

Atypical meningioma with destruction of cervical vertebrae inside the spinal canal: A case report and literature review

LIKUN MU, MINGXIN WANG, LIFENG CHENG, GUANGXU CHU and ZUYAO SONG

Department of Neurosurgery, Shengli Oilfield Center Hospital, Dongying, Shandong 257099, P.R. China

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Abstract. Atypical meningioma is a specific type of meningioma that is relatively rare in clinical practice. The clinical manifestations of atypical meningioma are non-specific and the imaging findings are similar to those for other intraspinal space-occupying lesions. It is usually misdiagnosed as schwannoma or metastases, and it is necessary to rely on intraoperative findings and pathological examination to confirm the diagnosis. Atypical meningioma invading the intervertebral foramen and causing bone destruction is rare. The present study describes the case of a patient with cervical intraspinal atypical meningioma combined with cervical bone destruction. A 50-year-old female patient presented with numbness of the extremities with unstable walking, and underwent magnetic resonance imaging (MRI) of the cervical spine. MRI showed a space-occupying lesion in the spinal canal at the C2-4 level. The patient underwent cervical intraspinal lesion resection, and postoperative pathology confirmed atypical meningioma (World Health Organization grade II). The patient received dehydration and hormone therapy, and was discharged after the symptoms improved. Radiotherapy was performed 2 months after discharge. Cervical MRI 3 months after the operation showed that the tumor resection was satisfactory. To the best of our knowledge, the present study is the first to report on this specific type of meningioma. The present case report may provide useful information for clinicians to diagnose and treat the disease.

Introduction

Meningioma is a benign spinal cord tumor that arises from cap cells of the arachnoid membrane or fibroblasts of the dura mater. Globally, meningiomas account for 10-30% of intraspinal tumors (1), and 85% of meningiomas are located in the

extramedullary subdural space (2). Atypical meningioma is rarely reported in the literature; its clinical manifestations are non-specific, and the imaging findings are similar to other intraspinal space-occupying lesions. Notably, atypical meningioma is usually misdiagnosed as schwannoma or metastases, and intraoperative findings and pathological examination are required to confirm the diagnosis. The tumor grows slowly and often presents with symptoms of chronic progressive spinal cord compression, resulting in limb motor, sensory and reflex disturbances below the level of compression. Meningiomas show low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and high signal intensity on enhanced images (3). Atypical meningioma with bone destruction in the cervical spinal canal is even rarer, and only a small number of relevant reports have been reported (4,5). At present, surgical resection is the main treatment for atypical meningiomas, and postoperative adjuvant therapy is mainly radiotherapy. The present case report describes the clinical data of a patient with cervical intraspinal atypical meningioma admitted to the Department of Neurosurgery, Shengli Oilfield Central Hospital (Dongying, China), with the aim of improving the understanding of this disease.

Case report

A 50-year-old female patient was admitted to Shengli Oilfield Central Hospital in June 2023, due to numbness of four extremities, unstable walking for 3 years, and aggravation of symptoms for 6 months. A total of 3 years prior, the patient had numbness in both hands without obvious inducement and did not receive specific treatment. A total of 6 months prior, the symptoms of the patient were gradually aggravated, and the patient had numbness of all four extremities, unstable walking, the sensation of walking on cotton wool, inflexible fine movements of both hands, a girdle sensation on the chest and back, self-perceived neck pain and discomfort, but no vertigo, palpitations, tinnitus or diplopia, and no radiating pain in both upper limbs. A total of 6 months prior to admission, the aforementioned symptoms were markedly aggravated and the patient had fallen due to unstable walking. The patient therefore visited a local hospital (Dongying People's Hospital, Dongying, China) for oral methylcobalamin (0.5 mg/time, tid) drug treatment with poor results, and was later admitted to Shengli Oilfield Central Hospital for further treatment. The patient had

Correspondence to: Professor Mingxin Wang, Department of Neurosurgery, Shengli Oilfield Center Hospital, 31 Jinan Road, Dongying, Shandong 257099, P.R. China
E-mail: sjwmx@126.com

