Case Report

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Ankylosing Spondylitis as a Deceptive Visage of Multiple Myeloma: A Case Report

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ABSTRACT

A 38-year-old male came to outpatient clinic complaining of a new-onset pain of his right buttock which has caused him to limp. Since 6 months ago, the pain had involved in order his cervical spine, ribs, sternum, and thoracic spine, and left them deformed. He also had constitutional symptoms and significant weight loss. On physical examination, there was a limited chest expansion, positive Schober's test, and multiple tender bony bumps along the ribs, costosternal and costochondral junctions. Laboratory tests showed anemia, elevated erythrocyte sedimentation rate, normal level of serum calcium, and creatinine, and 1+ protein in urinalysis. Serum protein electrophoresis was unexceptional. The imaging of the patient changed the diagnostic path. This article describes a deceptive visage of multiple myeloma presenting itself in the clothing of ankylosing spondylitis

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Introduction

Multiple myeloma is a neoplastic disorder of a single clone of plasma cells which produce monoclonal immunoglobulin. It is characterized by the proliferation of this clone of plasma cells in the bone marrow, which commonly invade the adjacent bone resulting in skeletal destruction with osteolytic lesions, pathologic fracture, and bone pain (1). The excessive amount of monoclonal immunoglobulin (M-protein) in urine can cause renal failure from Bence Jones proteinuria and, in serum, hyperviscosity (2).

The diagnosis is based on a clonal bone marrow with plasma cells ≥ 10% or biopsy-proven bony or extramedullary plasmacytoma and one or more of myeloma defining events: hypercalcemia (serum Ca >11 mg/dl), renal failure (serum creatinine > 2 mg/dl), anemia (Hb < 10 mg/dl), and bone lesions (3).

This article reports a patient which presented first with features of ankylosing spondylitis, but in a few months showed the full-blown picture of multiple myeloma. Ankylosing spondylitis is a chronic inflammatory disease, which affects axial skeleton with the consequence of progressive spine stiffness. It is manifested by back pain, postural abnormalities and spinal deformities (4) as in our patient.

Case Report

Not a month after the Persian New Year Nowruz, a 38-year-old male came to Imam Khomeini Hospital outpatient clinic seeking medical attention for "a crippling bone pain impeding his life" as he described. The pain had first commenced in the cervical spine 6 months before, but progressive in severity and additive in nature, moved further to involve the left ribs downwardly and then the same story had happened to his right hemithorax. The pain continued to involve in order the sternum, thoracic spine, and recently his right hip which has caused him to limp and come to doctor. It was more severe at night but improved upon arising and by doing daily activities. The full eccentricity of the pain is appreciated by adding the fact that, in each step, the pain not only emigrated the previous site to its new destination but also left its former dwelling deformed so that the patient developed pectus carinatum, thoracic kyphosis, and multiple tender bumps and irregularities along the spine and his ribs in only a couple of weeks (Figure 1). He had no complaint of anorexia, fever and chills, night sweats, and peripheral joint pain but had a significant weight loss of 20 kg (30%) in 6 months. His past medical and surgical histories were negative. He had no history of recent trauma and did not report any similar history in his family. He was a smoker (36 pack-year) and also opium-addicted.

On examination, the patient was a young wasted man who appeared pale and cachectic with bitemporal atrophy. He was not ill, and his vital signs were normal. He was underweighted (body mass index = 17.6). His conjunctivae were pale. No significant cervical, axillary, epitrochlear, and inguinal lymphadenopathy were detected. Pectus carinatum was evident, and chest palpation also revealed multiple moderately tender bumps and irregularities in the contours of the ribs and sternum including costosternal and costochondral junctions suggestive of enthesitis.

