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Case Report

Cervical Fracture C7-T1 in a Young Patient with Multiple Myeloma: Report of a Case

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Abstract

Multiple myeloma is a neoplasm with malignant characteristics, which affects plasma cells, infiltrating the bone marrow in the usual way, causing osteolytic lesions in the affected bone, and other multi-organ complications. Epidemiologically it is observed in ages between 50 and 70 years. We present a case of a 33-year-old male patient with C7-T1 vertebral fractures managed surgically; In his medical studies, plasmacytosis> 60% was evidenced in the medullogram, protein electrophoresis highlighted an increase in the Alpha 2 component with splitting in this same fraction, in addition the results of Protein M and Bence Jones were Negative.

***Keywords:** multiple myeloma, solitary plasmacytoma, monoclonal protein, Bence-Jons protein, pathological vertebral fractures.*

Introduction

According to the WHO, myeloma is a type of malignant neoplasm, included within the group of monoclonal gammopathies, distinguished by the abnormal proliferation of malignant plasma cells (PC) towards the bone marrow, born from a single clone, with varying degrees of immaturity, including atypical forms; where B lymphoid cells are most involved. These cells can form a tumor (plasmacytoma).[1,2,3] Thus, when there is more than one plasma cell tumor, according to Rustizky in 1873, the pathology is called Multiple Myeloma (MM). [4.5] There is no agreement between the authors regarding sexual predominance, although there is greater indication for the male sex; its incidence increases with age, reaching a peak between 50 and 70 years, its appearance being infrequent before the age of 35.[1,2,4,5,6,7] And their life prognosis is 2 to 3 years, and only 3% of patients survive more than 10 years.[5]

The objective of reporting this case is to present a patient with a diagnosis of multiple myeloma with characteristics little described in the literature, which include, among others, young age at diagnosis, the expression of the monoclonal band at the level of Alpha-2 globulin in the protein electrophoresis and the lack of previous symptoms suggestive of monoclonal pathology.

Clinical Case

A 33-year-old male patient, with a personal history of arterial hypertension, an active smoker since the age of 12, with a grade of elementary schooling, a bag-carrying trade, and no relevant family history. Who presented a clinical picture secondary to direct trauma to the cervical region after a fall from height, consisting of sharp stabbing pain, of moderate intensity, at the cervicodorsal level, associated with anesthesia and complete acute loss of muscle strength in the lower limbs, which prevented the standing and walking, as well as anal and bladder incontinence, without compromise in the upper limbs. Recovery of sphincter function was spontaneous at 2 weeks. Arrives at our hospital center 6 weeks after the trauma, At that time, he reported mild pain in the lower cervical and upper dorsal region, dysesthesia in the trunk and both lower limbs, synkinesias in both lower limbs, priapism and inability to walk due to paraparesis.

The positive findings on neurological examination were pain on palpation at the lower cervical and upper dorsal level over the supraspinatus region, in addition to Hypoesthesia from the D10 dermatome, Loss of epicritic touch from D10 down, apalestesia, abatesthesia in both lower limbs, reflex right extensor cutaneoplantar (Babinski positive) and Babinski substitutes also positive, Right exhausting foot clonus, decreased muscle strength in lower extremities 2/5, hyperactive patellar reflexes (++++ / +++), toned anal sphincter suitable. Dysesthesias in the trunk and both lower limbs, synkinesias in both lower limbs, priapism and inability to walk due to paraparesis. The positive findings on neurological examination were pain on palpation at the lower cervical and upper dorsal level over the supraspinatus region, in addition to Hypoesthesia from the D10 dermatome, Loss of epicritic touch from D10 down, apalestesia, abatesthesia in both lower limbs, reflex right extensor cutaneoplantar (Babinski positive) and Babinski substitutes also positive, Right exhausting foot clonus, decreased muscle strength in lower extremities 2/5, hyperactive patellar reflexes (++++ / +++), toned anal sphincter suitable.