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a history of hypertension for 3 years and was regularly treated with oral benazepril (10 mg/time, qd) every day with good blood pressure control. A neurological examination showed the following: Straightening of physiological curvature of the cervical vertebrae, no obvious limitation of cervical motion, no obvious tenderness or percussion pain in the spinous process and paravertebral region of the cervical vertebrae, cervical hyperflexion, and hyperextension test (+); hypoesthesia of distal skin in the shoulder joints of the bilateral upper limbs, normal sensation in the remaining limbs; muscle strength of the bilateral upper limbs, grade 4, muscle strength of the lower limbs, grade 5; hypertonia of four extremities; bilateral biceps reflex (+ + +), triceps reflex (+ + +), radial membrane reflex (+ +), bilateral knee tendon-reflex (+ + + +), Achilles tendon reflex (+ + +); bilateral ankle clonus (+), patellar clonus (-); bilateral Hoffman sign (+), bilateral Babinski sign (+). The diagnosis on admission was: i) Cervical spondylotic myelopathy; ii) hypertension. However, the patient's blood pressure was well controlled with medication and their blood pressure on admission was 111/72 mmHg.

The day after admission, a cervical spine CT showed a mixed density lesion in the spinal canal and spinal stenosis at the posterior edge of the cervical 3-4 pyramid (Fig. 1). A total of 3 days after admission, head and neck CT angiography showed no significant abnormalities (Fig. 2); on the same day, cervical spine enhanced magnetic resonance imaging (MRI) showed a space-occupying lesion in the spinal canal at the C2-4 level (Fig. 3). A total of 11 days after admission, cervical intraspinal tumor resection was performed under general anesthesia, during which the posterior arch of the atlas, C2-C4 spinous process and lamina were exposed, and the bilateral laminae of C2-C4 were grooved with a burr to completely remove the lamina including the C2-C4 spinous process. The lesion broke through the dura mater and grew into the right C3-C4 intervertebral foramen, and the base of the tumor was located in the dura mater, which was firm in consistency, calcified, poorly circumscribed and rich in blood supply. A piecemeal resection of the tumor was performed, the boundary of the lesion on the surface of the spinal cord was carefully separated, part of the C3-C4 facet was abraded, the intervertebral foramen was explored, and the intraforaminal lesion was subtotally removed. Finally, the patient underwent fixation and reduction surgery for the C2-C4 lamina. Postoperative pathology showed cervical spinal meningioma with local hot spot mitotic figures of ~3/mm, morphology consistent with atypical meningioma (World Health Organization grade II) (6), accompanied by bone tissue invasion, and no neoplastic necrosis or spinal cord invasion (Fig. 4). Immunohistochemical results (Fig. 5) were as follows: Broad spectrum cytokeratin (focal weak data not shown), epithelial membrane antigen (Fig. 5C), glial fibrillary acidic protein (data not shown), progesterone receptor (Fig. 5B), vimentin (data not shown), H3K27me3 (data not shown), STAT6 (data not shown), somatostatin receptor type 2 (Fig. 5A), S-100 (data not shown) and Ki-67 (~2%, data not shown). Immunohistochemistry was performed according to a previously described protocol (7). A total of 12 days after admission, cervical spine CT showed satisfactory tumor resection (Fig. 6). The

patient was treated with mannitol (125 ml/time, tid), methylprednisolone (500 mg/time, qd) and methylcobalamin (0.5 mg/time, tid) after surgery. Eventually, the condition of the patient improved and they were discharged.

In early August 2023, the patient was admitted to the oncology department (Shengli Oilfield Hospital) for radiotherapy due to postoperative atypical spinal meningioma of cervical vertebra. The target volume included C2-C4 and the radiation dose was 40 Gy. This patient underwent a total of 20 radiation treatments. A total of 4 days later, cervical spine enhanced MRI showed that the lesion was markedly smaller than before (Fig. 7). Repeated cervical spine MRI in mid-October showed no marked tumor recurrence (Fig. 8).