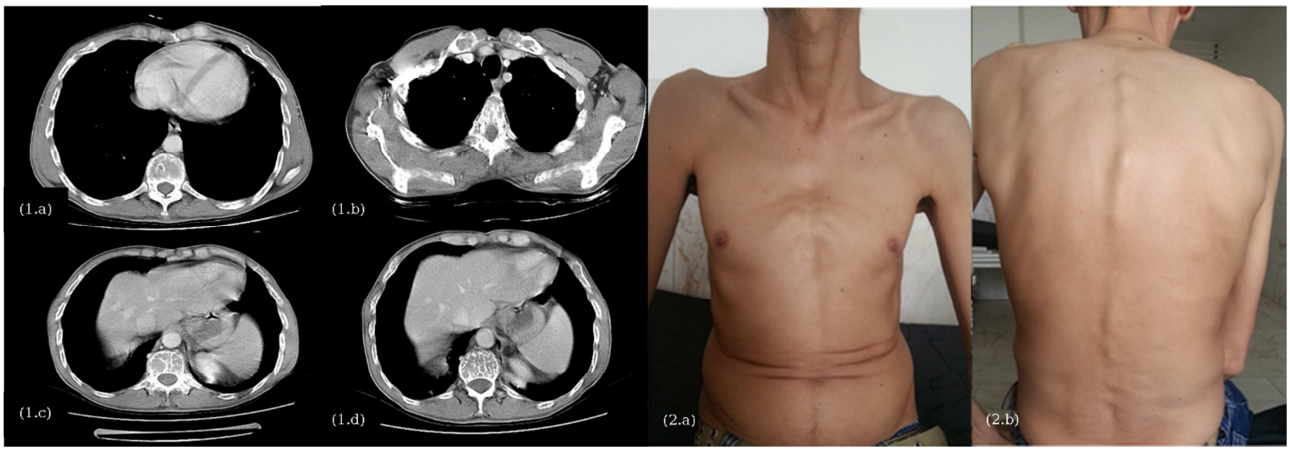

Figure 1. Spiral chest computed tomography scan of the patient showing multiple lytic foci in (1a, 1c) thoracic and lumbar vertebrae (1b) clavicles, (1d) ribs. Pectus carinatum (2a) and thoracic kyphosis (2b) are evident

The chest expansion was limited (3 cm) though symmetric. Despite the noticeable thoracic kyphosis, the occiput to wall test was normal. Schober's test was positive. The upper thoracic vertebrae were found to be tender on percussion. There was no sacroiliac tenderness. The tone and strength of muscles of the extremities were intact, and full examination of peripheral joints produced no positive result. The right hip joint though painful was normal on examination. The physical examination was further unexceptional.

A summary of important findings of the patient's laboratory data can be shown in table 1. It is to be noted that his serum creatinine, blood urea nitrogen, aspartate aminotransferase, and alanine aminotransferase were never elevated.

Imaging studies further unmasked more noxious pathologies. In spite of a normal pelvis plain radiograph, thoracolumbar spine showed degenerative changes, L_5 sacralization, collapse of T_{11}, and L_1 wedging. The spiral chest computed tomography (CT) scan with intravenous contrast showed multiple lytic foci in spine, ribs, scapula, sternum, clavicles, and humeri (Figure 1). The dual X-ray absorptiometry (DXA) scan of lumbar spine and femoral neck revealed osteoporosis with T-scores of −3.9 and −2.7, respectively. The ultrasonography of abdomen, pelvis, testes, thyroid gland, and cervical lymph nodes revealed no pathology. The results of capillary serum protein electrophoresis (SPEP) were non-specific: 1.5% increase in α_2 and 3.4% decrease in γ (0.64 g/dl) components. However, urine protein electrophoresis (UPEP) proved more decisive: there was remarkable Bence-Jones (light chain) proteinuria (57.8 mg/dl or 60.2%), immunoglobulin G (IgG) 3.2% (3.1 g/dl), and IgM 1.4% (1.4 g/dl). The SPEP was repeated with the results being much the same as the previous study: 5.7% increase in α_2 and 2.8% decrease in γ (0.8 g/dl) components.

Table 1. Represents the laboratory finding of the patient

Laboratory data	Patient	Normal range
RBC	3.11×10^6/µl	$4.5\text{-}5.8 \times 10^6$ µl
WBC	9.1×10^3 µl	$4\text{-}10 \times 10^3$ µl
PLT	162×10^3 µl	$150\text{-}450 \times 10^3$ µl
Hb	9.9 g/dl	14-17 g/dl
MCV	90 fl	80-100 fl
MCHC	35.1 g/dl	31 g/dl
Serum Ca	9.5 mg/dl	8.4-10.6 mg/dl
Serum phosphorus	3.6 mg/dl	2.5-5.0 mg/dl
Serum iron	134 µg/dl	60-150 µg/dl
TIBC	355 µg/dl	300-360 µg/dl
Serum ferritin	311 ng/ml	50-200
Transferrin saturation	37.7%	30-50
HLA-B27	Negative	-
25-OH vitamin D	2 ng/ml (very deficient)	< 20: Deficient
U/A	1+ Protein	Pr: negative
24 hours-urine protein	311 mg	< 150 mg
LDH	265 U/l	< 480 U/l
CPK	51 U/l	46-171 U/l
PSA	0.3 ng/ml	< 2.5 ng/ml
ESR 1st	47 mm/h	< 15 mm/h
ALP	320 IU/l	80-306 IU/l

