An Unusual Exhibit of Cervical Plasmacytoma with Trademarks of Cervical Myelopathy

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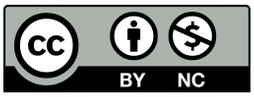

ABSTRACT

We reported a case of a 76-year-old man presented with cervical neck pain since 16 days. He had also weakness of all limbs since 4 days, insidious in onset. MRI Cervical spine revealed an expansile lesion in C2-C3 vertebral level and C2 lamina pathological fracture. He was suspected as a case of multiple myeloma. However, the histopathology report revealed morphological features compatible with Plasmacytoma.

Key Words : Cervical spine; Cervical Myelopathy; **Plasmacytoma.**

INTRODUCTION

Acclaimed to have its genesis from plasma cells, Plasmacytoma are infamous for forming discrete lesion either in bone or in soft tissue.[1,2] In the recent past, the International Myeloma Working Group in 2003 acknowledged anautonomous classification into three subtypes. When a single bone lesion is present mainly in the axial skeleton, they are termed Solitary Plasmacytoma of Bone (SPB). On the other hand, manifestation of plasmacytoma at the site of soft tissue / extra-osseous lesion are named Solitary extra-axial Plasmacytoma (SEP). To the contrary, multiple lesions within the bone and in soft tissues but without the evidence of bone-marrow engagement are labelled multiple solitary plasmacytoma (MSP).[3] Quoted above, the plasmacytoma, designated as SPB, SEP and MSP based on their liaison with the strains of tissues associated are exquisite clinical entities, portrayed by a monoclonal plasma cell infiltrate in bone or soft tissue and cytological depiction to multiple myeloma (MM). Nonetheless, they are disparate from Multiple Myelomaby the exclusion of hypercalcaemia, renal failure, anemia, pathological monoclonal plasmocytosis on a random (usually of the iliac crest) bone biopsy, bone lytic changes (except for the primary solitary lesion) and serum or urinary monoclonal protein (Bence Jones Protein) which are postulated to be obligatory in above-noted.

Epidemiological survey has disclosed that the incidence of plasmacytoma is 0.35/100 000/year, accounting to 5–10% of all plasma cell neoplasms.[4,5] The eminence of confirmation lies with the prospects of these frailty to evolveinto Multiple Myeloma. Astoundingly, as high as 52–80% of patients with SBP and 10–44% of patients with SEP are more seemingly presumed to progress into Multiple Myeloma later in life.

CASE REPORT

A 76-year- old gentleman, presented to our institution with the complaints of pain over the nape of the neck for 16 days. It was gradual in onset and progressive in nature followed by weakness of all four limbs for 5 days. However, his bowel and bladder habit is normal. Past history of fall injury 16 years back while carrying heavy weight. A thorough neuropsychological examination was carried out. All Cranial nerves were intact. Tone of the major muscle groups in all limbs were decreased. Evaluation of motor power revealed a decrease to 4/5 in the right upper limb and 3/5 on the left upper limb. Distal (handgrip) was decreased to 3/5 on the right and 1/5 on the left. Hoffman's Sign was positive bilaterally. Examination of lower limbs disclosed a decrease in motor to 3/5 on the right and 4/5 on the left. Planter response were extensor bilaterally. Evaluation of the sensory segment revealed a decreased sensation below D9.

Spinal pathology was contemplated preliminarily and radiological investigation was sought out. MRI of the cervical spine revealed an expansile lesion in C2-C3 vertebral level with pathological fracture of C2 lamina(Fig1A&B) This was an unanticipated discovery. A second thought of metastatic lesion was appraised and contrast enhanced CT Scan of chest and abdomen was performed. The finding was suggestive of an expansile lesion in body of the right 8th rib with slight enhancement of iso dense matrix of lesion, iso dense nonexpansile intramedullary isodense lesion in proximal shaft of the right humerus at metaphysis and collapsed D9 vertebral body with a calculus in lower pole calyx of left kidney.

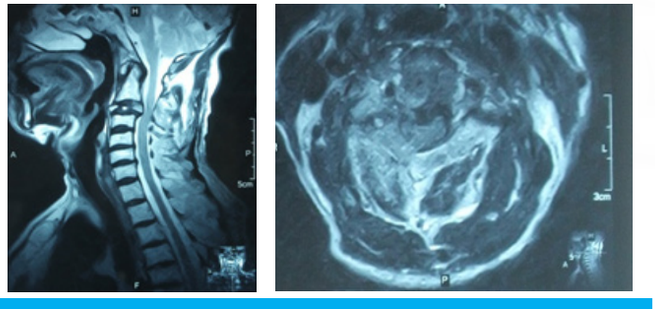

Figure 1 A&B: MRI of the cervical spine revealed an expansile lesion in C2-C3 vertebral level with pathological fracture of C2 lamina.

