Here we report the first case of the development of intracranial solitary plasmacytoma in the inner ear after allogeneic stem cell transplantation (allo-SCT) in a 39-year-old Japanese female with primary plasma cell leukemia (PCL). A point to note is that this is not a case on multiple myeloma but on PCL. She was successfully treated with local irradiation and survived more than 6 years from the time of diagnosis and transplantation. This case elucidates the biology of PCL and stresses the need for an individual approach to the clinical management of patients with plasma cell neoplasm undergoing allo-SCT.

**Key words:** allogeneic hematopoietic stem cell transplantation – plasma cell leukemia – intracranial extramedullary plasmacytoma – MRI

**INTRODUCTION**

Plasma cell leukemia (PCL) is known to have a very poor prognosis with a median survival of 6–8 months (1), even after autologous or allogeneic stem cell transplantation (auto- or allo-SCT) (2). Although meningeal involvement has been reported in multiple myeloma (MM) and primary PCL (3, 4), only two cases of isolated CNS relapse have been reported after hematopoietic SCT in PCL. Neither of these cases were solitary plasmacytoma (5). Hence, this is the first reported case of intracranial solitary plasmacytoma (ICSP) development after allo-SCT in a primary PCL patient, although CNS involvement has been reported after allo-SCT in MM patients.

**CASE REPORT**

A 39-year-old female was diagnosed with PCL when she developed generalized fatigue, headache and dyspnea on exertion without a past history of an otologic disease. Her white blood cell count was $9.2 \times 10^9/L$ with 29% plasma cells. Her hemoglobin and platelet counts were 6.2 g/dL and $126 \times 10^9/L$, respectively. Serum IgG was 12300 mg/dL with a monoclonal peak of IgG-κ, and β2-microglobulin was elevated at 19.2 μg/mL. Bence Jones protein of the κ type was also present in the urine. Bone marrow aspirate revealed 74.2% plasma cells with a normal karyotype. Renal function was normal and there were no bone lesions. She was diagnosed with PCL and treated with two cycles of VAD (vincristine 0.4 mg/day continuous i.v. days 1–4, doxorubicin 15 mg/day continuous i.v. days 1–4, and dexamethasone 40 mg/day days 1–4, 9–12, and 17–20) following myeloablative conditioning which consisted of melphalan 110 mg/m$^2$ intravenously on day −4 and total body radiation 12 Gy (six fractions) on days −3 to −1. She then received allogeneic peripheral blood stem cells containing $6.7 \times 10^9/kg$ of CD34-positive cells from her HLA-identical brother. GVHD prophylaxis was short-course methotrexate and ciclosporin. While she did not experience acute graft-versus-host disease (GVHD), she later had mild extensive-type chronic GVHD. One year after transplantation, she developed a recurrent left-sided sensory neural hearing loss (SNHL) that responded to steroid therapy, and a left vestibular system impairment. However, no abnormal findings...
were observed on Gadolinium-enhanced magnetic resonance imaging (Gd-MRI). Twenty-eight months after transplantation, she developed a left-sided facial nerve palsy and recovered completely after two months. Gd-MRI performed one month later revealed an enhanced lesion less than 6 mm in diameter in her internal auditory canal and inner ear (Fig. 1). The patient refused biopsy at that time. Nine months later, she developed left facial nerve palsy and left severe otalgia, which did not respond to steroids. Three months later, she developed vertigo and vomiting, and Gd-MRI detected a tumor which protruded from the internal auditory canal and pressed into the brainstem (Fig. 2). At this time, she had mild chronic GVHD without immunosuppressive treatment.

A biopsy was performed which revealed plasmacytoma. Immunohistochemical study revealed monoclonal protein of the IgG-κ type as well as primary PCL (Fig. 3). Laboratory findings, including bone marrow examination, were all within normal limits and neither monoclonal gammopathy nor Bence Jones protein were observed. There were no abnormal findings in the bone. On the basis of these findings, we diagnosed the patient with intracranial solitary plasmacytoma. Focal brain radiation therapy (2 Gy/day, for a total of 15 days) was performed followed by short-term administration of dexamethasone. She is currently well without recurrence 41 months after the last treatment.

DISCUSSION

Review of the literature revealed that the present case is one of the longest survivors of PCL (1,2). The present case also showed a very unusual relapse pattern involving the internal auditory canal and the inner ear of PCL not MM.

Extramedullary tumors may occur in patients with leukemia after allo-SCT as a manifestation of relapse (6). One of the possible explanations for occurrence and extramedullary localization is the relative inefficacy of immune surveillance, or the graft-versus-leukemia effect, outside the bone marrow cavity (7). The mechanism of solitary plasmacytoma occurrence in our case might be similar to that of leukemia and may be associated with the graft-versus-plasma cell effect (8). This effect and the use of steroid therapy for neuro-otological symptoms may also explain the slow progression of plasmacytoma in this case.

The relationship between PCL and plasmacytoma generally remains unclear. According to reported patterns of MM relapse after SCT, PCL occurred in six (2%) out of 280 and extramedullary plasmacytoma occurred in 40 patients (14%) with relapse (9), suggesting that leukemia and mass formation are interchangeable in plasma cell neoplasms. Our case is an example of this observation.

In the present case, the subtlety of the internal auditory canal and the inner ear lesion on Gd-MRI made the diagnosis difficult. Internal auditory canal lesions smaller than 6 mm on Gd-MRI, as in our case, are typically benign, even if the patient has neuro-otological symptoms (10). In order to detect drastic changes in the lesion, additional and periodic Gd-MRIs must be performed whenever new neurological symptoms are observed.

Conflict of interest statement

None declared.

References


