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Case Report

Intramedullary spinal cord germinoma clinically mimicking multiple sclerosis: A case report

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ABSTRACT

Background: It is important to differentiate intramedullary neoplastic lesions from nonneoplastic diseases such as multiple sclerosis (MS) and other demyelinating or inflammatory diseases.

Case Description: A 26-year-old Japanese male presented with a history of intracranial germinomas and obstructive hydrocephalus, treated with endoscopic surgery, and adjuvant chemotherapy and radiation therapy. Three years later, he developed paresthesias involving the right hand and both lower extremities. The cervical MR scan demonstrated a heterogeneously enhancing intramedullary C1-C2 lesion with surrounding edema. On cytological examination of the cerebrospinal fluid (CSF), there were no neoplastic cells. However, the fluid was positive for oligoclonal immunoglobulin G (IgG) bands. The patient received steroid pulse therapy to address the potential MS diagnosis. The follow-up MR showed reduced edema, but no change in the size of the intramedullary lesion. Therefore, the patient underwent a cervical laminectomy for tumor resection. The pathology was consistent with the same cranial germinoma treated 3 years previously. He subsequently received whole spinal radiation and three courses of chemotherapy.

Conclusion: Some spinal cord tumors may produce oligoclonal IgG bands in CSF. In this case, an intramedullary C1-C2 spinal cord germinoma was originally misdiagnosed as MS due to the presence of oligoclonal IgG bands in CSF. Differentiating this tumor from MS and initiating appropriate treatment were critical into the care of this patient.

Keywords: Germinoma, Multiple sclerosis, Oligoclonal band immunoglobulin G, Spinal cord tumor

INTRODUCTION

It is important to differentiate intramedullary neoplastic lesions from nonneoplastic diseases such as multiple sclerosis (MS) and other demyelinating or inflammatory diseases. Here, we report a drop metastasis from a cranial germinoma, resulting in an intramedullary C1-C2 cervical tumor documented on an enhanced MR. It was notably difficult in distinguishing this intramedullary metastatic germinoma from a potential MS lesion as the cerebrospinal fluid (CSF) was positive for oligoclonal immunoglobulin G (IgG) bands.

CASE DESCRIPTION

Original presentation

A 26-year-old Japanese male presented with headaches, anorexia, and diplopia. The enhanced computed tomography scan showed two small intracranial masses; one was a suprasellar lesion

and the other appeared at the aperture of the aqueduct, resulting in obstructive hydrocephalus. No lesions were found in the spinal cord. An endoscopic biopsy was performed of the suprasellar mass, and the accompanying third ventriculostomy resolved the hydrocephalus. The pathology revealed a germinoma and he received three courses of chemotherapy (carboplatin, 450 mg for 1 day; etoposide, 1100 mg for 5 days). This was followed by whole-brain radiation (24 Gy). Ultimately, the intracranial lesions disappeared.

New intramedullary lesion 3 years later

Three years later, however, the patient experienced vacillating paresthesia in his right hand and both legs, but without a focal neurological deficit. Human chorionic gonadotropin β-subunit (βhCG) and α-fetoprotein (AFP) were within normal limits in the serum (βhCG <0.1 ng/ml and AFP 2.2 ng/ml), CSF βhCG was 0.4 ng/ml, and AFP was 0.2 ng/ml. The cytological examination of CSF was negative. However, oligoclonal IgG bands were positive in CSF (IgG index, 0.66; myelin basic protein, 45.8 pg/ml).

Radiological diagnostic evaluation

The cervical MR revealed a heterogeneously enhancing, expansile intramedullary cord lesion at the C1-C2 level, accompanied by marked edema extending from the medulla oblongata to the C4 level [Figure 1]. There were no accompanying intramedullary or extramedullary lesions in the thoracic or lumbar spinal studies.

Differential diagnosis and treatment

The main differential diagnoses included; astrocytoma, ependymoma, or germinoma along with other nonneoplastic diseases (e.g., MS, other demyelinating diseases, or inflammatory myelitis). Due to the potential diagnosis of MS, the patient received steroid pulse therapy with methylprednisolone (1 g/day) for 3 days. The more likely diagnosis of a tumor was later confirmed when the follow-up magnetic resonance imaging (MRI) showed reduced edema around the unchanged contrast-enhancing C1-C2 intramedullary mass [Figure 1].

