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IgG,kappa monoclonal gammopathy of unknown significance with AL amyloidosis simulating giant cell arteritis

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Monoclonal gammopathies complicated by AL amyloidosis can mimic giant cell arteritis (GCA). We hereby present the case of a 63 year old woman in whom symptoms consistent with GCA were the first manifestations of a monoclonal gammopathy of unknown significance (MGUS) associated with amyloidosis.

A 63 year old woman was admitted for temporal headache, maseterine claudication, neck and shoulder stiffness. She was recently diagnosed with carpal tunnel syndrome. On physical examination she had prominent temporal arteries, macroglosia and orthostatic hypotension. Muscular strength was normal. She had high ESR and CRP; in this clinical context, GCA was suspected. A gamma spike on serum protein electrophoresis raised the suspicion of monoclonal gammopathy (MG). Immunoelectrophoresis revealed monoclonal bands for IgG and kappa chains. Massive deposits of amyloid and no inflammation were found on temporal artery biopsy. Multiple myeloma and lymphoma were ruled out. A diagnosis of AL amyloidosis complicating MGUS was formulated. She did well on therapy with bortezomib, cyclophosphamide and dexamethasone.

Cases published in medical literature reveal amyloidosis mimicking GCA in the setting of established MGUS. As far as we know, this is the first case of MGUS with IgG and kappa chains in which a GCA-like picture induced by amyloidosis was present from the very onset.

Key words: gammopathy, IgG, kappa chains, amyloidosis, giant cell, carpal tunnel, temporal artery, biopsy.

BACKGROUND

Monoclonal gammopathies (MG), such as: myeloma, Waldenstrom's macroglobulinemia and monoclonal gammopathy of unknown significance (MGUS) can masquerade giant cell arteritis (GCA). Usually GCA-like features develop in the setting of established MG. Very rarely they may be present from the very onset. We hereby present the case of a 63 years old woman in whom symptoms consistent with GCA were the first manifestations of a monoclonal gammopathy of unknown significance (MGUS) associated with amyloidosis.

CASE PRESENTATION

A 63 year old woman was admitted in our clinic because of maseterine claudication, odynophagia, symmetrical pain and stiffness in the neck and shoulders, recently aggravated headaches, mild weight loss (4 kg in the last 6 months). She also complained of orthostatic light headedness and also of xerostomia/xerophthalmia. Previous recent investigations disclosed: serum normal muscular enzymes, intense biological inflammatory syndrome, important hypergammaglobulinemia – with an electrophoretic peak (estimated at 0.7 g/dL), electroneuromyography-bilateral carpal tunnel syndrome, no signs of muscular lesions and no increment or decrement on repetitive stimulation.

On clinical examination she had normal weight; prominent temporal arteries but without local signs of inflammation; macroglosia with tooth indentations (Figure 1); muscular strength was normal; Tinel's sign was slightly positive bilaterally; hepatosplenomegaly; she had orthostatic hypotension. Peripheral lymph nodes were not palpable.

Blood tests showed (Table 1): slight normochromic hyposideremic anemia (attributable to chronic inflammation); moderate biological inflammatory syndrome; immunoelectrophoresis disclosed monoclonal increase in IgG and kappa chains. Bone marrow aspirate showed a hypercellular aspect with reactive plasmocytosis (8%).

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Temporal artery biopsy revealed massive deposits of Congo red-staining amyloid (Figure 2) with green birefringence in polarized light (Figure 3), but no inflammatory changes. X-rays of skull and pelvic bones revealed no lytic lesions. Computed tomography of the thorax, abdomen and pelvis showed no adenopathy, no hepatosplenomegaly and no other possibly malignant lesion.

Table 1
Patient's blood tests

Blood test	**Value**
WBC (/mmc)	6250
Hb (g/dL)	11.2
PLT(/mmc)	260000
ESR(mm/1h)	43
CRP(mg/L)	13.36
Glucose(mg/dL)	108
Creatinine (mg/dL)	1.11
GOT(UI/L)	24.4
GPT (UI/L)	13.6
Serum immunoelectrophoresis	Monoclonal bands for IgG, kappa

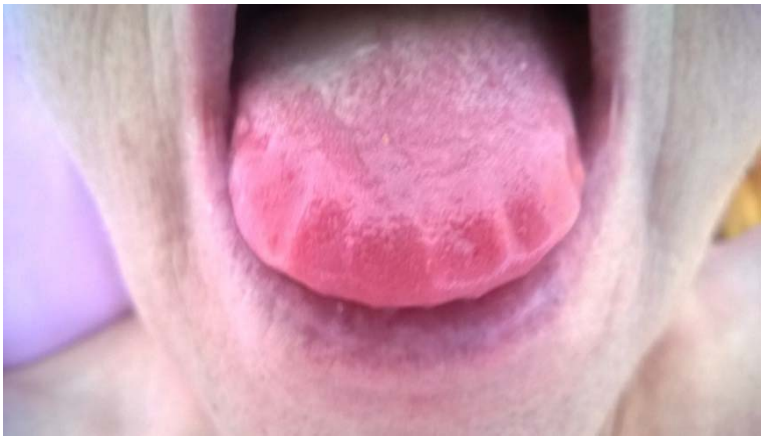

Figure 1. Macroglosia, with dental indentation.