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Among the examinations carried out, the following results of interest were obtained:

- Hemogram: no alterations.
- Serum Protein Electrophoresis (SPEP): Slightly high concentrations of Globulin Alpha-1 and even more of Alpha-2 with a splitting of this fraction, with levels in the upper limit of Globulin Gamma, normality of Globulin Beta, and low Albumin levels (Fig. 1)
- Serum light chain concentration = Kappa: Non-reactive and Lambda: 1.65 g / L.
- Total proteins: 30.5 g / dl.
- Monoclonal protein (Protein M): Negative.
- Bence – Jones protein in urine: Negative.
- LDH 420 U / L.
- FA 145 U / L.
- Serum calcium (3.0mg / dl).
- Serum creatinine (76mml / L).
- Echocardiogram: No alterations.
- Medullogram: Reports eosinophilic infiltration with abundant M (plasma) cells> 60%.
- Bone marrow biopsy: Elevated presence of plasma cells (medullary plasmacytosis)> 60%.
- Skull X-ray: Evidence multiple, scattered osteolytic images in a bite-out pattern in the cranial vault.
- Simple Skull Tomography: In the bone window, diffuse, variable size osteolytic bite-out lesions are observed in the cranial vault, middle and posterior fossa of the skull base. No intracranial or extraxial intracranial lesions are observed (Fig 2 a, b and c).
- Simple cervical spine tomography: C7 and T1 wedge fracture of the vertebra, with presence in the spinal canal of superior plate fragment and irregular isodense image in the spinous process of both vertebrae that invades the spinal canal and infiltrates the vertebral body, plus osteolytic images In bite-out in lower segments, with a predominance of vertebral bodies T6 and T7, there is no apparent loss of vertebral alignment (Fig. 3 a, b, c).
- Chest CT. Osteolytic images in the sternal region by three thirds, in the clavicles and ribs, no lesion in the mediastinum or lung (Fig. 4).
- Lumbo sacral spine CT: Small osteolytic images at L4 and L5.

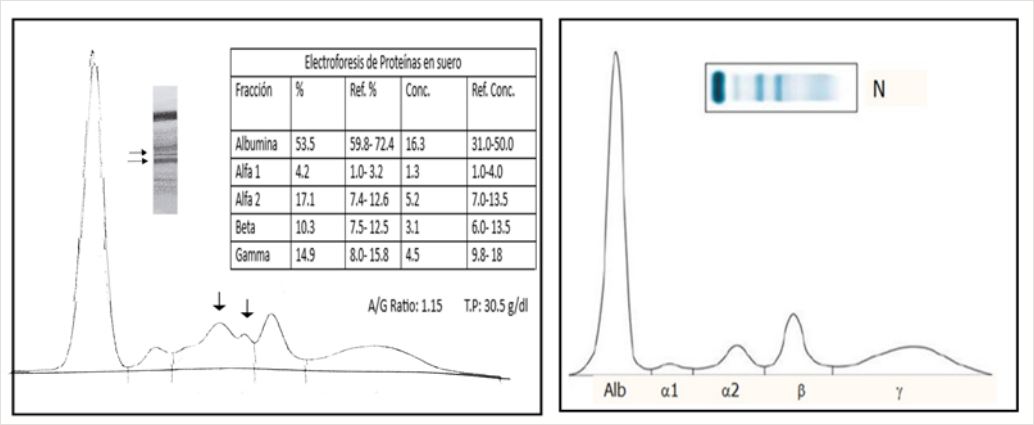

Fig 1: *Serum protein electrophoresis. Note the splitting of the Alpha-2 protein fraction on the right side vs a Protein Electrophoresis with normal results on the left side.*

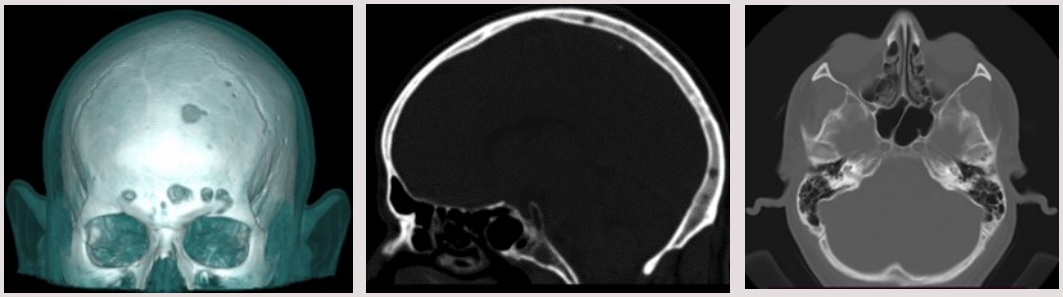

Fig. 2*. Simple Skull Tomography. A. Coronal view in reconstruction b and c Sagittal and Axial view, respectively, in bone window, where the osteolytic images are distinguished in a bite-out.*

