

Multifaceted Approach to the Management of C2 Vertebra Destructive Lesion in a Patient with Kappa IgG Multiple Myeloma with a Good Outcome: A Case Report and a Literature Review

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Abstract

Introduction: To present a rare case for a male in his fifth decade of life who was complaining of cervical swelling and neurological deficits attributed to a destructive lesion in the C2 vertebra, ultimately diagnosed as Kappa IgG Multiple Myeloma (MM).

Symptoms and findings: The patient presented with painless posterior cervical swelling associated with progressive weakness and loss of sensation on the left upper and lower limbs with intact reflexes and normal muscles tone. Upon conducting the appropriate images, he had a C2 destructive lesion mandating urgent surgical fixation.

Diagnosis and intervention: An extensive laboratory workup and imaging were conducted. Furthermore, surgical intervention was carried out

including occiput cervical fusion with C2 decompressive laminectomy and biopsy of the lesion. Postoperatively, the patient had a dramatic neurological improvement, and the biopsy revealed no malignancies, however, further workup revealed Kappa IgG MM.

Conclusion: MM affecting the cervical spine is uncommon, often presenting with neck pain. However, our case demonstrated a unique presentation of painless cervical swelling and deformity leading to acute neurological deficits. A systematic diagnostic approach, including imaging and surgical intervention, was crucial for the successful management of this case highlighting the importance of considering MM as a differential diagnosis in patients presenting with cervical

instability and neurological deficits, even in the absence of known malignancies.

Keywords: Plasma Cell Myeloma; C2 fracture; Cervical vertebrae fracture; Kahler disease; Multiple myeloma; Cervical Posterior decompression; Cervical instrumentation

Introduction

Multiple Myeloma (MM), known as Kahler disease, is an incurable hematological malignancy characterized by abnormal plasma cells proliferation and production of a numerous number of monoclonal immunoglobulins. It is the second most common hematological malignancy, and it is estimated to affect 0.9% of the population with an average age at presentation of 69 years and slight male predominance [1,2]. MM is usually preceded by premalignant, asymptomatic, quiescent condition named Monoclonal Gammopathy of Undetermined Significance (MGUS), which is identified by a monoclonal protein level less than 3g/l, clonal bone marrow plasma cells less than 10%, and the lack of end organ damage defined as absence of hypercalcemia, renal insufficiency, anemia, and bone lytic lesions (CRAB). MGUS could progress into Smoldering Myeloma (SMM), which is described as a monoclonal protein level greater than 3 g/l, clonal bone marrow plasma cells greater than 10%, and the absence of CRAB. The hallmark feature of MM is the presence of end organ damage [3]. MM is characteristic for inhibiting osteoblastic activity and enhancing osteoclastic activity. In contrast, other types of malignancies affecting the skeleton, such as breast and prostate cancers typically, activate both osteoblasts and osteoclasts. Therefore, MM exhibits minimal to almost zero bone production. This bone

destructive phenomenon was also observed in MGUS and during MM remission status [4].

In this case report, we present a 47-year-old previously healthy male who was diagnosed at an early age with MM after initially presenting with painless cervical swelling and acute neurological deficits that revealed destructive cervical osteolytic lesions and C2 fracture. Given that MM is still an uncommon malignancy globally, this case is distinctive for its rare initial presentation. A comprehensive search reveals only four cases of MM onset with cervical spine fracture [5-8], our case stands as the only one presenting free of pain. This report provides a detailed description of the presentation, management, and outcome of our case. Furthermore, a literature reviews of previous similar cases and an in-depth comparison.

Case Presentation

History and physical examination

In the bloom of his youth, a 47-year-old, previously healthy male presented to our hospital with one month history of progressive posterior cervical swelling associated with fixed neck flexing position for one week. In addition to the swelling, he reported left side weakness and diminished sensation in both upper and lower limbs. However, he had no history of trauma, urine or stool incontinence, weight loss, fever, night sweats, changes in bowel habit, or abdominal pain. Furthermore, he had an unremarkable family or personal history for malignancies or Tuberculosis (TB). Upon physical examination the patient was conscious, alert, and oriented to time, place, and person. His neck was maintained in a flexed position. The cranial nerve examination was unremarkable. Motor examination of upper limbs strength was graded as the following

left arm and forearm 4/5, left hand 3/5, right upper arm strength was full, and distally his right hand power was 4/5. Regarding lower limbs strength right lower limb strength was full, the left lower limb was weak, with prominent weakness distally graded as the following hip and knee 4/5, ankle (plantarflexion and dorsiflexion) 2/5, extensor hallucis longus 1/5. Additionally, Hoffman's sign was positive on the right side. Reflexes and muscle tone were normal bilaterally. Finally, a decreased sensation was observed in both left upper and lower limbs.

