Multisegmental Diffuse Intradural Extramedullary Ependymoma

An Extremely Rare Case

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Key words: intramedullary tumor, intradural extramedullary lesion, ependymoma

SUMMARY – *Ependymoma has been described typically as an intramedullary tumour derived from ependymal cells with a predominance in women in the fifth decade of life. Pain is the most frequent symptom. Intradural extramedullary presentation is rarely described and almost always as a unique lesion. We describe a 53-year-old man with multi-segmental diffuse intradural extramedullary ependymoma with progressive lower leg hypoesthesia with regular motility. The patient's neurologic condition improved after surgery.*

Introduction

Ependymomas are neuro-epithelial tumours of the central nervous system originating from ependymal cells lining the ventricles, choroid plexus, central canal of spinal cord, and the filum terminalis. They account for only 4% to 6% of primary central nervous system tumours, but about one third arise within the spinal canal where they represent the most common intramedullary neoplasm in the adult population. Although, ependymomas are unencapsulated glial neoplasms, the vast majority of spinal ependymomas exist in a histologically benign form with little infiltrative potential exhibiting a slow biological growth evidenced by a prolonged average symptom duration preceding diagnosis, preservation of neurological function in patients with spinal cord compression and characteristically long survival times following various forms of treatment. These features of spinal ependymomas, coupled with their relatively low incidence, hamper the evaluation of treatment¹⁻³.

Spinal cord tumours can be classified as extradural (55%) or intradural (45%), the lat-

ter being either intramedullary (5%) or extramedullary (40%). Neurofibromas, neuromas, and meningiomas account for roughly 60% of intradural extramedullary tumours and affect mainly adults, whereas intramedullary tumours are more common in children. Ependymomas and astrocytomas are the main histological diagnoses in children. Ependymomas are the most common intramedullary tumours and occur predominantly in adults. The reported cranial/spinal tumour ratio for ependymomas is 4:1, ranging from 3:1 to 20:1, depending on histological subtype. Intradural extramedullary ependymomas are extremely rare except for those located at the terminal filum or conus medullaris. An extensive literature survey yielded 18 such cases². Before this, there was just only one literature report of intradural extramedullary ependymoma (with the exception of those occurring at the conus medullaris and terminal filum) with multiple lesions at presentation.

We describe a 53-year-old man with multisegmental diffuse intradural extramedullary ependymoma and discuss the incidence, MRI pattern and treatment of this rare lesion.

Figures 1 and 2 A 53-year-old man with a ten-day history of progressive lower legs hypoesthesia with regular motility. The sagittal and axial T2W MRI cervical-thoraco-lumbar spine showed multisegmental diffuse intradural extramedullary hyperintense lesions predominantly isointense on T1W and on T2W and FLAIR images. The lesions were located anteriorly or posteriorly in the spinal canal at lower cervical, upper or lower thoracic level respectively, compressing and displacing the spinal cord contralaterally.

Figure 3 Sagittal T1W fat sat MRI with contrast injection showed moderate enhancement of the multisegmental intradural extramedullary lesions.

Case Report

History and Examination

A 53-year-old man presented with a ten-day history of progressive lower legs hypoesthesia with regular motility. The neurological examination showed negative Wasserman and Lasegue signs with preserved muscle strength.

In October 2010, a spinal MRI with contrast injection was performed (protocol sagittal and axial T1W T2W, and T1W fat sat) showing multisegmental diffuse intradural extramedullary lesions at cervical and thoracic level that were predominantly isointense on T1W and hyperintense on T2W and FLAIR images, with moderate contrast enhancement on T1W-fat sat (Figures 1-3).

Figure 4 Sagittal and axial T2W MRI control after the first surgery with decompressive laminectomy at lower thoracic level. The biopsy diagnosed a multisegmental diffuse intradural extramedullary ependymoma.

The lesions were located anteriorly or posteriorly in the spinal canal respectively at lower cervical, upper or lower thoracic level, compressing and displacing the spinal cord contralaterally.

Cranial MRI did not reveal any abnormality nor did the total body MDCT with contrast undertaken to disclose other primary lesions. The differential diagnosis was between lymphoma, metastasis, leptomeningeal diffusion by unknown cancer, and ependymoma. An open-surgery biopsy with decompressive laminectomy was performed to characterize the lesion.

Surgery

After few days later, a laminectomy was performed at lower thoracic level as the site of

major spinal cord compression. A brownish intradural extramedullary tumour compressing the spinal cord posteriorly was immediately apparent after opening the dura. Under microscopic assistance, no attachment of the tumour to the dura or spinal cord was observed, but arachnoid infiltration was noted. The tumour was soft. After internal debulking of the lesion, a sharp dissection at the tumour–spinal cord interface was performed, and the tumour was resected. Because of the multifocal characteristic of the lesion and arachnoidal infiltration, the aim of surgery was spinal cord decompression instead of total resection (Figure 4).