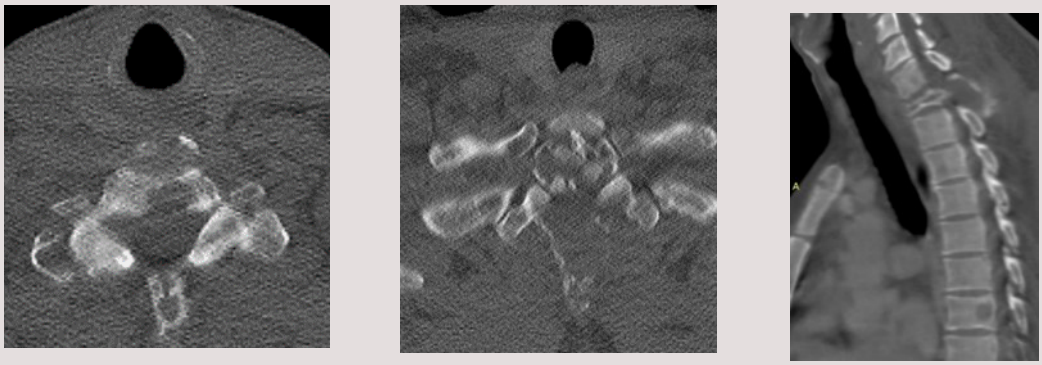

Fig 3.*to. CT cervical spine. Axial cut of the C7 cervical vertebra, showing complete loss of the anatomical image of the vertebra, due to the severity of its fracture, and large osteolytic images present. b. CT cervical spine. Axial cut of the C6 cervical vertebra, there is a marked osteolytic area that mainly involves the spinous process and the vertebral body. c. CT cervical spine Sagittal cut The complete destruction of the C7 vertebra was revealed, with infiltrative-type osteoclastic images in a punch in the C6, T6 and T7 vertebrae, and in the sternum.*

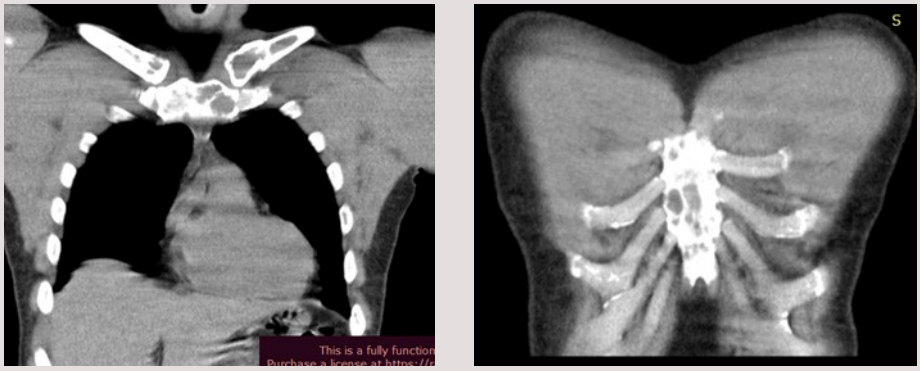

Fig 4*. Tomography of the thorax coronal sections. They show the appearance of osteolytic images of variable size in the Clavicle and Sternum.*

It was decided to perform a combined anterior and posterior approach to the cervical spine, in two stages. In the first stage, a C7 corporectomy was performed and implantation with an autologous tricortical iliac crest graft, tumor excision and anterior fixation with lamina and screws. In the second stage, a C7 laminectomy was performed, fixation from C5 to T4 with blades and screws, and tumor excision. It is of special interest to mention that during the surgical procedures there was great bleeding from the bone structures that caused acute anemia and the need for intra- and postoperative transfusion of blood products to the patient, in addition that lesions of soft consistency, amber color, moderately vascularized, and hardly collapsible due to their infiltrating character, located in the body and spinous process of Vertebra C7, and in the right iliac crest (autologous graft donor site), all samples were sent for further pathological study. The biopsies reported a plasma cell tumor with the presence of a large number of plasmoblasts in the three samples sent. (Fig. 5).

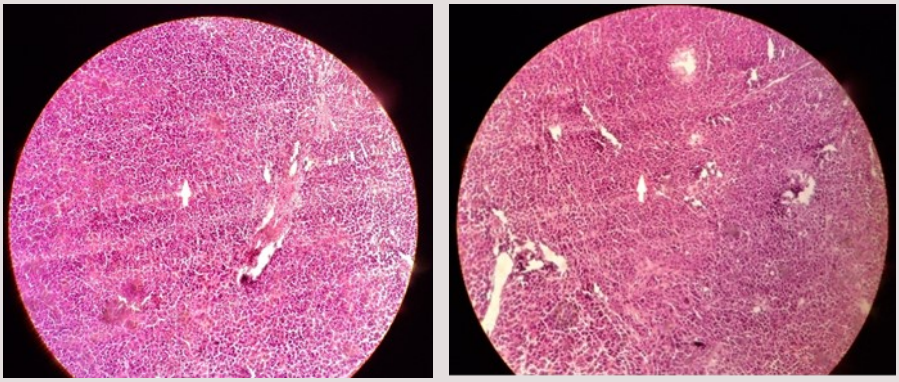

Fig 5. *Field seen in conventional microscopy of a sample taken from the vertebral body on the right and from the iliac crest on the left, hypercellularity is observed, constituted by tumor tissue with abundant atypical plasma cells that fill the visual field*

