A case of sarcoidosis with elevated serum IgM

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CASE REPORT

A 65-year-old woman treated for sarcoidosis was referred to our hospital for sustained serum IgM elevation. She had been diagnosed with uveitis seven years later after the onset of blurry vision. Besides, because of what appeared to be bilateral hilar lymphadenopathy on her chest X ray, a bronchofibroscopy was performed. Bronchoalveolar lavage fluid showed increased total cell count (8.44 × 10^5/mL) (1.0–2.0), lymphocyte ratio (36.5%) (0–10), and CD4/CD8 ratio (3.51) (2.0–3.0). Transbronchial lung biopsy revealed sarcoïdal granuloma, based on which we diagnosed sarcoidosis that made the diagnosis of sarcoidosis. Since then, treatment of uveitis with corticosteroid eye drops has continued. There was no record that serum IgM levels were measured at the time of diagnosis of sarcoidosis, and this was the first measurement. Subsequently, persistence of the serum IgM elevation was confirmed at her annual follow-ups. On the day of consultation after six months, blood laboratory examination showed: white blood cell count 5500/µL, Hb 15.5 g/dL, lactate dehydrogenase (LDH) 188 IU/L, Ca 9.8 mg/dL, angiotensin-converting enzyme (ACE) 12.9 U/L (8.3–21.4), IgG 912 mg/dL (870–1700), IgA 134 mg/dL (110–410), IgM 419 mg/dL, and total protein 6.9 g/dL (albumin 61%, α1-globulin 3.3%, α2-globulin 9.0%, β1-globulin 6.4%, β2-globulin 4.2%, γ-globulin 16.1%).

As the differential diagnosis for IgM monoclonal gammopathy, the possibility of primary macroglobulinemia, IgM heavy chain disease, IgM monoclonal gammopathy of undetermined significance (MGUS), and Schnitzler syndrome were considered. Serum protein electrophoresis showed a monoclonal (M)-spike in the γ-globulin region (arrowhead in Figure 1A), and serum immunoelectrophoresis showed an additional line (M-bow) in the same region. Bone marrow aspiration showed no abnormal findings, and plasma cells in the bone marrow were less than 10%. Hence, we diagnosed this case as IgM MGUS complicated with sarcoidosis.

DISCUSSION

The diagnostic criteria of MGUS are: monoclonal protein less than 3 g/dL, plasma cells in bone marrow less
than 10%, and no evidence of other lymphoproliferative disorders [1]. The risk of progression of MGUS to multiple myeloma is approximately 1% per year [1]. Although MGUS or multiple myeloma in association with sarcoidosis is not well elucidated, 10 such cases were previously reported [2, 3]. Additionally, 33 cases with both sarcoidosis and multiple myeloma were also reported [4]. Sarcoidosis is a T-cell-mediated immunological disease following an exaggerated response to an unknown trigger. Chronic immune stimulation associated with sarcoidosis might lead to lymphoproliferative disorders [5]. As the one side of sarcoidosis, lymphoproliferative neoplasm is known to occur concomitantly with sarcoidosis [5]. One explanation for the relationship between sarcoidosis and MGUS is that the hypergammaglobulinemia induced by the chronic inflammatory response with sarcoidosis results in prolonged plasma cell lifespan [2–4]. Due to the risk of progression to multiple myeloma, a careful follow-up is required.

CONCLUSION

We report a rare case of sarcoidosis with MGUS. In this case, serum protein electrophoresis helped the diagnosis. In this case, it was suggested that sarcoidosis might be associated with lymphoproliferative disorder.

Keywords: IgM, Monoclonal gammopathy of undetermined significance (MGUS), Sarcoidosis

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Author Contributions

Yasuyuki Taooka – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.