Discussion

Meningioma is the second most common benign tumor after schwannoma in the extramedullary subdural space, which is specifically common in women aged 40-70 years (8). In addition, meningioma is the most common tumor in the thoracic spinal canal. The mean age of onset is 45 years, indicating that endocrine hormones have an effect on the formation of meningioma (8). The common early manifestations of the disease include: Numbness of the extremities, fatigue, chest and back pain, and difficulty in walking. Later, there may be hypertonia, active or hyperactive key reflexes, paraplegia, and fecal and urinary dysfunction. In the present case report, the patient was a 50-year-old woman who developed a cervical spine meningioma. Previous studies have reported that 85% of meningioma cases are intradural, 7% extend epidurally and 8% are completely epidural (5,9). The present case was intradural with cervical bone destruction. By referring to the relevant literature, only one report of foraminal invasion and bone erosion was found. The patient was a 39-year-old woman who was found to have T3/4 and T5/6 meningiomas by MRI, with symptoms including progressive numbness and weakness of both lower limbs, and gait instability. The meningiomas were shown to be invading the left and right foramina of T3/4, and the tumors were found to have calcification and bone erosion during surgery (5).

The exact pathogenesis of meningioma is unknown. Some scholars hypothesize that tumors arise from arachnoid cells extending along peripheral nerves, and these ectopic or isolated arachnoid tissues initially accumulate in arachnoid villi, especially around the peripheral nerve root sleeves, where the spinal leptomeninges directly drain into the dura mater (10). Some scholars believe that the dura mater around nerve roots may contain residual superficial embryonic arachnoid villi, which may explain epidural and some periradicular meningiomas; others believe that arachnoid islands may migrate into the epidural space, such as orbital meningiomas without associated optic nerve sheaths (11-13). The present case of meningioma breakthrough into the intervertebral foramen can be explained by the fact that the tumor may arise from arachnoid cells around the spinal nerve roots (10,11), but such patients are extremely rare in clinical practice.



Figure 1. Cervical spine CT the day after initial admission.

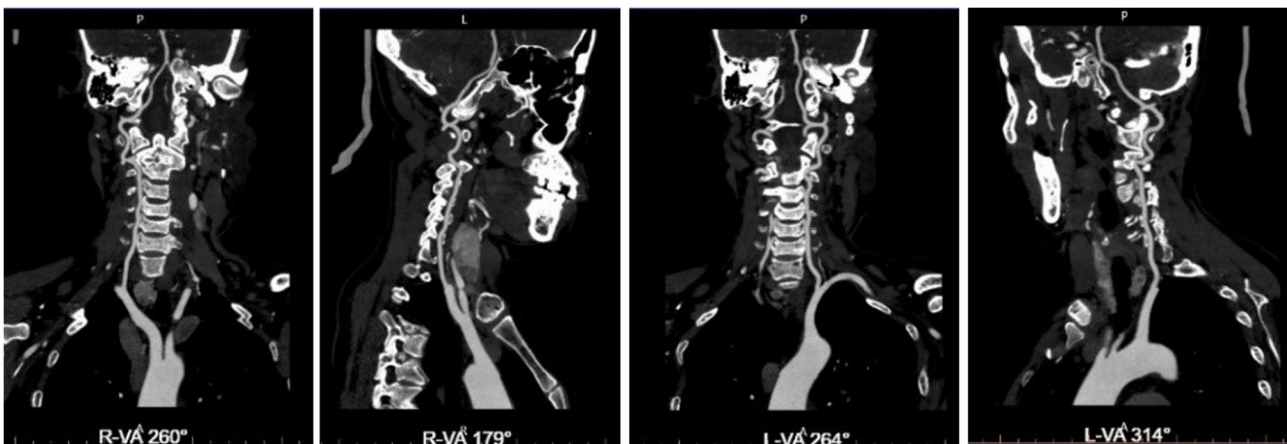


Figure 2. Head and neck CT angiography 3 days after initial admission.

