



CASE REPORT

Biclonal Gammopathy of IgA Kappa Variant in a Patient with Systemic Lupus Erythematosus: A Case Report

Hicham Esselmani^{1*}, Asmaa Yassine², Boutaina Akhtar³, Otmane Touzani⁴ and Abderrazak Lfakir⁵

¹HAFIR Laboratory of Medical Analysis, Morocco

²Medicine Service A, Regional Hospital of Beni Mellal, Morocco

³Laboratory of Medical Analysis, Provincial Hospital of Fkih Ben Salah, Morocco

⁴TOUZANI Laboratory of Medical Analysis, Boulevard of Moulay Ali Cherif, Morocco

⁵Faculty of Sciences and Techniques, University My Slimane, Morocco

*Corresponding author: Hicham Esselmani, HAFIR Laboratory of Medical Analysis, Lakouacem, street 28 n°12. Fkih Ben Salah, Morocco



Abstract

The association of systemic lupus erythematosus and multiple myeloma is an uncommon event. Biclonal gammopathy is a rare case, which accounts for only 1.5% of all myelomas but biclonal gammopathy of IgA kappa variant is extremely rare. We report this unusual case in which the simultaneous diagnosis of biclonal gammopathy of IgA kappa variants and systemic lupus erythematosus was made.

Keywords

Systemic lupus erythematosus, Multiple myeloma, Biclonal gammopathy, IgA, Kappa chain

Introduction

It has been suggested that patients affected by autoimmune diseases are more prone to developing malignancies, and especially lymphoma [1]. Although the coexistence of multiple myeloma (MM) and systemic lupus erythematosus (SLE) has been described in humans, it remains extremely rare [1]. MM is characterized by the production of M protein. The monoclonal protein is detected by serum protein electrophoresis as a single discrete band (M-band) most often in the gamma globulin region. However, it may rarely show a simultaneous presence of two distinct M-bands as seen in 1.5% of cases [2]. We report a case of simultaneous diagnosis of biclonal gammopathy with

a rare combination of immunoglobulin (IgA/IgA Kappa) in a 56-year-old man and SLE.