In the first postoperative week of the second surgical approach, the patient had completely recovered superficial and deep sensitivity, the muscular strength in the lower limbs increased to 3/5 according to the Daniels scale, and post-surgical evolutionary cervical spine tomography showed alignment of vertebral bodies, with correct position of blades and screws. (Fig.6)

When the patient was discharged, he was sent to Rehabilitation and adjuvant management by Hematology, being used in the treatment the VAD scheme: Vincristine, Doxorubicin (Adriamycim) x 1 day and Dexamethasone at a rate of 40 mg x 4 days, every 28 days, in total 6 cycles. At 12 months of follow-up, the patient returned to his work activities, even practicing non-demanding exercises, and manifested feeling asymptomatic; the physical examination showed improvement in the motor deficit with muscle strength of 5/5 in the lower limbs, recovery from ambulation , and the sensitivity both superficial and deep previously lacking, so far it does not present with any other related complication, and has not presented relapses.

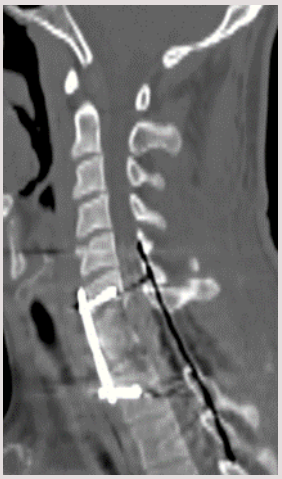

Fig. 6. *Sagittal section cervical spine tomography, under reconstruction. After 1 week post-surgery. Evidence of bone graft, blades and screws in proper position, with alignment of the cervical spine.*

Discussion and Conclusion

The pathologies that include the alteration in the monoclonal component are multiple, and sometimes they are difficult to differentiate from one another, having to meet the histological, clinical, imaging and other requirements that characterize them. These pathologies commonly become differential diagnoses of MM; Among them are monoclonal gammopathy of uncertain significance (MGUS), indolent, asymptomatic or quiescent multiple myeloma (MMQ), and solitary plasmacytoma (PSO). MGUS has a risk of 1% per year of progression to MM.[5,6,8,9] In 2008, the WHO granted the diagnostic criteria listed in Table 1 for monoclonal gammopathies. [1,6,8] It is taken into account that MM histologically is identical to plasmacytoma, and the difference between the two basically lies in the fact that the former is diagnosed when there is a single and isolated lesion in a specific bone tissue, but does not present other organic damage.[3.10]

Table:1

MULTIPLE MYELOMA (Symptomatic / Needs Treatment)	MONOCLONAL GAMMAPATHY OF UNCERTAIN SIGNIFICANCE (GMSI)
1. Presence of ≥10% clonal PC * in BM or a diagnostic plasmacytoma biopsy (bone or extramedullary) and 1 or more of the following criteria: to. Organic Damage Attributable to Myeloma (CRAB): • Hypercalcemia: serum calcium ≥1 mg / dL (≥0.25 mmol / L) of the upper limit of normal or ≥11 mg / dL (≥2.75 mmol / L) • Renal impairment: creatinine clearance <40 mL / min and / or serum creatinine ≥2 mg / dL (≥177 µmol / L). • Anemia: Hb <10 g / dL or> 2 g / dL below the lower limit of normal. • Bone lesions: one or more osteolytic lesions ≥5mm on CT, PET-CT or conventional X-ray **. b. One or more of these markers (even in the absence of CRAB): Clonal PC in MO ≥ 60% Altered serum free light chains (CLL): not altered ≥100 (in addition, the altered CLL must be ≥100 mg / L). Presence of> 1 focal lesion ≥5 mm on MRI.	They require the following 3 criteria: 1. Monoclonal component (CM) (IgG, IgA or IgM) in serum <3 g / dL. 2. Clonal plasma cells (CP) in bone marrow (MO) <10%. 3. No evidence of organic damage attributable to PC neoplasia (CRAB). **INDOLENT, ASYMPTOMATIC, or QUIESCENT MULTIPLE MYELOMA (MMq)** Requires the following 2 criteria: 1. Serum MC (IgG or IgA) ≥ 3 g / dL and / or urine MC ≥500 mg in 24 h and / or MO CP between 10–60%. 2. No evidence of organ damage attributable to PC neoplasia (CRAB) or amyloidosis

