Intramedullary Cervical Spine Germinoma: A Case Report

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SUMMARY

Primary intramedullary germinoma arising in the cervical spinal cord is a very rare entity. We present one such case arising in a young man who presented with radiculopathy and myelopathy, which was partially excised. Upon histological confirmation, he was treated successfully with radiotherapy alone. To our knowledge, this is only the second reported case worldwide which is histologically confirmed. Although extremely rare, differential diagnosis for intramedullary spinal cord tumor should include germinoma.

KEY WORDS:

Germ cell tumor, germinoma, intramedullary tumor, spinal cord tumor

INTRODUCTION

Germ cell tumors of the central nervous system are rare, and it usually arises intracranially. The pineal and suprasellar region are the most common site, constituting approximately 3% to 5% of all primary brain tumors in children and young adults [1]. However 5% are found to arise from other midline structures such as the third ventricle and thalamus. Germ cell tumors arising from the spinal cord is very rare, and case reports have been few and far between. Only two cases of cervical spine germinoma has been reported previously. Thus, we report a case of a primary intramedullary seminomatous germ cell tumor arising in the cervical spine of a young adult.

CLINICAL PRESENTATION

A 22-year-old Malay man presented to us with a 5-month history of neck ache, which was gradually progressing in severity. This was accompanied by progressive limb numbness, starting from the left lower limb with subsequent involvement of both upper limbs. He also has progressive difficulty in grasping an object with his left hand and difficulty in ambulating, whereby upon presentation to us he could only mobilize with the use of a wheelchair. He also had urge incontinence 4 weeks before presentation. He sought treatment from a private neurosurgeon, and magnetic resonance imaging of the spinal cord revealed a contrast-enhancing spinal cord mass extending from C3 to T1 level. He was then referred to the Neurosurgery Department, Sultanah Aminah Hospital, Johor Bahru, Johor for further management.

Physical examination revealed left hemiparesis (grade 3/5) with wasting of the small muscles of the hand. There was also decreased sensation from the level of C6 below, more prominent on the left side. There was hyper-reflexia of the left upper and lower limbs with clonus. No testicular mass was palpable and the rest of the clinical examination was unremarkable. The lesion on clinical examination was localised to be intramedullary.

IMAGING

Magnetic resonance imaging of the spinal cord revealed a hyperintense intramedullary lesion extending from the C3C4 junction down to the T1 level (Figure 1). The lesion was mainly solid and caused cord expansion. There was homogenous enhancement of the lesion upon administration of gadolinium. On the basis of these findings, the differential diagnosis was an ependymoma or astrocytoma.

INTERVENTION

The patient was started on high dose intravenous dexamethasone and an emergency spinal cord decompression was planned. He underwent a C3 to T1 laminoplasty and partial resection of tumor. The cervical portion of the spinal cord exposed upon opening the dura, was swollen. Posterior midline myelotomy of the spinal cord revealed a grey, soft tumor without a clear capsule. There was no identifiable clear margin between the tumor and the spinal cord. A frozen section biopsy revealed sheets of large, round tumor cells separated by lymphocytic infiltrates. Abundant nucleoli and numerous mitosis suggested a highly proliferative nature. In view of these findings and the inability to delineate a clear border between the tumor with normal spinal cord tissue, only partial removal of the tumor was done.

On histopathological examination, the tumor consisted of sheets and clusters of tumor cells intermingled with fibroconnective tissue stroma associated with dense lymphocytic infiltrates. The cells were round to polygonal with clear to eosinophilic cytoplasm and moderately pleomorphic nuclei. Occasional prominent nucleoli and mitosis was seen. There was no sarcomatous elements and no obvious choriocarcinomatous or other germ cell tumor component. Immunohistochemistry showed the cells to be positive for placental alkaline phosphatase stain with equivocal β-HCG staining. All other staining were negative. A diagnosis of intramedullary germinoma was made.

The patient recovered well but his neurological condition did not improve. Sensory impairment still persisted, as well as hemiparesis. Chemical analysis obtained early after surgery showed a normal level of serum β-HCG and α-fetoprotein level. A post-operative magnetic resonance scan confirmed

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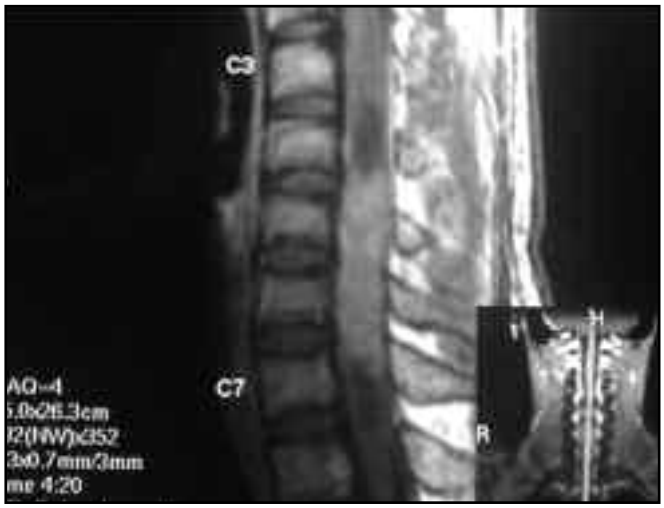

Fig. 1: A sagittal T1-weighted magnetic resonance imaging scan of the cervical spine after gadolinium administration demonstrating the homogenous hyperintensity of the tumor.