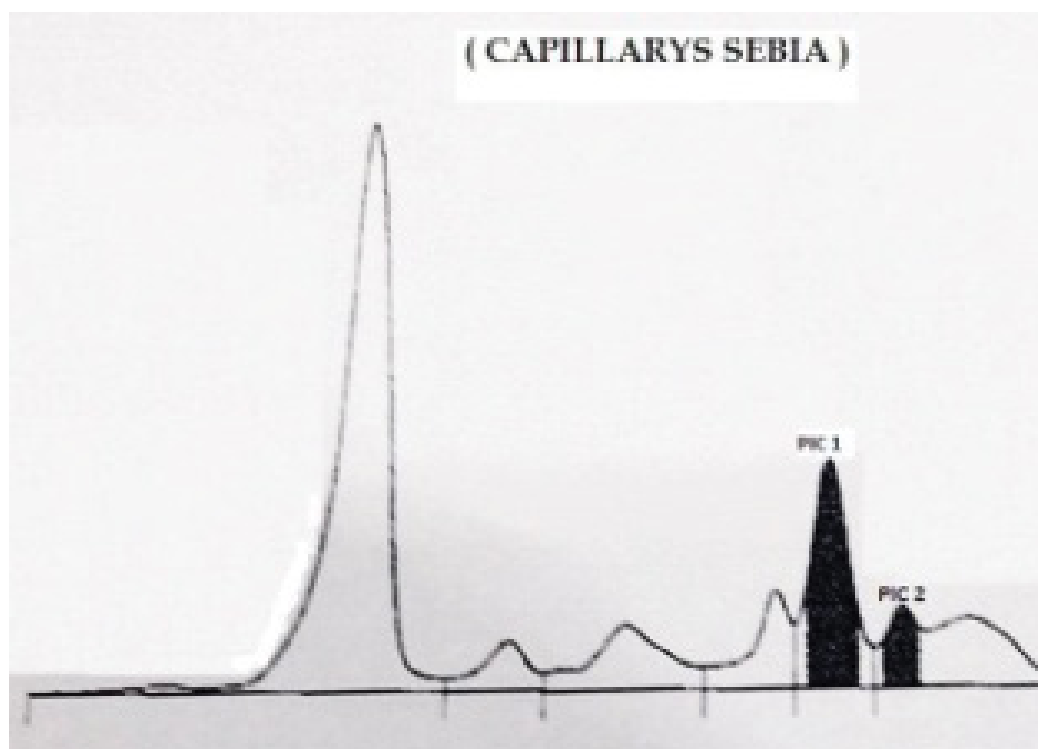
Case Report

A 56-year-old man has a past medical history of renal failure (secondary to type 2 diabetes diagnosed 11 years before) manifesting fatigue, general weakness, undocumented weight loss, a photosensitive rash and intermittent arthralgias to the hospital. His temperature was 38.5 °C, heart rate was 72 beats/min, and blood pressure was 135/85 mmHg. Physical examination revealed persistent malar erythema and transient indurated erythema on the upper extremities and trunk. He had non-erosive arthritis on shoulders, elbows, and knees. He did not present with swollen fingers, alopecia, lymphadenopathy or oral ulcers.

Complete blood count, blood biochemical parameters and auto-immune serologic test results are detailed in Table 1. Serum electrophoresis on agarose gel (Sebia) revealed two sharp discrete bands. Band-1 (10.21g/l) is seen in the Beta 2 region and Band-2 (3.03g/l) is seen in the gamma region (Figure 1). Serum electrophoresis repeated after the serum pretreated with beta mercaptoethanol showed two distinct bands. On subjecting to the Immunofixation, it is found that both bands are of IgA-Kappa type (Figure 2). The urine was strongly positive (++) for protein.

Table 1: Laboratory findings.

Variable	Results	Reference values
Red-cell count (*10 ⁶ /μL)	4.37	4.1-6
Hemoglobin (g/L)	130	130-170
Hematocrit (%)	0.88	0.8-0.95
White-cell count (*10 ⁹ /L)	6.6	4.0-10
Platelet count (*10 ⁹ /L)	126	150-450
Erythrocyte sedimentation rate (ESR) (mm/h)	93	< 10
Urea nitrogen (mmol/L)	14	3.6-9.5
Creatinine (μmol/L)	318	60-130
Proteinuria (g per 24 h)	0.83	< 0.15
ANA	Positive (1:640)	Negative
Anti-dsDNA (AU/ml)	183	< 25
Anti-Sm (U/ml)	1.19	< 25
Anti-SSA (U/ml)	5.34	< 25
Anti-SSB (U/ml)	2.9	< 25
Anti-Phospholipid (U/ml)	1.43	< 10



Fractions	%	ref.%	g/l	ref g/l
Albumine	51,5	< 55,8-66,1	38,11	40,20-47,60
Alpha 1	4,0	2,9-4,9	2,96	2,10-3,50
Alpha 2	8,5	7,1-11,8	6,29	5,10-8,50
Beta 1	6,5	4,7-7,2	4,81	3,40-5,20
Beta 2	15,6	> 3,2-6,5	11,54	2,30-4,70
Gamma	13,9	11,1-18,8	10,29	8,00-13,50
Pic etroit 1	13,8		10,21	
Pic etroit2	4,1		3,03	
A/G	1,06		Protein total: 74	

Figure 1: Serum protein electrophoresis shows Band-1 in beta2 region and Band-2 in gamma region.

