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Article

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A rare case of cervical epidural extramedullary plasmacytoma presenting with monoparesis

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Abstract: Multiple myeloma and other plasma cell disorders are characterized by production of a large number of plasma cells in the bone marrow. On the other hand, plasmacytoma results from proliferation of abnormal plasma cells in the soft tissue or skeletal system. Neurological complications are frequently observed in these diseases. The most commonly known complications among those complications are spine fractures, spinal cord compressions, and peripheral neuropathies. Although neurological involvements are common in plasmacytomas, extramedullary spinal epidural localizations have been reported very rarely. In this case report, we aimed to present a plasmacytoma case that presented with acute onset of upper extremity monoparesis. A 40-year-old woman was admitted to our clinic with complaints of sudden weakness and numbness in her left arm following neck and left arm pain. Emergency cervical magnetic resonance imaging (MRI) revealed an epidural mass and the patient underwent emergency surgery. The patient showed improvement post-operatively and the pathology was reported as plasmacytoma. Following hematology consultation, systemic chemotherapy was initiated and radiotherapy was planned after wound healing.

Key words: Cervical epidural, extramedullary, plasmacytoma

Introduction

Plasmacytoma, a member of plasma cell dyscrasias and a rare disease among monoclonal gammopathies, describes situations in which multiple myeloma disease is seen outside the bone marrow. Multiple myeloma is malignant and uncontrolled proliferation of plasma cells. The incidence of multiple myeloma is approximately 4-8/100,000 (1). Plasmacytoma is frequently observed in solitary bone tissue and

extramedullary tissue. While the incidence of solitary plasmacytoma in all plasma dyscrasias is 5%, the incidence of extramedullary plasmacytoma is 3% (1, 2, 4). The extramedullary spinal epidural localization of plasmacytomas is very rare and only 6 cases have been reported in the literature.

In this case report, we aimed to present a rare case of plasmacytoma who was evaluated in the emergency room with diagnosis of spinal epidural mass and paresis and underwent emergency surgery.

Case Report

A 40-year-old female patient was admitted to our emergency department with complaints of neck pain, weakness and numbness of the left upper extremity that last for 10 days. From the patient's history, it was learned that her complaints were present for the last 3 months and that the numbness in the left hand worsened and weakness developed during the last 5 days. The patient's neurological examination revealed painful neck movements in all directions, limited flexion and extension and 3/5 muscle strength in the distal and proximal of right upper extremity. Deep tendon reflexes were found to be increased in left upper extremity. Hoffman test was positive on the left side. Sensory examination revealed hypoesthesia conforming to the C3-C4-C5 dermatomes in the left upper extremity. Both non-contrast and contrast enhanced emergency cervical MRI scans were performed with the preliminary diagnoses of cervical disc herniation or spinal mass. Cervical MRI revealed a 5x4x5 cm, multilobulated, multicompartmental, well-demarcated soft tissue mass that fills the central spinal canal with expanding anterior epidural space at C3-C4-C5 levels, extends into prevertebral and parapharyngeal space through left neural foramina and rests against the posterior border of sternocleidomastoid muscle (SCM). The mass was hypointense on T1 and hyperintense on T2-weighted images and homogeneously enhanced on post-contrast examinations and it was evaluated as a tumor of peripheral nerve origin (Figure 1 A, B, C, D). The patient underwent emergency surgery with the diagnosis of schwannoma. Preoperative blood count blood test, sedimentation rate and C-reactive protein levels were normal. In the prone position, C3-C4-C5 laminectomy was performed with posterior approach. Following laminectomy, an off-white mass consistent with tumor that was compressing the dura posteriorly and laterally was seen. Laminectomy boundaries were expanded laterally with moving the operating microscope laterally. The soft tumoral mass was dissected from the dura and biopsies were taken. The tumoral mass was found to extend laterally and to anterior part of the dura. The surgery was completed after deciding achieving enough decompression with partial excision. The patient did not have any additional neurological deficit post-operatively and improvement was observed in the neurological deficit in the distal left arm. Pathology was reported as plasma cell tumor (Figure 2). The patient was referred to hematology, systemic chemotherapy was initiated and radiotherapy was planned after wound healing.

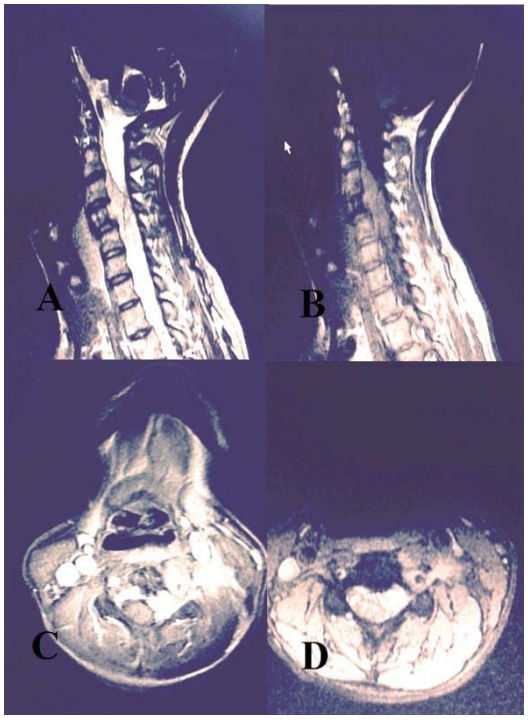

Figure 1 A, B,C,D - Mass lesion, compressing the spinal cord by filling the posterior C3-C4-C5 epidural space, that is hyperintense on sagittal T2-weighted MRI, hypointense on sagittal T1-weighted MRI