Initial laboratory workup and imaging

After detailed history and physical examination, urgent brain and cervical spine CT was done, neck

collar was applied to prevent further neurological damage, septic workup including Acid-Fast Bacillus (AFB) and QuantiFERON tests for TB were ordered, both came back negative. The CT head and cervical spine was done urgently in the emergency department, C2 vertebrae was almost undetectable with dramatic destruction and erosions of the C2 vertebral body and odontoid process, along with soft tissue fullness, haziness, and stranding in the surrounding peri-vertebral space and intra-spinal region. No definite collection pockets were seen, as shown in **Figure 1**.

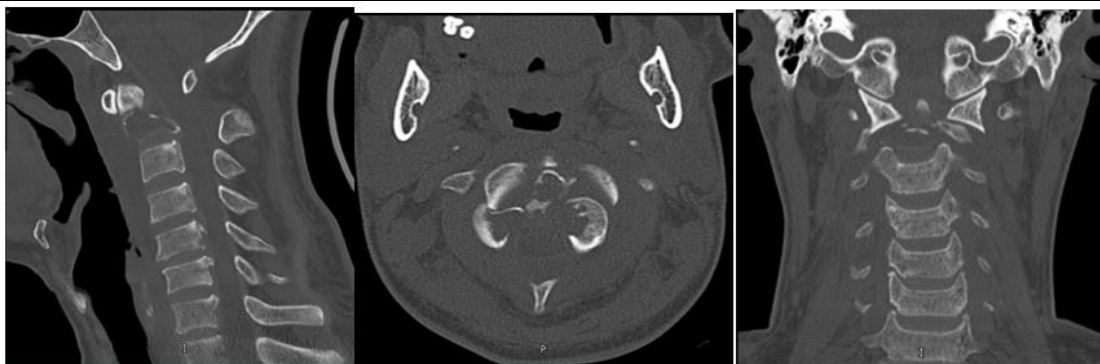


Figure 1: Brain and cervical spine CT scan conducted in the emergency department obviously showing destruction of the C2 vertebrae.

Further imaging was requested including cervical spine MRI which showed significant destruction and reduction of C2 vertebral body and posterior neural height. Additionally, heterogenous predominantly bright T2/STIR signal intensity, with heterogenous postcontrast enhancement, was noticed. Moreover,

there was an increase in the distance between the C1-C2 spinous processes, with partial posterolateral spondylolisthesis of C2 over C3, and linear high signal intensity in the cervical cord at the level of C4-C5 with volume loss suggesting myelomalacia as seen in **Figure 2**.