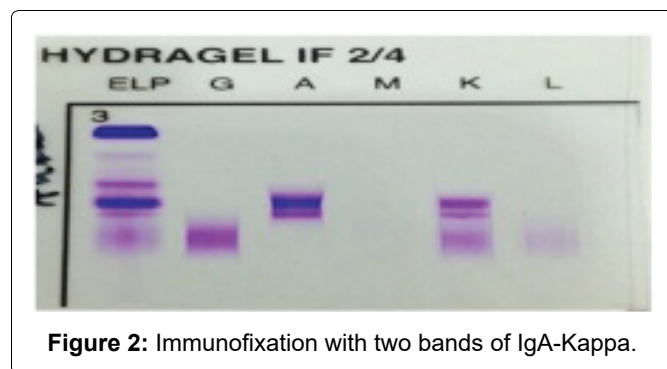


Figure 2: Immunofixation with two bands of IgA-Kappa.

Immunofixation performed on the urine demonstrated a kappa light chain not associated with IgA, IgG, or IgM heavy chains. Renal biopsy is not practiced. Red blood cells morphology showed anisocytosis and no rouloux formation. The bone marrow biopsy demonstrated normocellular marrow with a diffuse interstitial infiltrate of plasma cells comprising more than 30% of the marrow elements.

Autoimmune serologic test results showed positive tests for antinuclear antibodies (ANA) (title 1:640, homogenous pattern) and antibodies to double stranded DNA (dsDNA) (183AU/ml). Anti-Phospholipid (Antibeta-2-Glycoprotein, Anticardiolipin, Antiphosphatidic acid, Antiphosphatidyl inositol and Antiphosphatidyl serine) antibodies were negative (Table 1).

SLE was diagnosed according to criteria for SLE set by the American College of Rheumatology (ACR). Based on the above, the simultaneous diagnosis of biclonal gammopathy of IgA kappa variants and systemic lupus erythematosus was made.

Discussion

Although monoclonal gammopathy is often detected in SLE patients the coexistence of MM has rarely been reported. The prevalence of monoclonal gammopathy of undetermined significance (MGUS) in SLE patients varies from 2.2% to 3.3% and the outcome of these patients seems not to differ from the other SLE patients [3].

MM possibly diagnosed roughly a decade after the diagnosis of SLE is made, but it may be diagnosed simultaneously as in our case or even the MM precedes the diagnosis of SLE. A careful literature review confirms that the association of these two diseases has been reported only in a few cases. Pehamberger, Sendagorta, Vaiopoulos and Humayun diagnosed SLE and MM simultaneously [3-6]. To our knowledge, this is the first ever-reported case with biclonal gammopathy of IgA kappa variant and simultaneous SLE.

The pathogenesis of the association between these two disorders is mainly speculative. The genetic factors have been claimed to play a role in the pathogenesis, while some other researchers think that it is the prolonged use of immunosuppressant drugs in previously diagnosed SLE patients which causes MM to

develop [3].

Biclonal gammopathies are a vanishingly rare group characterized by the production of two distinct monoclonal proteins. This rare finding can result, for example from either a proliferation of two clones of plasma cells with each producing an unrelated monoclonal spike or from the production of two monoclonal spikes by a single clone of plasma cells. Approximately 1.5% of MM cases present with biclonal paraproteinemia [2].

IgA found selectively in the seromucous secretion, tears, saliva and GI secretions. IgA has two subclasses which are differentiated immunochemically as IgA1 and IgA2. Normally the serum contains approximately 90% of IgA1 molecules [7].

Donald R. Hoffman, Muhammad Younas, Kumar ML and Yadav were reported a case of biclonal gammopathy of IgA-kappa variant [7-10].

Conclusion

Our case report highlights an extremely rare clinical presentation of biclonal gammopathy of IgA kappa variant and SLE. The association of SLE and MM has rarely been studied so more research needs to be done in this regard so that the definite pathogenetic mechanisms involved can be established and the effective treatment guidelines could be devised.

Conflict of Interest

None of the authors has any conflict of interest to disclosure.

Authors Contributions

All the authors have read and agreed to the final manuscript.

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