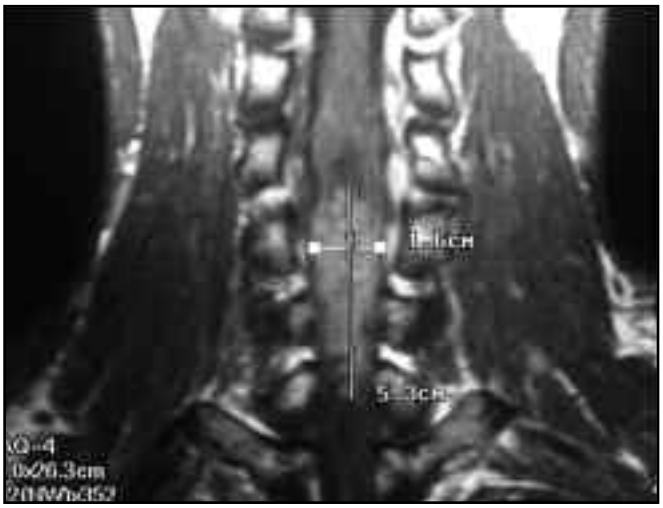

Fig. 2: A coronal T1-weighted magnetic resonance imaging scan of the cervical spine showing the size of the intramedulary tumor.

partial resection of the tumor. Further diagnostic imaging of the brain, mediastinum, testes and the rest of the body were negative for any tumor. With the histopathological confirmation and radiological evidence of tumor residual, radiation therapy was started. Local irradiation of the spinal cord was commenced, from C1 to T3 level (including two vertebrae above and below the tumor site), to a total of 36 Gy in 20 fractions for a total duration of 4 weeks. Neither prophylactic craniospinal irradiation nor total spinal irradiation was undertaken due to the absence of obvious craniospinal dissemination on imaging. Magnetic resonance imaging of the spine was done 3 months after radiation therapy and showed that the tumor has completely disappeared. Although the patients overall clinical status improved vastly, his left-sided hemiparesis with hyperreflexia and clonus still persists.

DISCUSSION

Extragonadal germ cell tumor usually occurs at midline structures of the body such as the mediastinum or retroperitoneum. In the central nervous system, the pineal region is the most common site of origin, followed by the suprasellar compartment. These tumors usually spread to the spinal cord first if they metastasize. The spinal cord is an extremely rare primary site of origin for germinoma. Recent literature review [2] have found 26 reported cases of primary intramedullary spinal cord germinoma, mostly as a single case report. Of the 27 cases, including our case, 16 of them were from Japan.

Out of all the reported cases of primary intramedullary spinal cord germinoma, only 2 cases shows involvement of the cervical spine. One of the cases was a presumed germinoma based on the cytological analysis of the cerebrospinal fluid and complete eradication of the tumor with radiotherapy. No pathological diagnosis was ever made. The other more recent case involved the C3 to C6 spinal level, surgically treated with partial resection with additional chemotherapy and radiotherapy [3]. Pathological examination confirmed the diagnosis of germinoma. The present case, to our knowledge, is the only other case of primary intramedullary spinal cord germinoma of the cervical spine with confirmed pathological diagnosis.

Treatment of primary intramedullary spinal cord germinoma should follow that of primary intracranial germ cell tumor. This includes surgery for tissue diagnosis followed by radiation therapy with/without chemotherapy. In our case, intraoperative frozen section show findings which are consistent with germinoma, therefore total resection was not attempted. Thereafter he was subjected to local radiation treatment to the tumor bed. Craniospinal radiation and chemotherapy was not commenced.

Radiation therapy has been the primary curative treatment for intracranial germinoma, achieving disease control rate of more than 90% [4,5]. Several major series also incorporated the use of low-dose craniospinal irradiation followed by a local-field booster dose to the primary tumor of approximately 50 Gy [4,5]. However, the patient may develop significant late effects from radiotherapy on their cognitive and neuroendocrine function, even in older patients. Although radiation therapy proved effective, there is currently no standing consensus regarding the need for radiation to the craniospinal axis or the radiation dose to the spinal tumor bed in primary intramedullary spinal cord germinoma. This is also true for the efficacy of chemotherapy use in this group of patient. Intracranial germinoma is highly sensitive to chemotherapy, especially platinum and alkylating agents. As such, chemotherapy followed by lower dose radiation therapy has been proposed as it may reduce the endocrine and neurocognitive side effects of craniospinal radiation. As yet, the role of adjuvant chemotherapy with radiation in spinal cord germinoma remains to be conclusively proven.

Primary intramedullary spinal cord germinoma is extremely rare but should be considered as a differential in primary spinal cord tumors. Radiotherapy and chemotherapy appears to be highly effective, however further study needs to be done to determine the efficacy of this treatment and its possible long term outcome.

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