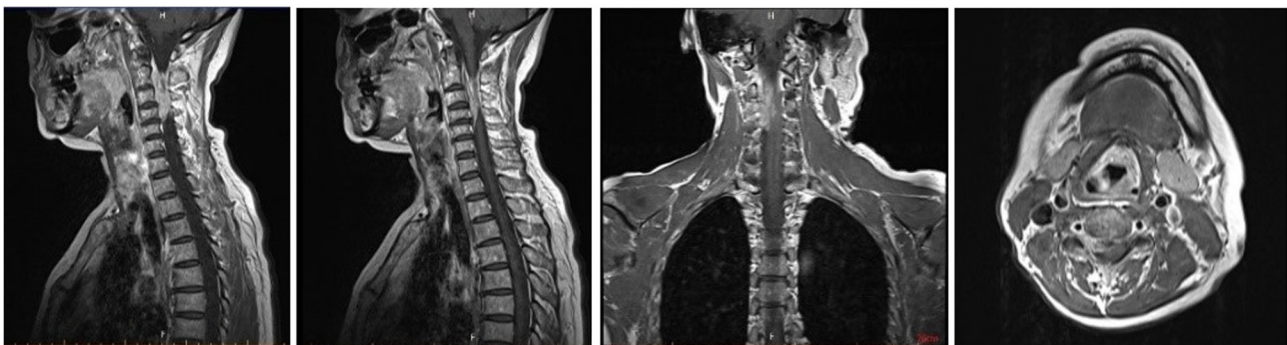


Figure 3. Cervical spine enhanced magnetic resonance imaging 3 days after initial admission.

Differential diagnosis of meningioma includes schwannoma, metastases of cancer, such as breast cancer and lung cancer, in the spinal canal, lymphoma and tuberculoma (14). MRI is the first imaging choice for this disease, and can show the location, size and shape of intraspinal tumors, whether there is bleeding or cystic degeneration in the tumor, and the relationship with neural tissue. Meningioma exhibits low signal intensity on T1-weighted images and high signal intensity on T2-weighted images,

and exhibits moderate homogeneous enhancement after gadopentetic acid injection (3). Intraoperative findings and pathological examination are the main basis for definitive diagnosis.

The main treatment of atypical meningioma is surgical resection, and whether the tumor recurs and the survival time of patients is closely related to the degree of the first surgical resection of the tumor (15,16). In the present case report, the patient was a middle-aged woman; total resection should

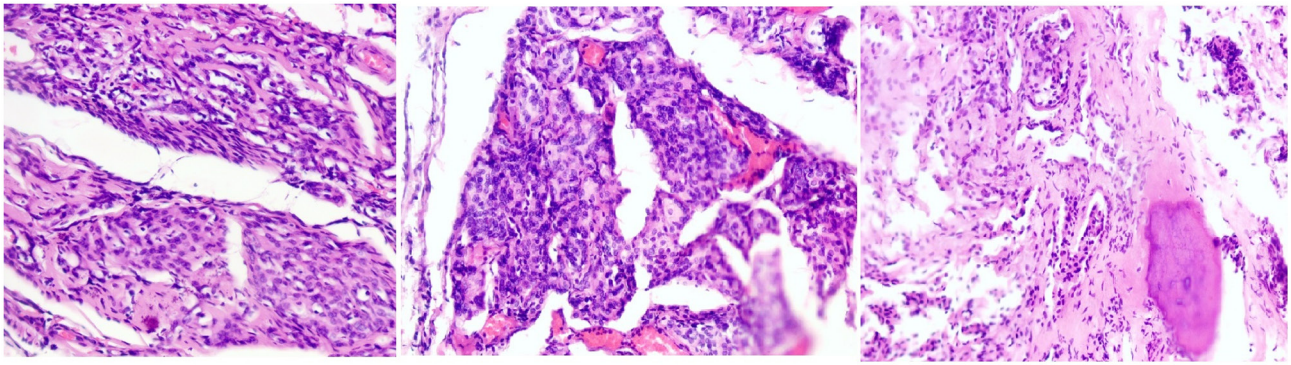


Figure 4. Hematoxylin and eosin staining of pathological sections (magnification, x200).