One month later another laminectomy with tumour resection was performed at upper thoracic and lower cervical level (Figure 5).

Figure 5 Sagittal T2W MRI control after the second surgery with decompressive laminectomy showed spinal cord decompression.

Histological examination revealed a proliferation of ependymal cells with eosinophilic cytoplasm and hyperchromatic nuclei and papillary structures, the presence of a pseudo-rosette with many mitoses, vascular proliferation and a significant proliferation index (Ki 67), a WHO Grade II ependymoma (Figure 6).

Postoperative Course

The patient's neurologic condition improved after surgery, and he was able to stand on his own and walk without assistance.

The patient received adjuvant chemotherapy consisting of four cycles of carboplatin (175 mg/m²/week for four weeks), followed by a two-week break.

Discussion

Intradural extramedullary ependymomas are extremely rare and predominate in women in the fifth decade of life². Our literature survey yielded 18 cases of ependymoma at the same location; the first of which was reported by Cooper in 1951⁴. Only one was described as multifocal at presentation^{2,4,6}.

Pain is the most frequent initial symptom^{1,3,6}. The origin of intradural extramedullary ependymoma has been attributed to the presence of heterotopic glial cells, since the earliest report by Cooper et al.⁴.

Our case is an atypical, rare presentation of ependymoma in terms of clinical onset, sex distribution and lesion location and distribution.

Figure 6 Histological examination revealed a proliferation of ependymal cells with eosinophilic cytoplasm and hyperchromatic nuclei and papillary structures, the presence of a pseudo-rosette with many mitoses, vascular proliferation and significant proliferation index (Ki 67), a WHO Grade II ependymoma.

It represents the second case of intradural extramedullary ependymoma (except those occurring in the conus medullaris and terminal filum) with multiple lesions described in the literature⁴.

Generally, this lesion is more frequent in women and symptoms are neck or back pain. Of the 19 patients described, 15 were women and four were men. A higher incidence among women was observed in earlier studies. In a review paper of intradural extramedullary ependymomas, Duffau et al. postulated that a hormonal factor was involved^{2,8-9}.

The initial symptoms were pain in 13 patients and medullary syndrome (with motor and/or sensory and/or sphincter control deficit) in 12 (data were not available for one patient). Dorsal pain and radicular pain are the most frequently reported clinical symptoms, regardless of compartment (intra or extradural) or histological subtype. Intradural extramedullary ependymomas tend to follow the same pattern, with pain being reported as the earliest symptom in the majority of cases described (13 of 19 cases)².

Our case concerns a middle-aged man without pain but affected by progressive lower leg hypoesthesia with preserved motility. Typically, ependymomas are mostly at cervical level and are generally intramedullary. The thoracic location is the most common site for extramedullary ependymomas, unlike intramedullary ependymomas^{2,4}. In the literature, a thoracic location for these lesions was found only in 11 cases, while intradural extramedullary thoracic-lumbar ependymomas have been described in women. Although a hormonal influence on the histogenesis of these lesions has been postulated no hormonal assessment data has been published to date^{2,4}.

Multisegmental diffuse presentation at diagnosis had been previously described in one literature report². In our case, the lesion was disseminated from lower cervical to thoracic and lumbar levels. The prognosis seems to be related to the extent of resection and the presence of meningeal infiltration. Clinical improvement occurred in 11 patients².

Magnetic resonance imaging is the gold standard for diagnosing spinal tumours. MRI of the entire neuraxis is essential to rule out intracranial ependymomas with metastasis to the neuraxis. Ependymomas usually enhance uniformly after contrast administration. Tumour location was distributed as follows: cervical in two patients, cervico-thoracic in three, thoracic in 11, lumbar in two, and multiple in two patients².

Intradural extramedullary ependymoma is often histopathologically benign, and patients enjoy a good neurologic recovery after tumour resection. However, malignant transformation of this neoplasm can occur, leading to cerebrospinal fluid dissemination and recurrence⁵.

The major differential diagnosis for intramedullary ependymoma is spinal cord astrocytoma. Ependymomas often haemorrhage and are typically more sharply delineated than astrocytoma, but there is no specific pattern that permits reliable differentiation between these two common intramedullary tumours and a surgical biopsy is recommended².

Conclusion

We describe the second case of multisegmental diffuse extramedullary ependymomas. This etiology must be taken into account in the differential diagnosis of intradural extramedullary tumours, and a complete imaging study

of the entire neuroaxis should always be performed to exclude tumour dissemination. In multisegmental presentation, asymptomatic

lesions of this histopathological variant may be observed at appropriate time intervals due to no or slow growth.

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