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Case Report

A Case of Refractory Multiple Myeloma with Hypoglossal Nerve Involvement

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Abstract:

When the case was re-examined after the literature scan, it was seen that there was a prominent paranasal sinus wall thickening on the right side that developed during the period when the patient's complaints began, without any mass lesion or leptomeningeal involvement. It was observed that the patient had regressed in the imaging after treatment.

Key words: radiotherapy; chemotherapy; cells; bone

Introduction

Multiple myeloma (MM) is a clonal stem cell disease originating from plasma cells. The development of MM neurological findings is mostly caused by hyperviscosity, hypercalcemia, amyloidosis, vertebral bone involvement and spinal cord compression due to fractures or nerve compression, neuropathy due to paraproteinemia. Brain involvement is very rare. It may present as cerebral lesion, parenchymal disease or leptomeningeal involvement. A case of isolated hypoglossal nerve involvement under MM treatment at the age of 52 will be presented.

Case

A 46-year-old male patient presented with complaints of pain in the left shoulder and chest that started on January 0, 2019, in addition to weight loss, weakness in the legs, and difficulty in walking. In the examinations performed, wbc: 8.24 10e3/uL hbg: 9.23 gr/dl, plt: 118 10e3/uL, pnl: 5.65 10e3/uL, urea: 117 mg/dl, creatinine: 3.85 mg/dl, albumin: 2.79 gr/dl, globulin: 3.81 gr/dl, corrected calcium: 9.16 mg/dl, beta2 microglobulin: 0.44 mg/dl, serum free lambda light chain: 1190 mg/l increased and serum free kappa light chain: 5.25 mg/l, 24-hour urine immunofixation electrophoresis revealed free lambda light chain: 5.65 mg/l, Chain band was detected. In the MR imaging, there were nodular lesions in the iliac bone and sacrum, immunoglobulin values were IgA: 25 mg/dl, IgG: 293 mg/dl, IgM: 68 mg/dl, 80% plasma cells in the bone marrow. The patient was diagnosed with multiple myeloma and started on bortezomib, cyclophosphamide, dexamethasone (VCD) chemotherapy. After 4 cycles of VCD and radiotherapy (RT), an increase in light chains was observed in the control evaluation, and a bortezomib, lenalidomide and dexamethasone (VRD) course was started. The patient, who underwent autologous BMT in December 2019, was followed up under lenalidomide cordexa maintenance treatment, and a relapse was detected in the control evaluations in June 2022. Ixazomib, lenalidomide and dexamethasone (IRD) treatment was started, and daratumumab VCD treatment was switched to due to lack of response. The patient, who was followed up under daratumumab VCD treatment, developed complaints of decreased hearing and numbness in the jaw. In addition to the atrophy in the left half of the tongue and the complaint of shifting to the left when the tongue was taken out of the mouth, speech and swallowing were impaired. Neurology consultation and detailed brain imaging showed increased thickness in the right maxillary sinus. In the PET-CT imaging, although there was no pathological involvement in the head, neck and mediastinal structures, widespread lytic lesions were seen especially in various vertebrae, femur and tibia, and it was evaluated as progressive disease. No plasma cells or other pathology was detected in intrathecal sampling. Peripheral smears were examined daily with suspicion of plasma cell leukemia, but plasma cells were not detected. The patient was evaluated in the council with neurology and it was evaluated that there was isolated myeloma involvement of nervus hypoglossus and the treatment was arranged as dartumumab, pomalidomide and dexamethasone. Brain RT was performed. After 2 cycles of chemotherapy and radiotherapy, the patient's tongue numbness, speech and swallowing disorders improved. The complaint of left shift when the tongue came out of the mouth regressed. The patient's isolated nervus hypoglossus involvement improved with treatment, and his follow-ups are continuing.

Conclusion

Brain involvement may very rarely develop in 1% of patients with multiple myeloma [1]. Parenchymal involvement may be in the form of

mass compression or leptomeningeal involvement due to plasmacytomas [2]. The most common cranial nerve involvements in the literature are the oculomotor nerve (CN 3), abducens nerve (CN 6) and hypoglossal nerve (CN 12) [3]. Involvement of these cranial nerves is most commonly due to plasmacytomas originating from the skull base and sinuses [4]. When other cases of multiple myeloma with hypoglossal nerve involvement were scanned in the literature, it was seen that there was plasmacytoma or leptomeningeal involvement [5,6]. In our case, it is a very rare condition in terms of the absence of a mass lesion, plasmacytoma and leptomeningeal involvement in the brain. When the case was re-examined after the literature scan, it was seen that there was a prominent paranasal sinus wall thickening on the right side that developed during the period when the patient's complaints began, without any mass lesion or leptomeningeal involvement. It was observed that the patient had regressed in the imaging after treatment. This finding could not be associated with hypoglossal nerve involvement. However, in a case report, a patient with a soft tissue mass in the right paranasal sinus had significant plasmacytomas at the skull base, and clinically, there was oculomotor, facial and hypoglossus nerve involvement [7]. This situation suggests that involvement in the paranasal region may require differentiation from classical infection conditions in patients at risk and close monitoring in terms of intracranial events. Plasmacytomas of the skull base occur as an extension of plasmacytoma originating from the clivus, petrous part of the temporal bone or from the submucosa of the sinonasal and nasopharyngeal (extramedullary plasmacytoma) region. Extramedullary plasmacytomas are most commonly seen in the nasal and paranasal sinuses, nasopharynx, tonsils and larynx [8]. Due to the rarity of extramedullary plasmacytomas, data on treatment and prognosis are limited. However, studies have shown the effectiveness of radiotherapy. There are publications showing that 30-50 gy radiation most effectively reduces tumor sizes caused by multiple myeloma [9]. In our case, symptoms were evaluated acutely and it was thought that starting RT and appropriate chemotherapy would prevent possible involvement of other cranial nerves and the development of leptomeningeal spread. In cases of relapsed refractory disease, cranial nerve involvement can be seen very rarely, and radiotherapy and chemotherapy constitute treatment options.

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