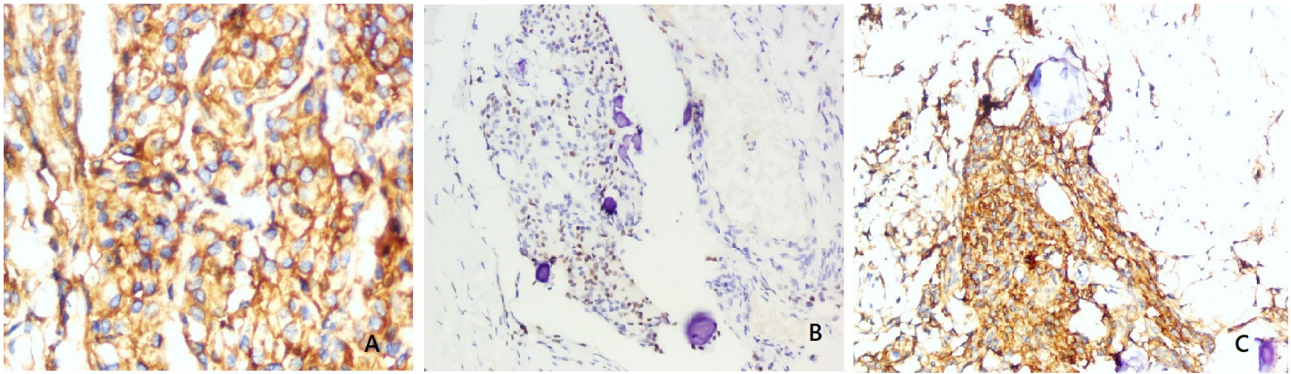


Figure 5. Immunohistochemical sections. (A) Recombinant somatostatin receptor 2 stain (magnification, x400). (B) Progesterone receptor stain (magnification, x200). (C) Epithelial membrane antigen stain (magnification, x200).

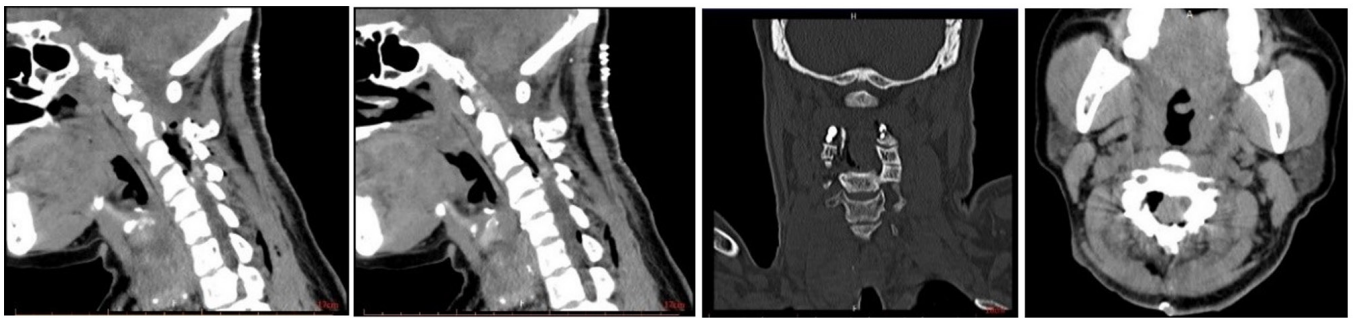


Figure 6. Postoperative cervical spine CT in July 2023.

