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PHOTO QUIZ

Facial numbness as a symptom of a systemic disease

Y.H.M. Poel[1*], W.A.T. Slieker[2], H.P.J. Visser[1]

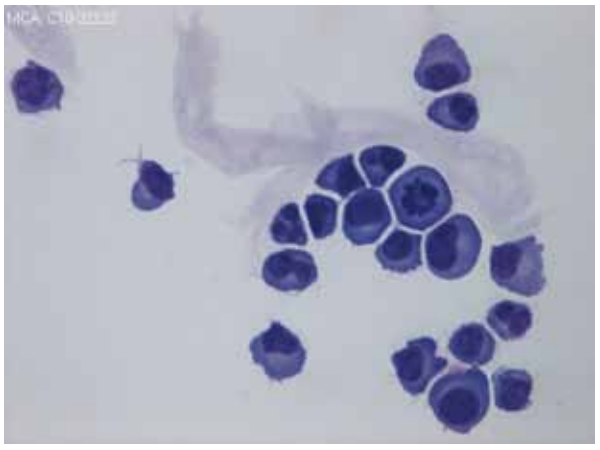
Departments of [1]Internal Medicine and [2]Clinical Chemistry, Hematology and Immunology, Medical Centre Alkmaar, the Netherlands, [*]corresponding author: tel.: +31(0)72 548 4444, fax: +31(0)72 5482165, e-mail: y.h.m.poel1@mca.nl

CASE REPORT

A 71-year-old woman was referred to the neurology department with unilateral sensibility loss of the face, which had developed over several days. Her medical history included arteritis temporalis and multiple myeloma, with an IgA-Lambda M-protein level of 49 g/l at onset, one year before presentation. The patient was treated with melphalan, thalidomide and prednisolone, and she was in complete remission five months before presentation.

Physical examination revealed a subjective sensibility loss of the face, without other abnormalities. Laboratory investigations showed no abnormalities in kidney function, liver enzymes, electrolytes or inflammatory markers. No M-protein was detected. Magnetic resonance imaging of the brain and spine revealed a mass close to the left nervus trigeminus, suggestive for a schwannoma or a meningeoma. However, the patient showed a rapid deterioration of her neurological condition, with the onset of dysphagia and a paresis of the right foot. Because of this symptomatology, analysis of the cerebrospinal fluid (CSF) was performed (*figure 1*).

Figure 1. *Cerebrospinal fluid showing abnormal cells*



WHAT IS YOUR DIAGNOSIS?

See page 405 for the answer to this photo quiz.

DIAGNOSIS

Analysis of the CSF showed atypical plasma cells suggestive for the diagnosis multiple myeloma (MM) with involvement of the central nervous system (CNS). Additional flow cytometric analysis of the CSF revealed a monoclonal plasma cell population with a strong expression of CD38 and cytoplasmic IgA-Lambda (*figure 2),* which confirmed the diagnosis mentioned above.

MM is characterised by the presence of monoclonal proliferating plasma cells, usually restricted to the bone marrow. The current treatment and prognosis of this disease have improved due to the introduction of a novel generation of drugs. These drugs are combined with more traditional chemotherapy and in younger patients possibly autologous stem cell transplantation.[1] CNS involvement of MM is a rare complication with an estimated incidence of approximately 1%.[2] It is defined by the presence of monoclonal plasma cells in the CSF. Evidence of monoclonality is mandatory, as plasma cells can be seen in several infectious and non-infectious conditions. The exact aetiology remains unknown. Several hypotheses are 1) direct continuous spread of osteolytic skull lesions; 2) haematogenous spread of plasma cells seen in plasma cell leukemia, or the spread of lymphoid cells, progenitors of plasma cells; and 3) continuous growth of plasma cells in the CNS during the course and treatment of MM, while the drugs used in MM cannot pass the blood-brain barrier.[3] The clinical presentation covers a diffuse array of neurological symptoms and signs. Treatment options include combinations of systemic chemotherapy, intrathecal chemotherapy and cranial irradiation. Autologous stem cell transplantation can be considered when the patient is in a good clinical condition. Despite treatment, CNS involvement of MM has a poor prognosis with a median survival of two months.[3,4] Our patient started intrathecal chemotherapy (cytarabin); however, the neurological symptoms worsened rapidly and she died three weeks after the diagnosis was made.

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Figure 2. *Immuno-flow cytometric analysis of CSF showing strong expression of CD38 and cytoplasmic expression of Ig light chains Lambda (CyLambda) (left figure) and IgA (CyIgA) (right figure) by the monoclonal plasma cells*