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Case Report

# Cervical intradural recurrence of multiple myeloma during hematological remission: A case report and literature review

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## 1. Introduction

The average overall survival of patients with Multiple Myeloma (MM) was around three years in the US in the early twentieth century. Driven by access to better medication, median overall survival in younger patients (aged < 50 years) was exceeding ten years by 2014. At the time of diagnosis, extramedullary MM is found in approximately 7% of patients, while another 6% may develop extramedullary lesions during the course of their disease. However, the central nervous system (CNS) is a scarce location of extramedullary involvement and is diagnosed in less than 1% of MM patients, especially younger patients who are more prone to develop lesions in the CNS with a median age of 53 years. Our patient was 57 years old when initially diagnosed with MM, and had CNS involvement ten years after the diagnosis.

## 2. Case report

A 67-year-old woman with a past medical history of stage III Multiple Myeloma (Intraosseous, Kappa subtype) diagnosed in May 2009 and treated with chemotherapy and autologous stem cell transplant (ASCT), had a complete hematological remission in April 2016.

In April 2019, the patient presented with a ten weeks history of lower neck pain with a recent onset of left C7 radiculopathy. She had no signs or symptoms of myelopathy. Radiographs of the cervical spine were normal. However, Magnetic Resonance Imaging (MRI) confirmed a contrast-enhancing, intradural, extramedullary $13 \times 11 \times 10$ mm mass lesion at the level of the C7 vertebral body, compressing the spinal cord and the left C7 nerve root posteriorly (Fig. 1).

Microsurgical resection of the mass lesion was performed under general anesthesia, the patient placed in a prone position using a Mayfield head holder. A midline approach was performed, and the posterior elements of the C6 and C7 vertebrae were exposed. A high-speed burr was used to drill through between the junction of the lamina and the lateral mass. The laminae were removed en bloc with a Leksell Rongeur, and the spinal cord was exposed. Opening the dura showed an intradural and extramedullary mass with an identifiable cleavage caudal and cephalad of the tumor without attachment to the dura. The tumor was removed off the spinal cord and the exiting nerve root, after excision of the denticulate ligament and meticulous dissection between the lesion and the spinal cord. Using microdissection, the entire tumor was removed and sent for histopathological analysis. Sheets of neoplastic plasma cells with few mitoses, Russell bodies, and plasmablasts diagnostic of a plasma cell malignancy were observed. Immunohistochemical studies confirmed the diagnosis of the MM of kappa subtype (Fig. 2A).

The patient was discharged three days after surgery with no neurological deficit and was able to ambulate independently without support. Post-operative MRI showed complete resection of the lesion (Fig. 2B). A complete workup performed before surgery including the brain, thoraco-lumbar MRI and a PET-Scan showed only three small suspicious lesions in the vertebral bodies of L3, L4, and L5 vertebrae, with serum Immunoglobulin G levels staying within a normal range (680 mg/dL). The 6 months clinical follow-up showed that the patient is neurologically intact, and had started systemic chemotherapy.

## 3. Discussion

The average overall survival in patients with multiple MM was

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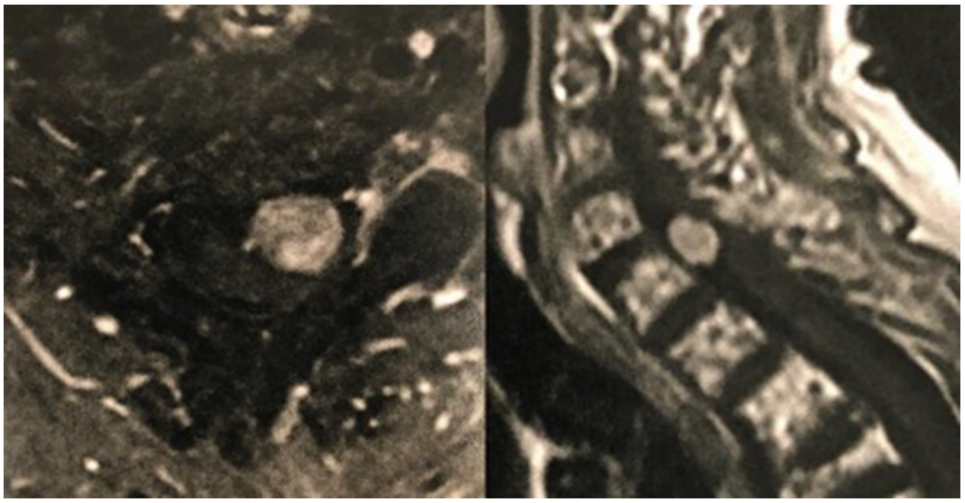

**Fig. 1.** Pre-operative axial and sagittal gadolinium enhanced T1 weighted magnetic resonance imaging showing the intradural mass lesion at the level of the C7 vertebra.