be completed as much as possible if the diagnosis is not completely confirmed; however, in the present case, the tumor invaded the intervertebral foramen and was accompanied by bone destruction, thus making total resection difficult. In the present case, the tumor was surgically removed under general anesthesia using a microscope. Extramedullary intradural tumors are most commonly located in the anterolateral aspect of the spinal canal (17), and the spinal cord often protrudes backwards due to compression from the meningioma. After opening the dura mater, the relationship between the tumor and the spinal cord and nerve roots should be carefully explored. In the present case, after separating the spinal cord and tumor, the spinal cord was protected with a piece of cotton, the base of the tumor was fully exposed, electrocoagulation

was performed to stop bleeding, and the tumor was removed together with the attached dura mater. Notably, in the case of large tumors, it can be removed in pieces. During tumor resection, the nerve surrounded by the tumor should be carefully separated, and when the nerve root adheres tightly to the tumor or penetrates out of the tumor, the tumor capsule should be incised, the tumor should be removed piecemeal in the capsule and the nerve should be separated as much as possible. In addition, the nerve root should be protected as much as possible, and the tumor should be removed as completely as possible during surgery. When the boundary between the tumor and the spinal cord is unclear, surgeons should be careful and avoid removing the tumor that adheres to the spinal cord to avoid causing spinal cord injury. For

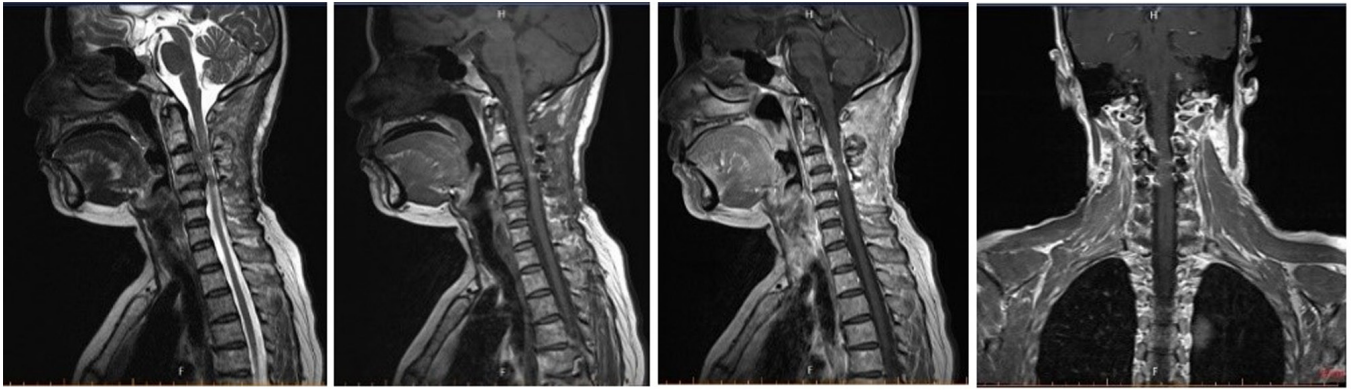


Figure 7. Cervical spine enhanced magnetic resonance imaging in August 2023.