RBC: Red blood cell, WBC: White blood cell, PLT: Platelet count, Hb: Hemoglobin, MCHC: Mean corpuscular hemoglobin concentration, TIBC: Total iron binding capacity, U/A: Urinalysis, LDH: Lactate dehydrogenase, CPK: Creatine phosphokinase, PSA: Prostate specific antigen, MCV: Mean corpuscular volume, ESR: Erythrocyte sedimentation rate, ALP: Alkaline phosphatase

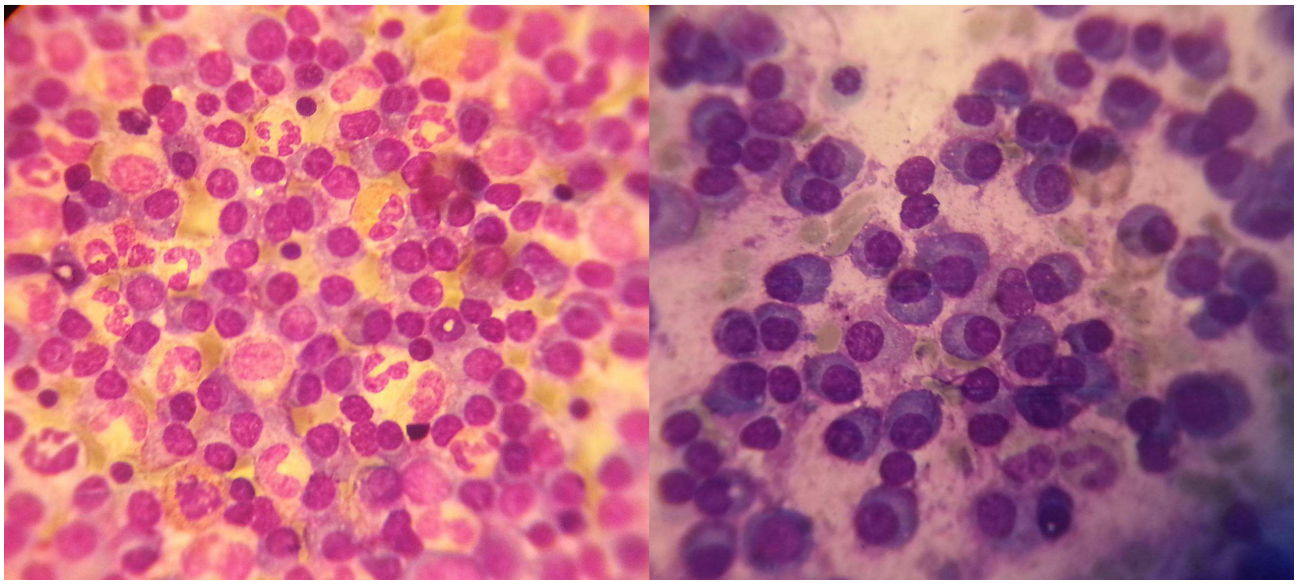

Figure 2. The bone marrow biopsy of the patient showing a preponderance of mature-appearing plasma cells (H and E, left: ×400, right: ×1000)

It was interesting that not only there appeared nothing as an M component in SPEP but also it was diminished below the normal range. To achieve the definitive diagnosis, bone marrow aspiration/biopsy (BMB) was performed. Meanwhile, serum-free light chain (FLC) assay was requested, which indicated a decreased λ-FLC of 4 mg/dl (5.7-26.3) and normal κ-FLC of 10 mg/dl (3.3-19.4) with the κ to λ ratio being 2.5 (0.26-1.65) which is highly suggestive of abnormal or tumoral plasma cell activity (1). The diagnosis of multiple myeloma (MM) was eventually validated by BMB result of 70% plasmacytosis (Figure 2).

