MULTIPLE AUTOIMMUNE SYNDROME

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Background: Multiple autoimmune syndrome (MAS) is a rare entity, defined by the association of three autoimmune diseases in the same patient. MAS can be classified into three groups.

Objectives: The objective of this work was to describe the autoimmune diseases profile in MAS in an Internal Medicine department.

Methods: We report a retrospective analysis including 14 cases of MAS seen in The Internal Medicine department at Tahar Star Hospital, Mahdia, TUNISIA over a period of 10 years.

Results: We followed 14 patients with MAS. They were 14 women. The mean age of patients was 52 years. SAM was type 3 in 12 patients (85%), type 2 in one patient (7.1%) and one patient satisfied both type 2 and type 3 MAS criteria (7.1%). No cases of MAS type 1 had been reported. We found 13 patients with 3 associated autoimmune diseases and one patient with 4 associated autoimmune diseases. The autoimmune diseases were: Sjögren’s syndrome in 14 patients (100%), Hashimoto’s thyroiditis in 11 cases (78%), systemic lupus erythematosus in 7 cases (50%), Addison’s disease in 4 cases (28.5%), scleroderma, rheumatoid, and renal neoplasms. Sternal puncture showed a rich bone marrow of normal cells without further infiltration. Bone biopsy of the detected lesion showed tumour cells made of mature plasmocytes confirming the diagnosis of solitary plasmocytoma of the bone. Without progression to multiple myeloma and keep a hawk-eyed guard.

Disclosure of Interests: None declared

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A RARE CASE OF A SOLITARY PLASMOCYTOMA OF A LUMBAR VERTEBRA

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Background: Solitary plasmocytoma is a rare tumour that represents around 2 to 5% of all plasma cell dyscrasias. It normally affects soft tissue but rarely the bone. Diagnosis is based on histology, the absence of bone marrow involvement.

Objectives: To drag attention to think of solitary plasmocytoma of bone when dealing with a vertebral fracture in the absence of the CRAB criteria of multiple myeloma.

Methods: We report a rare case of a vertebral fracture of the 4th lumbar vertebra (L4) revealing a solitary plasmocytoma of bone.

Results: A 67-year-old female patient presented to our rheumatology department with back and left radicular pain of brutal onset, 15 days prior to her visit. Pain was severe and awakened her at night. On examination, mobility of the spine was unchanged but on palpation she had exquisite pain of L4. Laboratory tests showed a normal sedimentation rate of 15mm the first hour, a negative c-reactive protein, normal calcemia and kidney tests. X-rays of lumbar spine showed a vertebral fracture with a destruction over 50% of the vertebral size and cortical rupture. MRI of the spine showed the absence of other lesions or other fractures or spinal cord compression and showed the total destruction of the anterior vertebral body of L4 (Figure 1). Protein electrophoresis was in normal range and 24h urinary proteinuria was negative. Other tests rules out gynecological, thyroid, and renal neoplasms. Sternal puncture showed a rich bone marrow of normal cells without further infiltration. Bone biopsy of the detected lesion showed tumour cells made of mature plasmocytes confirming the diagnosis of solitary plasmocytoma of the bone. The patient was treated with radiation therapy. The evolution after 24 months showed a stabilised lesion and the absence of progression to multiple myeloma.

Conclusion: It is important to keep in mind the diagnosis of solitary plasmocytoma of bone when facing a solitary lesion or vertebral fracture despite relatively non aggressive radiological signs. It is also important to note the possible evolution to multiple myeloma and keep a hawk-eyed guard.

REFERENCES:

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CAN INFLAMMATION COEXIST IN PATIENTS WITH PROGRESSIVE PSEUDORHEUMATOID DYSPLASIA?

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Background: Progressive pseudorheumatoid dysplasia (PPRD) is considered as a degenerative genetic bone disorder. It is caused by loss of function mutation in WNT-1 inducible signaling pathway protein-3 (WISP-3). WISP-3 gene function is required for the normal function of cartilage and skeletal development. The patients are normal at birth and start developing symptoms around 3-6 years of age. The disease is characterised by stiffness, pain, deformity due to enlargement of the ends of short and long bones. Often, such patients are misdiagnosed as Juvenile idiopathic arthritis (JIA). In general, PPRD being considered as non-inflammatory disease, immunosuppressants or disease modifying anti rheumatic drugs (DMARDs) like methotrexate treatment are not used for treatment.

Objectives: We report a patient with characteristic findings of PPRD but with coexisting clinical and imaging evidence of inflammation.

Methods: 16 year old male boy born of third degree consanguineous asymptomatic parents presented with progressive swelling, deformity of bilateral small and large joints of upper and lower limbs. He also had pain in both hip and knee for past two years. Pain is associated with difficulty in walking and squatting. On examination he had bony enlargement around bilateral elbow, wrist, proximal and distal interphalangeal joints(Figure 1A). He also had restriction of bilateral hip movements and swelling of bilateral knee with effusion. He had exaggerated lumbar lordosis and flexion deformity of bilateral hip, knee. His blood counts, ESR, CRP were normal. Analysis of Knee joint synovial fluid showed cell count of 200/mm3 with no crystals and sterile culture. USG knee showed evidence of synovial thickening with increased power Doppler signals. Skeletal survey showed typical findings of PPRD with enlargement of epiphysis and osteoarthritis.