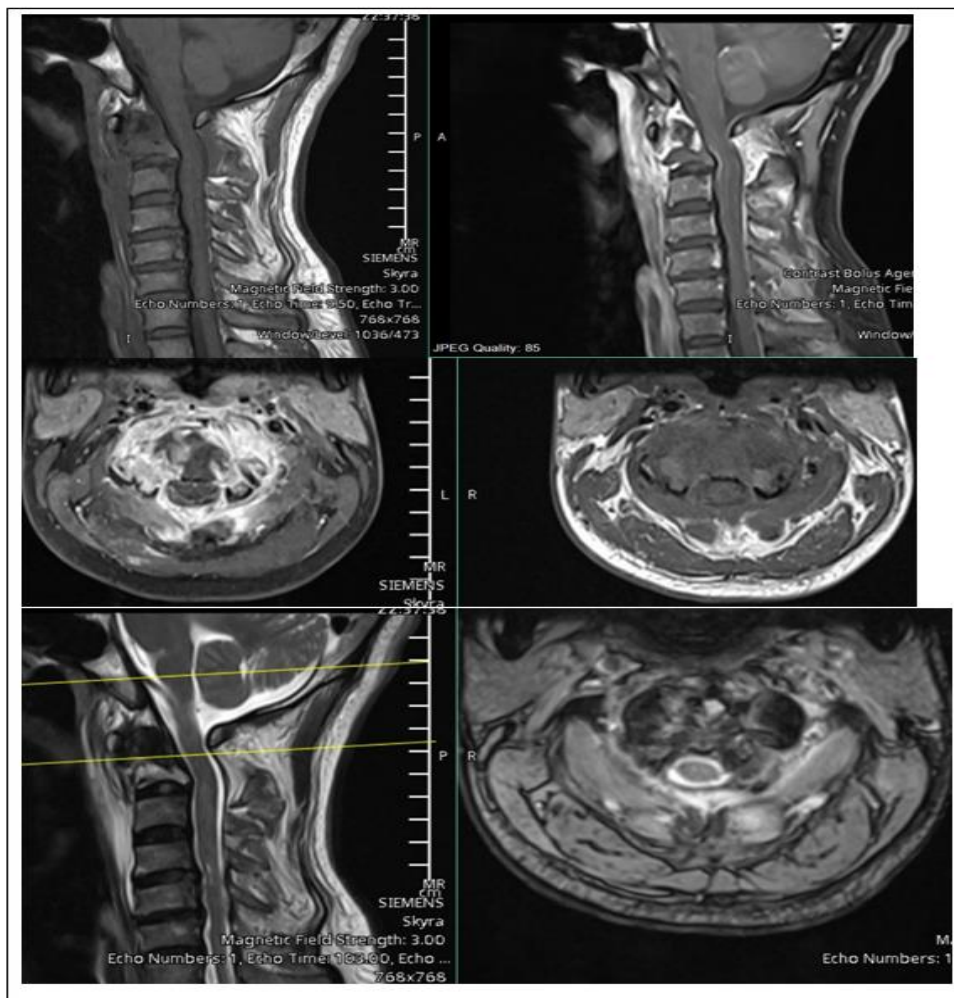


Figure 2: Cervical spine MRI showing prominent C2 vertebrae destruction and reduction in the height of its body. Extensive soft tissue swelling with heterogeneous post-contrast enhancement is seen in the prevertebral soft tissue, extending to the retro/parapharyngeal space with no definite collection. Similar but to a lesser extent soft tissue swelling and enhancement were noted in the posterior paraspinal muscles. Increase in the distance between the C1-C2 spinous processes, with partial posterolateral spondylolisthesis of C2 over C3 was also seen. Linear high signal intensity was seen in the cervical cord at the level of C4-C5 with volume loss suggesting myelomalacia.

Management

Halo-vest was applied as preoperative precaution to maintain the position then the patient underwent occiput-cervical fusion from the sub occiput to C4

under fluoroscopy with C2 decompressive laminectomy and biopsy of C2 destructive lesion. Postoperative X-Rays are shown in **Figure 3**.