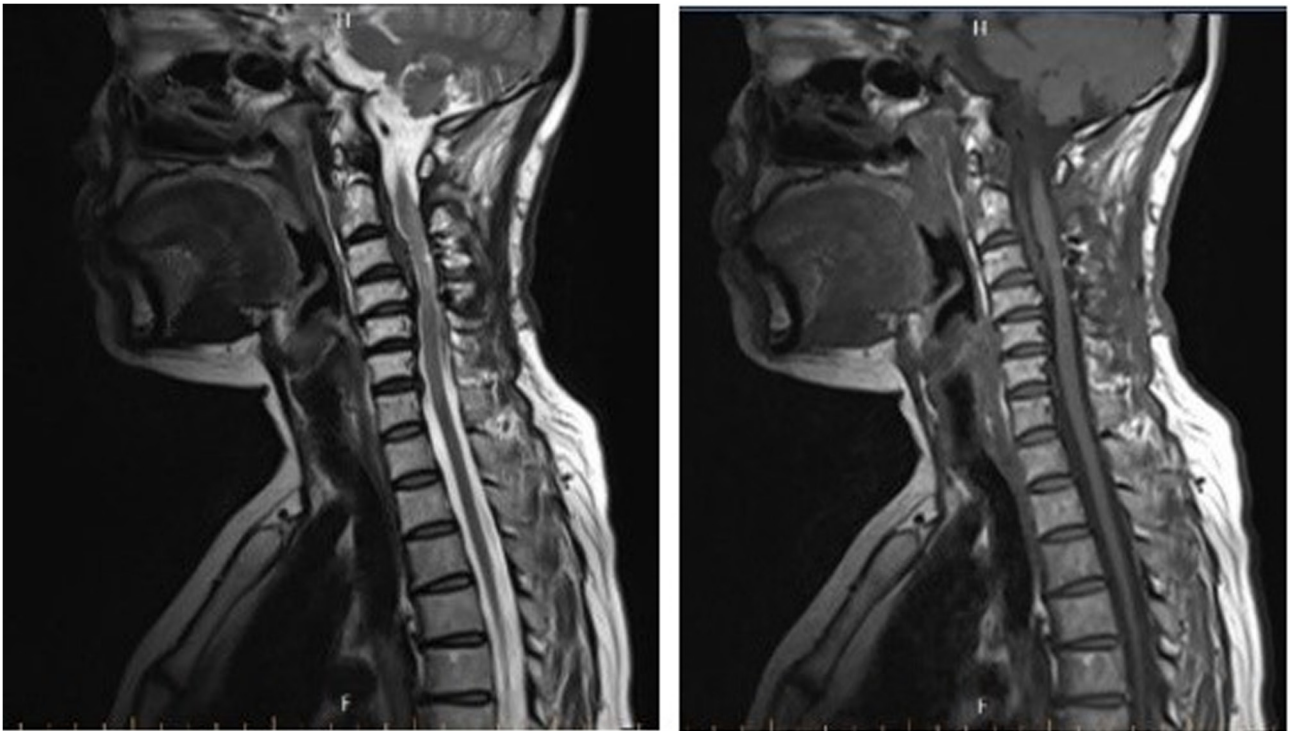


Figure 8. Cervical spine magnetic resonance imaging in October 2023.

patients with subtotal resection, postoperative radiotherapy can achieve a certain period of remission. The irradiation dose is generally 40-50 Gy for 4-5 weeks; however, radiotherapy can cause radiation myelitis. In the present case, adjuvant radiotherapy was performed after surgery because some tumor cells remained in the intervertebral foramen and the patient did not develop radiation myelitis during the treatment.

The long-term prognosis of atypical meningioma is still unclear. Early MRI signals are often affected by surgery. In order to accurately show the extent of tumor resection, MRI should be performed 3 months after surgery in patients with meningioma. MRI is also recommended to be performed every 1-2 years for 10 years after surgery. The global recurrence rate of meningioma is 6-15% and recurrence may depend on whether the tumor is completely resected (18). Globally, if the tumor is completely resected, the 5-, 10- and

15-year recurrence-free survival rates are 93, 80, and 68%, respectively (19); in cases of subtotal resection, the 5-, 10- and 15-year recurrence-free survival rates are 63, 45 and 9%, respectively (20). Other factors associated with poor prognosis are epidural invasion, age <50 years, multiple lesions, calcification, ossification and anterior location of the lesion (8,14,20). In the present case report, the patient was reexamined 3 months after surgery, and no tumor recurrence was observed.

In conclusion, atypical meningioma exhibits nonspecific symptoms before surgery, and the diagnosis mainly depends on intraoperative macroscopic findings and pathological examination. Surgical resection is the main treatment for atypical meningioma and radiotherapy is the main adjuvant therapy after surgery. Notably, total tumor resection can markedly reduce the recurrence rate of postoperative tumors and prolong the survival time of patients.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

LM and MW conceived and designed the study. ZS, LC and GC provided study materials and treated the patient. LM and MW wrote the manuscript. LM and MW confirm the authenticity of all the raw data. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

The patient signed an informed consent form.

Competing interests

The authors declare that they have no competing interests.

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