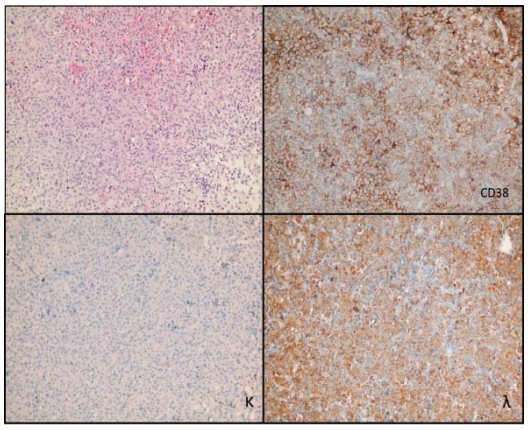

Figure 2 - Histopathological examination of the specimen showing plasma tumor cells with atypical nuclei (400x, H & E)

Discussion

Uncontrolled proliferation of B-lymphocytes and plasma cells, which are involved in antibody production, may result in development of plasma cell dyscrasias and plasmacytoma. Diseases observed in plasma cell dyscrasias are divided into three groups. These are multiple myeloma, solitary plasmacytoma and extramedullary plasmacytoma, in descending order. There are proliferating malignant plasma cells in the bone marrow and diffuse lytic bone lesions in multiple myeloma. The incidence of plasmacytoma, one of the plasma cell dyscrasias, is 3%. The prognosis of plasmacytoma, in which solitary bone and extramedullary tissue involvement is also seen, is better than multiple myeloma. Multiple myeloma usually accompanies extramedullary plasmacytoma and close follow-up is necessary for cases without accompanying multiple myeloma. A transformation rate of 32-70% to multiple myeloma in the following years has been reported in the literature (1,2,4).

Diagnosing plasmacytoma patients with preoperative MRI scans and clinical findings is extremely difficult. MRI reveals spinal epidural metastasis or spinal mass image. Patients can be admitted with cord compression and myelopathy due to multiple myeloma. Spinal cord compression can occur due to vertebral body compression fractures, spinal epidural tumor extension or extension of multiple myeloma in another tissue to epidural space (3,6,7). Compression fractures can be seen in 55-70% of patients with

multiple myeloma with or without spinal cord compression. The incidence of paresis related to plasma cell dyscrasias is 5-10%. Spinal column is one of the regions that involvement is seen in plasmacytomas. Extramedullary plasmacytomas usually involve submucosal areas and necks of various organs. Extramedullary plasmacytoma localized only in the epidural space is very rare (8).

Review of the literature revealed only 14 cases of cervicothoracic localization (cervical localization with 6 case and thoracic localization with 8 case). All these cases were observed to have myelopathy due to compression. The first case was reported by Sod and Wiener in 1957. It was a patient with metastatic spinal epidural tumor at thoracic 5 level and who presented with paraparesis (10).

The first case of cervical region localized plasmacytoma was presented by Kim et al. A 69 year-old female patient presented with paraparesis and was operated for a detected lesion extending from C6 to T10. In 1992, another 52 years old patient, who was presented with paraparesis and was found to have a lesion extending from C7 to T4, was reported (5,9). The last patient with cervical epidural plasmacytoma was reported in 2011. The 45-year-old patient was admitted with progressive paresis and an epidural mass extending from C4 to C7 was determined on MRI (8). While the youngest patient was 40 years old, the oldest patient was 85 years old. However, all the patients except one were accompanied by multiple myeloma. Patients with plasmacytoma should be consulted to hematology department and multiple myeloma screening should be performed. It should be kept in mind that multiple myeloma develops in 70% of such cases during follow-up (8).

The optimal treatment of extramedullary plasmacytoma cases is not clear. Radiation therapy should be added to treatment, as they are highly radiosensitive tumors. There are some tumor cases in which local control was achieved with only radiotherapy. Besides, chemotherapy may also extend survival and recurrence time, thus preventing the transformation to multiple myeloma. The treatment is surgical excision and postoperative radiotheraphy - chemotheraphy in patients with cord compression (8).

Conclusion

We presented a 40-year-old patient who was admitted with neck pain and left upper limb paresis and who was diagnosed having a very rare case of plasmacytoma with epidural metastases. Extramedullary plasmacytomas must be kept in mind in differential diagnosis of masses with spinal epidural metastasis and especially extension to neck.

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