury and a history of smoking in men more than women, it is rare to have low back pain as a presenting feature of serious medical pathologies like cancer,[30] thus diagnosis of solitary bone plasmacytoma (SBP) can be delayed (low index of suspicion).

The clinical presentation of solitary plasmacytoma is varied and depends on the site of collection of the malignant cells, infiltration of the spinal cord or compression of tissues near the tumour. The hallmark of solitary bone plasmacytoma (SBP) is a single site of lytic bone destruction brought on by the proliferation of clonal plasma cells, without any signs of the disease spreading throughout the body, as in the index case. The most significant risk of SBP is the development of multiple myeloma (MM). Solitary plasmacytoma is the most common distinct entity in the group of primary tumors that affect the spine, making up about 30% of all cases.[31] This hematopoietic cancer develops in bone tissue and has a high predilection for the spinal vertebrae as in the index case. It is a rare disease and so the diagnosis may be missed or delayed. The diagnostic standard for solitary plasmacytoma involves a multifaceted approach, including clinical, histological, and imaging assessments to confirm the localized nature of the disease and differentiate it from multiple myeloma. This comprehensive evaluation is essential for accurate diagnosis and appropriate management of solitary plasmacytoma.[32] The accurate diagnosis and treatment of solitary plasmacytoma are important to prevent progression to multiple myeloma, which can be life-threatening.

There is no uniform consensus on the treatment of solitary plasmacytoma, and there is also the paucity of information about the prognosis because the disease is rare.[12,33] The index case was treated with chemotherapy only due to patient's state of health and logistic concerns. Chemotherapy, radiotherapy or a combination of both, as well as surgical interventions; have been used in the treatment of solitary bone plasmacytoma. In a study by Fregonese et al. that reviewed the efficacy of radiotherapy alone for controlling SBP, their data showed that definitive radiotherapy for SBP can provide excellent local control. Consistent with previously published literature, progression to MM was the primary concern and was more prevalent in patients with SBP than extramedullary plasmacytoma.[34] Sharpley et al. reported a cohort of 66 patients with the majority (96.2%) of them receiving radiotherapy, their study showed that 90.7% of the patients had 5 years overall survival. However, some progressed to multiple myeloma despite treatment, and the average time to progression was 61 months.[35]

A recent report showed that modern radiotherapy techniques combined with an RT dose >40 Gy may enhance local control and lower the relapse rate without appreciably lowering survival rates. In this study, patients with SBP had higher myeloma-free survival and progression free survival rates with the addition of novel systemic therapies.[36] Interestingly, some reports using a small number of patients have demonstrate the efficacy of using novel chemotherapeutic agents containing Proteosomes Inhibitors – bortezomib-based regimen. The findings from a study by Mheidly et al. showed that adjuvant chemotherapy for the treatment of SBP had a positive effect on progression-free survival (PFS) but not overall survival (OS) and was unable to stop SBP from progressing to MM, although the prognosis with the adjuvant chemotherapy with novel medications was better than with older forms of chemotherapeutic regimen.[37] The index case received only novel chemotherapy, with a Bortezomib-based regimen and had maintenance with lenalidomide after 8 cycles of chemotherapy. He had full neurological recovery and recovery of the function of his two lower limbs. He has not progressed to multiple myeloma at the time of this report, after 7 years of follow-up.

CONCLUSION

Solitary bone plasmacytoma is a rare disease that has a predilection for the spine, a high index of suspicion is required, especially if it presents with non-specific symptoms such as low back pain. Its diagnosis requires a rigorous, stepwise approach, although the plasma cells respond well to radiotherapy, where this is not easily available, patients should be offered treatment with novel chemotherapy with bortezomib-based regimen and the necessary supportive care by a multidisciplinary team. More multi-centre studies will be needed from this region to determine outcome with available treatment modalities.

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