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A huge ependymoma of the cervical spinal cord with subtle atypical manifestations and hyperhidrosis: Case report

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ABSTRACT

INTRODUCTION: Ependymomas are the most common neuroepithelial tumors of the spinal cord, accounting for 50–60% of spinal cord gliomas. The nonspecific clinical presentation of a spinal cord tumor frequently results in delay of diagnosis with opposing outcomes.

PRESENTATION OF CASE: We report a 34-year-old man presented with abnormally enhanced sweating on the left side of his neck, upper extremity, and chest that had been occurring for 1 year. In the sagittal MRI there were a centrally localized mass lesion extending from medulla and C1 to T2 vertebra level and expanding the cord. Surgical elimination of the tumor was performed with posterior midline approach and near total resection of tumor was achieved.

CONCLUSION: Cervical intramedullary ependymal is a rare, slow growing spinal cord tumor. Attention to uncommon characteristics like hyperhidrosis might be an important key to early diagnosis of this rare spinal tumor. Surgical resection is the choice of treatment with infrequent recurrence.

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1. Introduction

Ependymomas are the most common neuroepithelial tumors of the spinal cord, accounting for 50–60% of spinal cord gliomas [1]. In adults, ependymomas characterize the most common intramedullary spinal tumors. Almost two thirds of spinal cord ependymomas comprise cervical levels [2].

Liabile on location, the spinal cord tumors present with diverse symptomology, counting neck or back pain and motor symptoms antedated by sensory symptoms [3]. The nonspecific clinical appearance of a spinal cord tumor often results in delay of diagnosis with opposing outcomes. We present a case of a 34-year-old male with a cervical spinal ependymoma. The objective of this case report is to prove uncommon characteristics of a rare complaint in a patient that may present to spine clinics or other health care specialists.

2. Presentation of case

This study has been approved by the institutional review board of the Mazandaran University of Medical Science in Iran.

This was a 34-year-old cabdriver man presented with abnormally boosted sweating on the left side of his neck, upper extremity, and chest that had been happening for 1-year. On extra

questioning, the patient described he protested of chronic recurrent cervical pain from 8 years ago and after 4 years he had subtle clinical signs of onset of pain along the left side of the neck radiating to the upper lateral aspect of left arm. Recently, he was referred to our department because MR images discovered an intramedullary tumor of the cervical spinal cord. His medical and family histories were ordinary.

We did physical and neurological examinations and found hyperhidrosis in nearly the right C5–T6 dermatomes. The patient had isocoric pupils, and both direct and indirect light reflexes were normal. He had mild motor faintness of the left upper extremity and slightly diminished sensation in the left lower extremity and also criticized of pain in the left thigh.

In the sagittal MR images there were a centrally localized mass lesion spreading from medulla and C1 to T2 vertebra level and expanding the cord. The lesion was iso to hypointense in T1 weighted images and hyperintense in T2 weighted images. The lesion showed heterogeneous appearance with multiple cavities and cystic lesions. These images finding suggested an intramedullary lesion. Extending of lesion in upper cervical cord and medulla was significant (Fig. 1).

Surgical removal of the tumor was achieved with the patient placed prone with a Mayfield skull lock after induction of general anesthesia. We performed midline incision between occiput to T2 vertebra level, total laminectomy of C1 to T1 performed, and the facets conserved.

We opened Dura in the midline under the operative microscope and a standard midline myelotomy performed through the

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Fig. 1. Sagittal T2 weighted MRI without contrast showed heterogeneous high signal lesion in upper cervical and cervicothoracic cord.