Discussion

This case describes a rare presentation of MM as ankylosing spondylitis (AS). To the best of our knowledge, no similar case has so far been reported except one which is discussed below.

MM is well known as a disease of older adults with the median age at diagnosis being (3). However, in our case, the age of presentation was 38-year-old, which is instead in favor of AS, known as a disease of younger adults with the median age of 23 years (5). It is worth noting that while only a minority of MM patients (2%) are affected under the age of 40, almost all (nearly 95%) of the patients afflicted with AS are below 40 years (2, 5).

According to International Myeloma Working Group (IMWG) updated criteria for the diagnosis of MM, the defining events of MM include calcium, renal insufficiency, anemia, and bone (CRAB) features, i.e., hypercalcemia, renal failure, anemia, and bone lesions (3); of those, our patient had solely two, namely bone lesions and anemia. Serum calcium and renal function were intact. The observable normocytic normochromic anemia (normal iron profile) is in congruence with MM, as it occurs in 80% of MM patients. Bone pain is established to be the most common symptom of MM, affecting 70% of patients (6). Our patient had bone pain, but his pains were all clearly of inflammatory nature and totally incongruent with myeloma pain, as they were worse at night and improved during the day and with exercise, not by rest. It is also well known that the most characteristic clinical presentation of AS is inflammatory back pain (7). Although the initial involvement of the disease was not typically located in the lower back or buttocks, and in fact, his cervical spine and thoracic cage were the prime targets of the malady, his principal reason to come to the

doctor was his new-onset right buttock pain. However, it is well established that sometimes pain and stiffness in the cervical and mid-thoracic spine or chest wall might be the initial presentation in AS patients (7). Bacterial infections are the second most common clinical problem in MM patients. Our patient had, nevertheless, no past history of infection (6). He also had significant weight loss, which occurs in 24% of MM patients and though not frequently, it is also the case, along with other constitutional symptoms, in AS (2).

It is known that bony tenderness of enthesitis is the predominant sign in some AS patients (7). The tender bony bumps along the patient's ribs, costosternal and costochondral junctions, and also tender thoracic spinous processes were suggestive of enthesitis, particularly, as they are common places of it. The limited chest expansion and positive Schober's test were also in favor of AS, while lack of sacroiliac joint tenderness was against it.

Thus far, the overall clinical image depicts a misleadingly rosy picture of a patient with AS, but the imaging modalities would offer the physician a helping hand. There was no typical involvement of the sacroiliac joint in the plain radiograph of pelvis, but it is well known that it could be normal during the first initial years of the disease as in a study of newly diagnosed AS patients, only 33% of the patients had typical AS radiographic changes (8, 9). Similarly, none of the AS-specific radiographic changes of spine was seen, and instead, there were observed collapse and wedging of two vertebrae and multiple lytic foci in CT scan of spine, ribs, and other bones, not explained by AS. Casting serious doubts on the diagnosis of AS, these lytic bone lesions put the metastatic bone disease on the top of the list of differential diagnoses and so the ultrasonography of probable sites of malignancy was performed and PSA was checked though no pathology was detected. According to the IMWG criteria, osteolytic lesion(s) < 5 mm (B of CRAB) fulfills the end organ damage criterion by itself (3). The patient also had severe osteoporosis on DXA which is frequently seen in MM though it is also a common complication of AS (2, 10). The definitive diagnosis of MM was finally established based on the IMWG updated criteria with its three criteria being met: (11) presence of M-protein in UPEP, (3) bone marrow clonal plasma cells infiltration of at least 10% (70% in our case), and (5) presence of CRAB features (3).

It is also worth mentioning that the decrease seen in γ component in SPEP, which remained constant even with repetition and made the diagnosis further complex, is an event observed in one among every 10-20 myeloma patients, namely the "oligosecretory myeloma group" (2). A similar case was found in the literature describing a 44-year-old male with a history of lower back and buttock pain, and radiographic evidence of sacroiliitis – with a presentation of AS - which was further revealed to be MM. However, he had obvious M component in SPEP, which made the diagnosis more straightforward (12).

Conclusion

This article describes a deceptive visage of MM presenting itself in the clothing of AS. It emphasizes the fact that MM, malignancy, can atypically though rarely, manifest itself as AS, a rheumatologic disease. The timely diagnosis will not be achieved unless the physician thinks of such a possibility in advance.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

None.

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