Surgery

One month later, the patient underwent a C1 laminectomy/C2 partial laminectomy with revised laminoplasty of the C2 spinous process for resection of the intramedullary cervical lesion. A myelotomy was performed along the posterior median sulcus; just under the cord surface, the tumor was grayish, soft, and nonhemorrhagic and appeared to grow into the central canal. As the intraoperative frozen section diagnosis was consistent with germinoma, a sufficient biopsy/decompression was performed without the need for total resection.

Pathology

The tumor consisted of a mixed population of large cells (e.g., large round nuclei, hyperchromasias, with clear cytoplasm) and mature lymphocytes [two-cell pattern, Figure 2]. There were no syncytiotrophoblastic giant cells. Immunohistochemical studies positively stained for placental alkaline phosphatase and C-kit [Figure 2]. Staining for βhCG and AFP was negative, while the Ki-67 labeling index was approximately 30%. These findings confirmed the diagnosis of a germinoma that was unchanged compared to his prior intracranial surgery. Most likely, the cervical tumor resulted from dissemination of germinoma cells through the central canal. The patient recovered from surgery without any sequelae. The postoperative MRI revealed partial resection of the tumor, and he was treated with three courses of chemotherapy (e.g., carboplatin and etoposide) and begun on holospinal radiation (30 Gy).

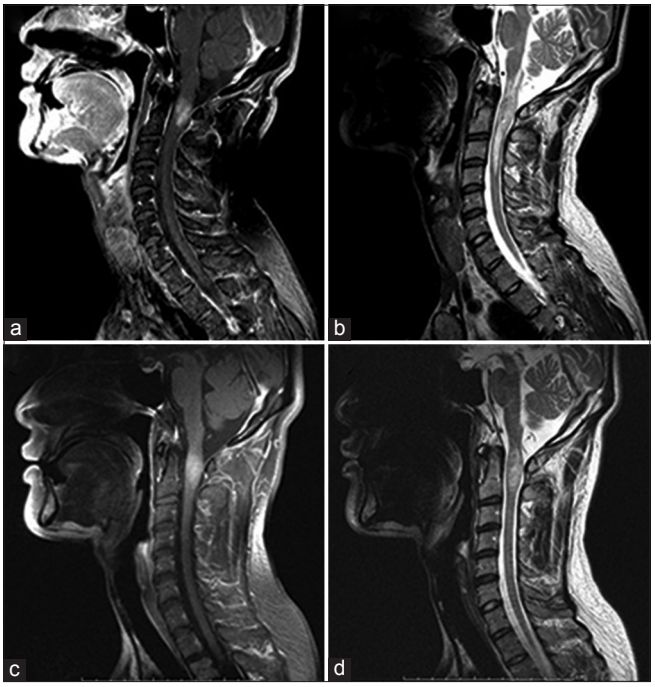

Figure 1: (a) Sagittal T1-weighted postgadolinium magnetic resonance (MR) images through the cervical spine showing intense contrast enhancement of an intramedullary lesion from the C1 to C2 level. (b) Sagittal T2-weighted MR images demonstrating the heterogeneous intramedullary lesion extending from the medulla oblongata to the C4 level, which was thought to represent spinal cord edema surrounding the enhanced mass. (c and d) Scans after steroid pulse therapy. (c) Sagittal T1-weighted postgadolinium MR images showing no change in the enhanced lesion. (d) Sagittal T2-weighted MR images showing a decrease in cord edema.

Table 1: Cases of germinomas that showed oligoclonal IgG bands before surgery.

Series	Age (year)/sex	Lesion	First diagnosis
Krolak-Salmon *et al.*[5]	35/M	Right optic nerve	Optic neuritis
Krolak-Salmon *et al.*[5]	?	Pineal lesion	?
Krolak-Salmon *et al.*[5]	?	Midbrain	?
Birnbaum *et al.*[2]	34/F	Pineal lesion, hypothalamus, medulla oblongata	Multiple sclerosis
Akiyama *et al.*[1]	13/M	Bilateral thalamus	Demyelinating disease
Present case	29/M	Cervical spine (C1-C2)	Multiple sclerosis susp.