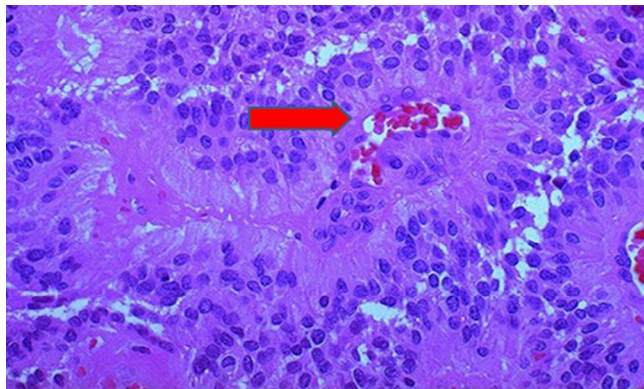


Fig. 2. Photomicrograph of endependymoma showing pseudorosettes around the central blood vessels, Hematoxylin and eosin X 400.

posterior median septum. Sensory motor evoked potentials were used to guide to diminish the neurological deficit intraoperatively. Between C1–T1 levels a soft, brown colored, no cleavable mass lesion was seen. We resected the tumor up to 90%. Homeostasis was skillful and the Dura sutured continuously.

Macroscopically, tumor was soft, brown colored constituents. Microscopically circumscribe of tumor was well and it organized in sheets of spindle and epithelioid cells with round to oval nuclei containing small nucleoli. Some cells had clear cytoplasm. Perivascular pseudorosettes which consist of nuclear free zones around central blood vessels were recorded. Hyper cellularity and frequent mitoses were not seen (Fig. 2).

The pathological diagnosis was well-matched with WHO, grade II endependymoma [4].

Postoperatively, the patient showed worsened motor weakness of the right and left extremity (I/V and II/V, respectively) and decreased sensation in the left lower extremity. The patient could

walk on the second day. After the two day, arm weakness gradually improved and he discharged and referred to a physical therapist on the 4-day. Hyperhidrosis gradually improved. With rehabilitation the patient could walk without support. Then, he was undertaken a course of cervical field external beam radiotherapy at a dose of 45–54 Gy for adjuvant therapy.

Three, six and twelve months MRI with contrast showed no obvious mass lesion (Fig. 3). There was no tumor recurrence during the 1-year follow-up period.

3. Discussion

Classical spinal ependymomas mainly disturb middle-aged adults with an equal sex distribution. Although the cervical cord represents only 22.5% of spinal cord tissue, about 68% of tumors arise from or spread into the cervical cord [4].

Intra medullary spinal cord tumors can exist with a diversity of symptoms. Neck or back pain is often the earliest symptom [5]. Sensory symptoms frequently ante date the motor symptoms and are consistent with the central location of the lesion within the spinal cord. Hyperhidrosis is a rare symptom of intramedullary spinal cord tumors; we have seen only 3 cases of hyperhidrosis caused by this type of tumor in the literature. The histopathological analysis in these cases was astrocytoma in two [6] and gangliocytoma in one of them [7]. It seems that our cases maybe was the first ependymomal spinal cord tumor causes hyperhidrosis.

Involvement of the descending autonomic pathways, which are positioned between the corticospinal and spinothalamic tracts, may reason both sympathetic and parasympathetic disturbances below the level of the lesion. Hyperhidrosis has been described in spinal cord injured patients [8] and also in post-traumatic syringomyelia [8]. In our patient it is likely that there was involvement of the sympathetic fibers in the upper cervical cord or in the ciliospinal center of Budge at the C8–T2 segmental level. Hyperhidrosis in this instance may be assumed to have occurred as a



Fig. 3. Sagittal T2 weighted MRI without contrast 12-month after surgery just shows linear high signal intensity in the upper cervical cord.

result of over activity of the sympathetic fibers due to irritation by the tumor. It may be hypothesized that had no treatment been done, the hyperhidrosis may have gradually progressed to anhydroses.

4. Conclusion

Cervical intramedullary ependymoma is a rare, slow growing spinal cord tumor. Attention to uncommon characteristics like hyperhidrosis might be an important key to early diagnosis of this rare spinal tumor. Early detection of the tumor is essential for optimal postoperative functional status.

Conflict of interest

None declared.

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None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

All step of article from design to writing made by Kaveh Haddadi.

Guarantor

None.

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