A Case of Primary Macroglobulinemia Initially Presented as Sensorineural Hearing Loss

Hiroki Tanaka, Yukiyoshi Hyo, Dai Shibata and Tamotsu Harada

Primary macroglobulinemia is called Waldenström’s macroglobulinemia (WM), which is defined as lymphoplasmacytic lymphoma (LPL). The excessive and abnormal production of immunoglobulin M (IgM) monoclonal macroglobulin protein in bone marrow is characteristic of WM.

Clinical signs and symptoms are presented by hyperviscosity with excessive IgM protein disordering the vessel flow with sensory and motor peripheral neuropathy derived from the myelin-associated glycoprotein (MAG) in IgM.

The clinical features in the hyperviscosity syndrome are the different kinds of mucosal hemorrhage, retinal vein thrombosis and hemorrhage, headache, vertigo, hearing loss and so on. However, sensorineural hearing loss is rarely caused by WM, and WM with hearing loss has been reported only in eight studies to the best of my knowledge. Although the etiology of the hearing loss is unclear, but the past reports have concluded that the most probable of the cause of the hearing loss is the hyperviscosity syndrome and reported that hearing loss improved with plasma exchange treatment or chemotherapy.

We experienced a case which proved to be WM after the recovery of sensorineural hearing loss with steroid therapy followed by plasma exchange and chemotherapy. The patient reached remission status for the WM and the hearing loss has not recurred.

Keywords : macroglobulinemia, sensorineural hearing loss, hyperviscosity

References

13) Vital C, Deminière C, Bourgouin B, et al.: Waldenström’s macroglobulinemia and peripheral neuropathy: deposition of

Pathological findings
a: HE stain ×100, b: HE stain ×400
Proliferation and diffuse infiltration of small lymphoid round cells and plasma cells is found in the trabecular bone.
c: immunostaining for CD138, ×400
Cells express CD138, which is a marker for plasma cells.
d: immunostaining for IgM, ×400
Specimen is diffusely positive for IgM.

Findings of FDG-PET
FDG has accumulated in lymph nodes in the neck, axilla, and mediastinum, in addition to the vertebral bones (arrows).