IgG: Immunoglobulin G

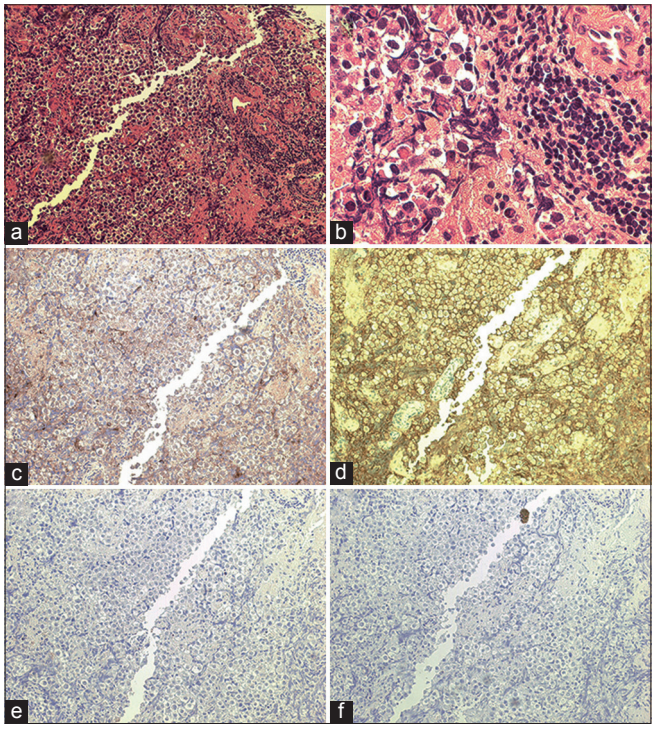

Figure 2: (a) Photomicrographs of the operative specimen at low power (original, ×100) displaying a mixed population of large atypical cells and small lymphocytes (two-cell pattern). Large atypical cells had proliferated with a solid to sheet-like pattern. The small lymphocytes had invaded the stroma. There were no syncytiotrophoblastic giant cells. (b) High-power micrograph of the tumor (original, ×400) displaying large round cells characterized by nuclear atypia with hyperchromatic large ovale and a clear cytoplasm. Invading lymphocytes were matured. (c-f) Immunohistochemical stains of the tumor. The large atypical cells were diffusely positive for placental alkaline phosphatase and C-kit (original, ×100). (c) Placental alkaline phosphatase. (d) C-kit. (e) α-fetoprotein. (f) Human chorionic gonadotropin.

DISCUSSION

Differential diagnosis tumor versus MS

For intramedullary spinal cord lesions, it is important to differentiate tumors from nonneoplastic diseases (e.g., MS, demyelinating disease, acute and subacute myelitis [viral, bacterial, tuberculosis, fungus, and parasites], atopic myelitis, and sarcoidosis). Here, we were initially unable to differentiate demyelinating diseases from a tumor. As the patient demonstrated oligoclonal IgG bands and his clinical symptoms were suggestive for MS versus other demyelinating diseases, steroid pulse therapy was initially performed before surgery.[7]

Frequency of oligoclonal bands and tumor versus MS/other

Several case reports show that 5.8% of neoplastic lesions of the central nervous system (CNS) may produce oligoclonal IgG bands as do 9.3% of patients with paraneoplastic syndromes.[8] The production of IgG may be attributed to tumor of the CNS (e.g., lymphomas or carcinomatous myelopathy), spinal arteriovenous malformations (AVMs), cervical spine disease, and miscellaneous factors.[3,6] For AVMs, recurrent bleeding into the CNS can damage the blood-brain barrier, leading to the immune system synthesizing IgG.[3] Space-occupying lesions, which destroy the surrounding normal tissue and blood vessels/blood–brain barrier, can further lead to release of CNS antigens.[3] Including the present case, there are six reported cases of germinoma that showed oligoclonal IgG bands [Table 1].

Differential diagnosis of intramedullary spinal cord germinoma versus MS with IgG antibodies

Intramedullary spinal cord germinoma is rare and has been reported in just 16 primary cases.[4] If CSF is positive for oligoclonal IgG bands, an intramedullary spinal cord germinoma can be initially misdiagnosed as demyelinating disease/MS. Thus, germinomas should be considered among the differential diagnoses for cases of intramedullary spinal cord lesions with oligoclonal IgG bands, particularly when MS-based steroid pulse therapy is ineffective.

CONCLUSION

Here, we report an intramedullary cervical spine germinoma that represented a drop lesion from an original intracranial tumor. Notably, the tumor produced oligoclonal IgG bands in the CSF making it difficult to differentiate between metastatic germinoma and MS.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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