* Clonality: it will be established based on the restriction of κ / λ light chains by flow cytometry, immunohistochemistry or immunofluorescence. It is recommended to estimate the infiltration (in percentage) of PC by BM biopsy. In case of disparity between aspirate and biopsy, the higher value result should be used. ** If there is <10% clonal PC in BM,> 1 bone lesions are required to differentiate it from solitary plasmacytoma with minimal medullary infiltration

Table 1. Diagnostic criteria for monoclonal gammopathies according to who (2008)

In multiple myeloma, monoclonal protein (component M or protein M) can be evidenced by quantifying serum immunoglobulins, urine / serum protein electrophoresis or both, serum / urine immunofixation electrophoresis or both, total protein in urine of 24 hours. With the exception that said protein is considered to be measurable if it is ≥ 1 g / dL in serum and ≥ 200 mg / day in urine.)[2.6] Highlighting that in monoclonal gammopathies they present a monoclonal band, most of the time, located in the gamma globulin emigration zone, either at the beginning, in the middle or at the end, but exceptionally it can be found in the beta zone. or alpha-2 globulin. A pathologic monoclonal gammopathy is almost always accompanied by total hyperproteinemia (90-110 g / l). However, there is a small percentage that present with normoproteinemia, there may even be a monoclonal gammopathy with a normal quantitative gamma globulin.[eleven] Y Immunofixation and serum free light chain test will be indicated in patients with a strong suspicion of myeloma, with the presence of target organ damage and / or greater cellularity of plasma cells in the bone marrow, but serum protein electrophoresis of routine is negative (No detection of M protein). [5]

The bone disease observed in multiple myeloma, present in 80% of patients, reflects the existing imbalance between osteoblasts and osteoclasts, which end up forming osteolytic lesions, and is distinguished by severe bone pain (25%), pathological vertebral fractures (30%) and extravertebral (12%) and hypercalcemia. These skeletal events not only have a negative effect on the quality of life of patients, but also decrease their survival time.[5]

Regarding the treatment of MM, there are several proposed schemes. Among them, and for many years, the combination of vincristine, dexamethasone and adriamycin (VAD) has been used, the highest activity of the combination being attributed to dexamethasone, with an overall response of 55% to 65%; which can increase to an overall response of 91% if an analog of Thalidomide, such as lenalidomide, is incorporated into this scheme. However, it is demonstrated in the work by Koreth et al, that first-line autologous transplantation benefits both progression-free survival and overall survival, even double autologous transplantation is only recommended for those young patients who did not achieve a very good answer with the first.[12]

The case reported in this review presents a type of Multiple Myeloma with unusual characteristics, that said, due to two main factors, among others: the age of the patient at diagnosis, being well below the minimum age range referred to in this entity, Likewise, the result of his protein electrophoresis that the typical increase or monoclonal band in the Gamma globulin fraction was not evidenced (with normal values in it), if not, an increase in the level of Alpha 2 globulin with special attention to the presence splitting in the morphology of this fraction. Knowingly, that according to the literature, said pattern may signify the appearance of a monoclonal immunoglobulin band or a Bence Jones protein that migrated to this area;[11.12]. In the patient, the clinical form was marked by the evidence of plasmacytoma infiltrating the bone marrow from two different anatomical sites (vertebral body / iliac crest) and plasmacytosis was highlighted in the medullogram and the tumor lesion biopsy> 60%, but with negativity Monoclonal protein in serum, and Bence-Jons protein in urine; Likewise, the calculation of the free light chain index showed a slightly unequal Kappa / Lambda relationship, with an increase in Lambda over the non-appearance of Kappa as a sign of Myeloma. When carrying out an analysis of the aforementioned, the patient was included in the diagnosis of active, symptomatic, oligosecretory and / or light chain Multiple Myeloma, with grade 3 bone involvement,[2,6,7]also defining its treatment according to the therapeutic algorithms, establishing aggressive management, and auspicious follow-up to detect signs of disease progression. Hence the importance of thinking about this entity as a diagnosis in patients with vertebral fractures whose causal mechanism does not seem to obey the severity of the trauma suffered, and therefore act in a timely manner, looking for this type of pathologies, and thus be able to reduce morbidity and mortality and increase survival when initiating appropriate and timely treatment.

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