These findings shed light upon the probable plasma cell pathology. Henceforth, biochemical investigation in reference to multiple myeloma was carried out which unveiled negative for Bence jones protein. Comprehensive myeloma protein panel disclosed a yotal protein of 9gm/dl, Albumin of 3.12, Alpha 1 globulin of 0.28, Alpha 2 globulin of 0.74, Beta 1 globulin of 0.31, Beta 2 globulin of 0.19, gamma globulin of 4.26 along with detection of Myeloma Band- M Band and M -spike-3.87. Additionally, Mantoux test-1 mm, CRP- 50.86 and Vitamin D- 45.81 was found to rule out other pathology. With the hope of pursuing a decompression of cervical spine, multi-level cervical laminectomy was performed. Under general anesthesia, using a Mayfield neurosurgical skull clamp in prone position, a posterior midline incision extending from inion to 4th cervical vertebrae was made. The wound is then extended along the paravertebral muscles until the periosteal layer and exhibiting the pathological fracture of C2 lamina. The cervical bones were soft, osteoporotic and edematous. Serial laminectomy was performed from C2-C4. Dense adhesion was noted over the epidural space compressing the cord. A meticulous tissue dissection was performed andadequate decompression is achieved. Further, a lesion measuring approximately 5 cm x4 cm in the right cervical region encroaching up to the left hemi cord was excised. Epidural drain was kept. Wound was closed in layers and skin stapled. A tight compression bandage was applied. As anticipated, the histopathology report revealed morphological features compatible with plasmacytoma.

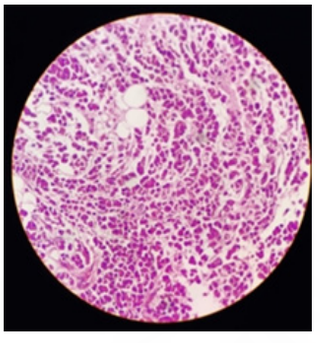

Figure 2: Histopathological examination showing mature and immature plasma cells. The cells have abundant cytoplasm and eccentrically placed nuclei. Some of the cells show centrally placed nuclei with prominent nucleoli.

DISCUSSION

Multiple solitary plasmacytoma, a subtype of the genre plasmacytoma is characterized by monoclonal plasma cell infiltrate in one or more lytic bone lesions. Moreover, these are found to spreadto soft tissueswithin its vicinity. Remarkably, there is no evidence of plasma cell proliferation on random bone marrow biopsy and without the systemic abnormalities of multiple myeloma (hypercalcaemia, anemia, renal failure, serum and/or urinary monoclonal proteins or monoclonal light chains).[6] Nevertheless, presence of M- band/ chain is a salient feature.

In our case, M Band was detected. However, it was negative for urinary Bence Jones protein. Myeloma protein panel showed slight elevation of total protein and Gamma globulin with a decreased level of albumin, beta 1 and 2 globulin. Rest of the biochemical parameters were unremarkable. Radiological survey revealed multiple

bony lesion in the body of the right 8th rib, proximal shaft of the right humerus, with features of pathological fracture in D9 vertebral body and C2 lamina with osteolytic lesion of C3 vertebra causing compression over cervical cord with a consequent myelopathic and hence the hallmarks of cervical myelopathy evident in clinical examination. Histopathologicalexamination was positive with morphological picture of plasmacytoma. Alongside the presence of myeloma band and M- spike with multiple bony lesion which anachronise plasma cell disorder, the histological picture of our patient corroborated to the diagnosis of multiple solitary plasmacytoma. With reference to our case, the constitutional symptoms of fatigue and generalized weakness eventually evolved tomore specific symptoms probably owing to the osteolytic lesion causing pathological fracture of C2 lamina with resultant compression over cervical spinal cord. As cervical myelopathy came to fore,a full blown progressive weakness evoked in all four limbs. He did stated a history of fall while carrying heavy weight around 17 years back not associated with any symptoms back then presumably resorting to a trivial nature of the trauma.

To outreach the diagnosis, besides the clinical and biochemical survey, the use of imaging modalities viz. CT, MRI and PET in the evaluation and staging of apparently solitary plasmacytoma will reveal multiple soft tissue lesions or additional bone lesions and will enhance in the future the correct diagnosis of MSP.[1,7]No clear guidelines for the treatment of Multiple Solitary Plasmacytoma has been formulated till date, seemingly due to the rarity of the ailment and the heterogeneous presentation of this frailty. Most of the information available regarding the best treatments are found in single or multicentre retrospective series. Treatment modalities in the form of chemotherapy, radiotherapy and surgery have been tried with an inconsolably variable results.[1,2,8,9] While for single-lesion cases of plasmacytoma (SBP and SEP) an upfront surgical excision followed by radiation therapy appears a judiciousoutlook, for multiple-lesions disease (MSP) a blend of surgery, radiotherapy and chemotherapy is morecoherent.[1,9]

CONCLUSION

Being a rare ailment with a heterogeneous mode of presentation, a conscientious workup is obligatory to attain a final diagnosis of plasmacytoma. Despite the paucity of befitting guidelines for its treatment, individualization can be done based on its subtype such that an affirmative aftermath can be accomplished.

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