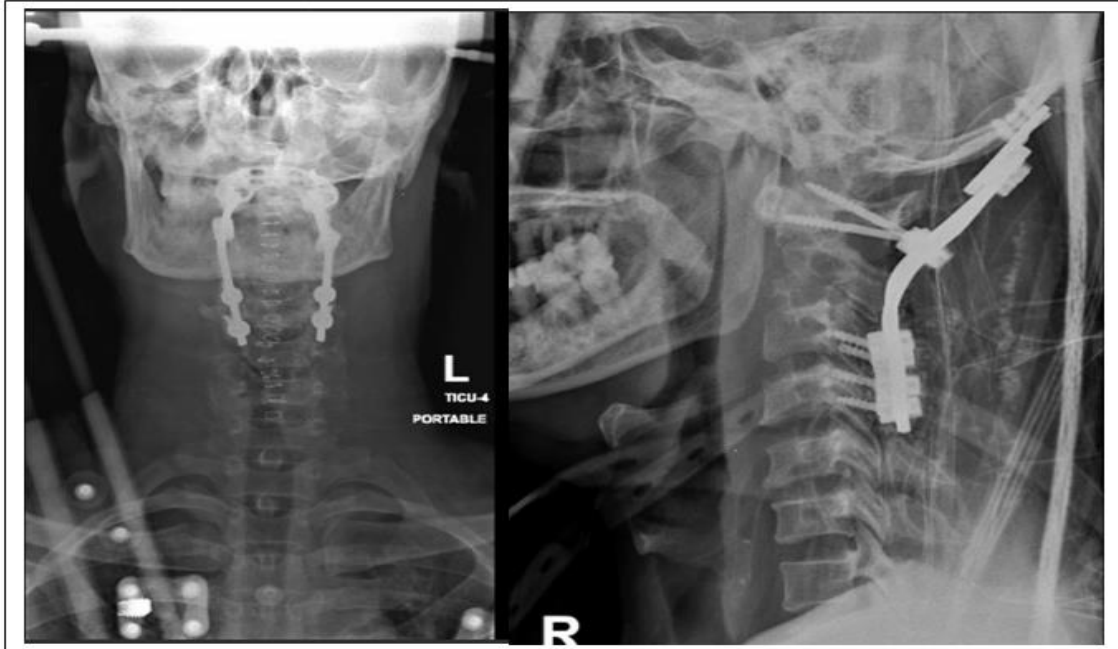


Figure 3: Post operative X-rays showing cervical spine fixation.

Multiple myeloma workup

After surgery, histopathology results came as fibroadipose tissue with mild chronic inflammation and no evidence of malignancy. However, pan body CT scan with contrast was done afterward to check for other associated lesions elsewhere, which showed only one osteolytic lesion at the posterior part of the S1 vertebra suggestive of MM or metastasis. Therefore, an extensive MM workup was initiated involving a protein electrophoresis test which showed a polyclonal increase in gamma globulins, but no monoclonal bands were detected even after immunofixation. Also, a urine protein electrophoresis test was conducted which showed the presence of free Kappa light chains and was positive for Bence Jones proteins. Then, a Positron Emission Tomography (PET) scan was done and showed diffuse increased Fluorodeoxyglucose (FDG) uptake in the bone marrow with multiple lytic lesions showing increased focal uptake denoting active

disease sites, a picture suggestive of MM. Eventually, Iliac crest biopsy was ordered, the flow cytometry on sacral bone biopsy showed approximately 54% kappa monotypic plasma cells consistent with plasma cell neoplasm and the final pathology report confirmed Kappa IgG MM.

Outcome and follow up

Postoperatively, the patient showed significant improvement in limbs strength. On the third day, he was neurologically intact. Regarding his MM, he was prescribed Velcade, Revlimid, and Dexamethasone (VRD) in addition to Daratumumab by A hematologist. Currently, the patient is treated as outpatient with regular follow ups.

Discussion

Skeletal Related Events (SREs) are common complications of MM. SREs are defined as spinal cord compression, pathological fractures, and palliative radiotherapy or surgical management for

bone related pathologies. At presentation 70% to 80% of MM patients suffer from osteolytic bone lesions and over 90% of patients develop bone lesions through the disease course, most commonly the lesions are painful and involve the axial spine, particularly chest and back with vertebral bodies being most involved 49% followed by skull 35%, pelvis 34% and ribs 33% [9-12]. In contrast to the painful nature of MM bone involvement our patient exhibited a distinctive painless initial presentation. MM is the most common cause of bone metastatic lesion that could present with pain, fractures, and spinal cord compression causing a substantial effect on the patients' quality of life. Metastatic lesions mostly affect the spine with the thoracic spine being most likely involved in 70% of the cases, followed by lumbosacral and cervical spine 20%, 10%, respectively [13]. Herin, we describe a previously healthy male in his fifth decade of life presented with painless posterior cervical swelling revealing an extensive C2 vertebrae fracture which was ultimately

diagnosed with Kappa IgG MM. Fractures in the cervical region are associated with high mortality rates due to the presence of many vital structures in this area, particularly C2 fractures, which have a 40% mortality rate within two years. Early detection and proper management of such fractures is critical, as delays can mean the difference between a severe, irreversible outcome and a minor, treatable complication. However, detecting C2 fractures can be challenging and requires a vigilant, experienced physician [14,15]. In this case report cervical CT scan was initially conducted revealing the deleterious extensive C2 fracture, thereafter MRI was ordered to further visualize the lesions.

Among the four reports with cervical spine fracture as an initial presentation of MM as shown in **Table 1** our case marks as the only one presenting with painless swelling. Moreover, only one case was reported in young age for a female while all other patients were between the ages of 60-67 years [5-8].

Table 1: The table describes a comprehensive mapping of the literature for cases of MM who their first presentation was cervical vertebral fracture.