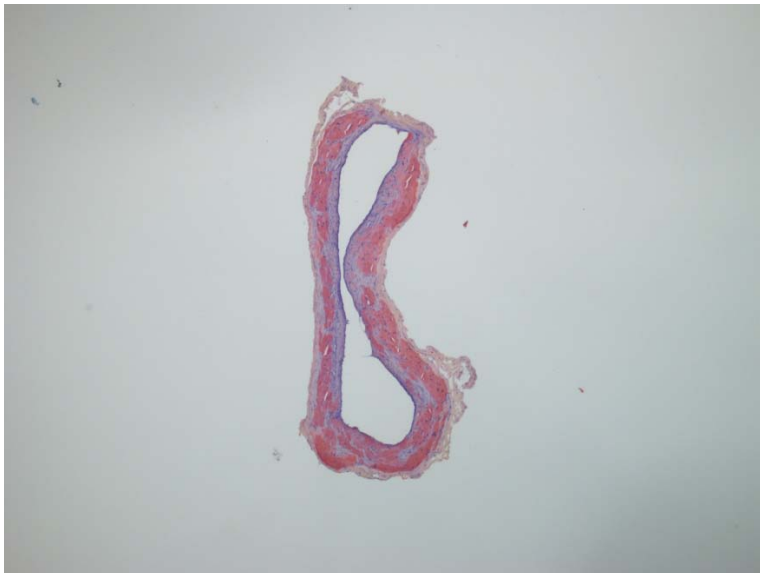

Figure 2. Temporal artery biopsy- Congo red staining: massive amyloid deposits.

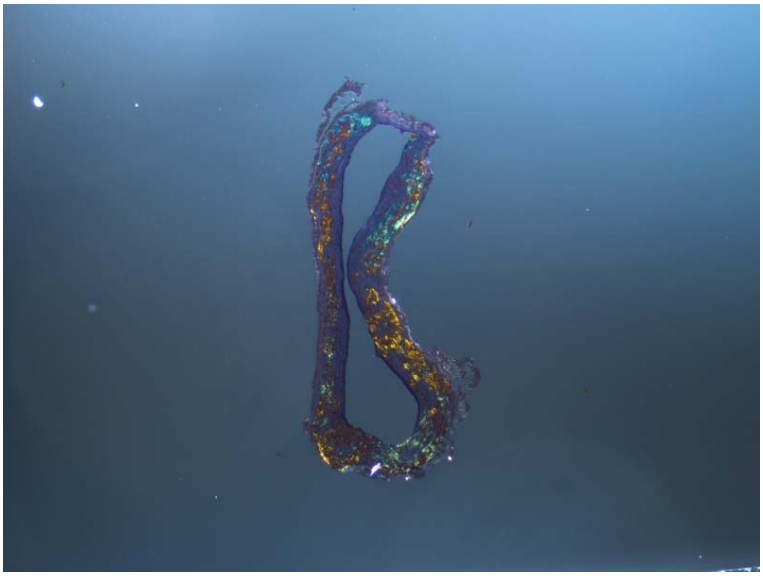

Figure 3. Same patient, temporal artery biopsy: amyloid deposits showing green birefringence in polarized light.

A monoclonal IgG-kappa increase with no arguments for myeloma or lymphoma sustained the diagnosis of MGUS. Histologically proven amyloidosis was responsible for temporal headache, maseterine claudication, neck and shoulder stiffness – which initially suggested giant cell arteritis; amyloidosis also caused carpal tunnel syndrome, macroglosia, hepatosplenomegaly, sicca symptoms and orthostatic hypotension.

Therefore, she was transferred in a hematology clinic where she was treated with bortezomib, cyclophosphamide and dexamethasone with favorable results.

DISCUSSION

The possibility that AL amyloidosis complicating a MG may be a mimicker for GCA is not a new item. However, most published cases were in the context of established multiple myeloma [2, 3] or established MGUS [1]. As far as we know, it is the first case of MGUS with IgG and kappa chains in which a GCA-like picture induced by amyloidosis was present from the very onset. The multitude of symptoms explainable by amyloidosis suggest widespread disease.

CONCLUSIONS

1) AL amyloidosis complicating MG may be a mimicker of GCA

2) AL amyloidosis may be encountered in all forms of MG:multiple myeloma, Waldenstrom's macroglobulinemia, MGUS .

3) Most often AL amyloidosis develops in the setting of established MG; rarely it may be present from the very onset (such is our patient's case).

4) We highlight the importance of performing Congo red staining of temporal artery biopsies if diagnosis of GCA is doubtful

5) Distinction between AL amyloidosis and GCA must be performed quickly because the treatment is radically different.

Gamapatiile monoclonale complicate de amiloidoză tip AL pot mima arterita cu celule gigante (CGA). Prezentăm cazul unei paciente de 63 de ani la care simptomele ce mimau CGA s-au dovedit a fi manifestări ale gamapatiei monoclonale de cauză necunoscută (MGUS) ce asocia amiloidoză.

O pacientă de 63 de ani a fost internată pentru cefalee, claudicație maseterină, redoare de umăr. Ea fusese recent diagnosticată cu sindrom de tunel carpian. La examenul clinic pacienta prezenta artere temporale proeminente, macroglosie, hipotenstiune ortostatică. Forța musculară era normală. Pacienta

avea niveluri crescute ale VSH-ului şi a CRP. A fost suspectată CGA. Un vârf gamma la electroforeza proteinelor serice a adus în discuţie gamapatia monoclonală (MG). Imunelectroforeza a relevat bande monoclonale pentru IgG şi lanţuri kappa. Depozite masive de amiloid în absenţa inflamaţiei au fost găsite la biopsia arterei temporale. Mielomul multiplu şi limfomul au fost excluse. A fost pus diagnosticul de amiloidoză tip AL ce complică MGUS. Pacienta a avut răspuns clinic bun la terapia cu bortezomib, ciclofosfamidă şi dexametazonă.

Din cunoştiinţele noastre este primul caz la care MGUS cu IgG şi lanţuri kappa se prezintă ca CGA.

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