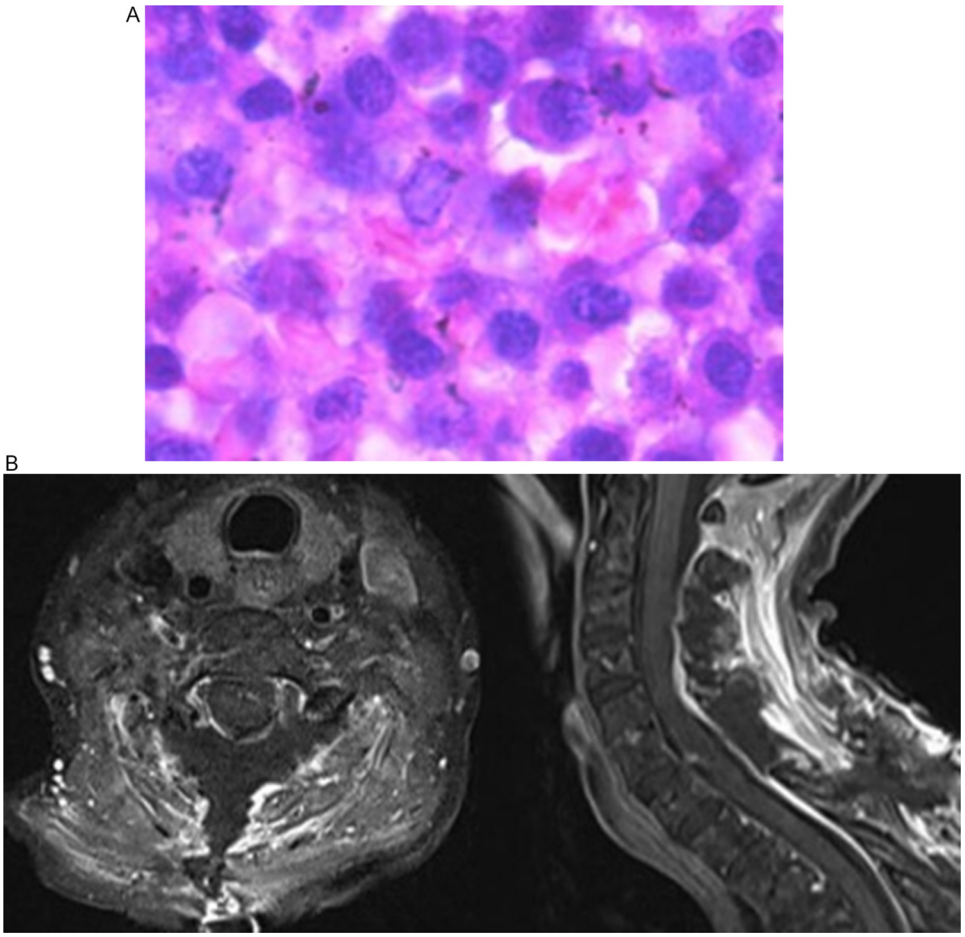


**Fig. 2.** (**A**) Histological appearance of MM (hematoxylin and eosin) showing sheets of plasma cells with eccentric nuclei and cartwheel chromatin. (**B**) Post-operative axial and sagittal gadolinium enhanced T1 weighted magnetic resonance imaging showing complete resection of the mass lesion.

around three years in the US at the beginning of the twentieth century. Driven by access to better medication, median overall survival in younger patients (aged < 50 years) was exceeding ten years by 2014 [4]. At the time of diagnosis, extramedullary MM is found in approximately 7% of patients, while another 6% may develop extramedullary lesions during the course of their disease. However, the central nervous system (CNS) is a very rare location of extramedullary involvement and is diagnosed in less than 1% of MM patients, especially younger patients who are more prone to develop lesions in the CNS with a median age of 53 years [3]. Our patient was 57 years old when initially diagnosed with MM, and had CNS involvement ten years after the diagnosis, and three years after complete hematological remission. Gross total resection was achieved in this case.

The differential diagnosis for intradural extramedullary spinal

tumors includes spinal meningioma, nerve sheath tumors such as schwannoma or neurofibroma, and metastasis. Schwannomas are the most frequent intradural tumors, often found in patients aged between 30 and 40 years, and typically present on MRI as iso-T1, hyper-T2, moderately enhanced lesions. Meningioma is the second most common intradural tumor, more often found in older patients. Leptomeningeal metastases of solid tumors include breast, lung, melanoma, gastrointestinal, genitourinary, and head and neck cancers [1]. Although clinical and imaging characteristics of a metastatic tumor depend on their origins, they usually appear in people aged between 50 and 70 years, with MRI showing enhanced lesions isointense on T1 and hyperintense on T2.

There are only five cases of intradural MM, including one case of intramedullary involvement [2]. No clear guidelines have emerged in the management of intradural MM because of the small number of cases published in literature with various treatment options. Complete surgical resection of the lesion was performed in only one reported case of solitary plasmacytoma. Other management alternatives include chemotherapy (systemic and intrathecal) and radiation therapy. A retrospective study including 17 patients with CNS myeloma showed significant improvement in overall survival of patients treated with intrathecal chemotherapy (IT) with an overall survival of 20 months compared to 2 months in patients who did not receive IT [5]. In our case, complete microsurgical resection will be followed by a systemic pharmacotherapy associating Daratimumab, Pomalidomide and zoledronic acid, and IT was judged unnecessary at this point since the CSF workup was negative at the time of the lesion diagnosis.

CNS involvement in MM is a negative prognostic factor. Paludo et al. found that Overall survival (OS) from the diagnosis of MM was significantly shorter in the CNS-MM group (median 40 months) than in a control group (median, 93 months; 95% CI, 67–129 months). OS was 3.4 months from the detection of CNS disease.

## 4. Conclusion

Intradural, extramedullary MM is an infrequent entity and should be included in the differential diagnosis of intradural mass lesions. Radiological findings are not characteristic, and the diagnosis is only confirmed by immunohistochemical analysis. Although there is no consensus on the optimal management strategy for these lesions, complete surgical resection, when possible, should be considered the treatment of choice, followed by adjuvant therapy including systemic and intrathecal chemotherapy and radiation therapy. The prognosis of patients with myelomatous involvement of the central nervous system remains poor despite the advances in the management of Multiple Myeloma and the novel agents improving overall survival.

Patient consent was taken before the inscription of this case report.

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