<i>Case number</i>	<i>Year of publication</i>	<i>Authors</i>	<i>Age</i>	<i>Gender</i>	<i>Comorbidities</i>	<i>presentation</i>	<i>Fracture site</i>	<i>Management</i>	<i>Outcome</i>
1	1994	Lisa Chan, Howard S Snyder, and Vincent P Verdi	61 years	Male	Angina and hypertension	The patient had acute onset neck pain after a sneeze, it was progressive. On the next few days, he	C2 fracture involving the body and lamina	Posterior cervical fusion of C1 and C2 in addition to a left iliac crest graft to support fracture instability.	The patient showed improvement after surgery and was discharged home in a good health despite post-

		le.				developed neurological deficits and eventually quadriplegia.			operation complication (pneumonia and cholecystitis)
2	2006	Jiri Pavlu, Riaz Jan-Mohamed, and Richard Kaczmariski.	67 years	Male	–	A 67 male patient complained of neck pain of 14 months duration associated with recurrent vertigo. Symptoms started after a minor trauma and tended to be progressively worse, particularly with neck motion. Additionally, the cervical spine was tender upon physical examination without	C2-odontoid peg fracture causing compression over the spinal cord	Occiput-cervical fusion was performed to control the fracture followed by chemotherapy for the MM	After 7 months of chemotherapy for his MM there was a noticeable improvement, and the patient remained asymptomatic.

						noticeable neurological deficits.			
3	2007	Miriam Rodriguez-Catari et al.	47 years	Female	-	After 6 weeks of suffering from neck pain her pain reached maximum severity the night prior to her presentation, minimal movement led to a cracking noise. Next day, an X-ray showed C2 fracture which was unstable. However, there were no neurological deficits, and a neck collar was placed to prevent spinal cord compression or further	C2 unstable fracture	After applying MM treatment guidelines, the patient was on a neck collar for 9 months she was experiencing discomfort, pain, and the fracture was unstable which necessitate a surgery to relieve her symptoms. A percutaneous vertebroplasty (PVP) was performed to stabilize the fracture and relieve the pain.	The patient was able to perform her ordinary daily activity after PVP on 18 months follow up and she was free of pain during the follow up period.

						damage.			
4	2014	Andrew James Berg, Miguel Hernandez, and Chandra Bhatia.	60 years	Female	-	The patient had neck pain for several weeks it was getting more painful and associated with left upper arm pain but no neurological deficits. were observed	Complete destruction of C4 vertebrae leading to unstable cervical spine. Furthermore, she had a humerus fracture.	The patient underwent C4 corpectomy with cervical spine stabilization from C3-C5 vertebrae. Moreover, upon discharge she was started on the MM treatment regimen.	She had a remarkable improvement on the follow up visit 8 weeks post operation, she was free of pain and her spine was stabilized with no evidence on cord compression.

Regarding the management of the previous similar cases, all of them had surgical management in addition to chemotherapy and radiotherapy. The technique used intraoperatively was cervical and occiput cervical fusion which coincides with our case, while one case was managed with percutaneous vertebroplasty (PVP). However, the definitive management option whether conservative or surgical depends on the site of the fracture, presence of other complications, and fracture stability. The main goal is to stabilize the fracture and prevent further damage, some cases can be managed with palliative radiotherapy which produces excellent results and symptoms relieve. In the four previous cases the management yielded notable improvement, in our case the patient was neurologically free of any deficits in the third day after surgery, which was a

rapid and excellent response in comparison to the previous similar cases [5-8,16,17]. Our patient was not diagnosed with or investigated for MM or other malignancies before his latest presentation to the emergency department with painless cervical swelling and neurological deficits thus, the first presentation of MM in this patient was C2 vertebrae fracture which was managed urgently by surgery and the appropriate MM chemotherapy. Furthermore, the painless nature of his MM lesions and the early age at which he was diagnosed add to this case uniqueness. However, case reports provide weak evidence, and we cannot generalize the findings of our case nor confirm the causality between painless cervical swellings and MM, this case encourages considering atypical disorders when encountered by an atypical presentation and furnishes the literature by a stepwise

organized approach on how to deal with similar cases.

Conclusion

In this case report we shed light on the importance of systematic based approaches in evaluating the patient, especially those presenting with unusual complaints, considering multiple myeloma as a differential diagnosis in patients presenting with cervical instability and neurological complaints, even in the absence of known hematological malignancies. A multidisciplinary approach, involving orthopedic and hematological expertise, is essential for effective management and favorable outcomes.

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