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Case Report

Intradural Extramedullary Metastasis of the Upper Thoracic Spine – Case Report and Literature Review

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Abstract

Intradural extramedullary metastases of systemic neoplasms are extremely rare and associated with progression of the underlying disease.

We report a case of 46-year-old male operated on for moderately differentiated lung adenocarcinoma. Postoperatively, he underwent six courses of chemotherapy and radiotherapy. He developed progressive severe inferior paraparesis accompanied by excruciating pain between the shoulders two years later. Magnetic resonance imaging revealed metastases in the bodies of T2 and T3 vertebrae with adjacent intradural extramedullary lesion compressing the spinal cord. The patient underwent surgical decompression and vertebral body cement augmentation that lead to pain relief and partial neurological recovery. The histological examination was consistent with metastases from low differentiated pulmonary adenocarcinoma. Surgical resection of intradural extramedullary metastasis improves patient quality of life by reducing pain intensity and neurological deficit.

Key words

intradural extramedullary metastasis, pulmonary adenocarcinoma, surgery

INTRODUCTION

About 70% of patients with neoplastic disease develop spinal metastases, and incidence tends to increase due to population aging, progress in treatment of the primary lesion, and increased patient survival.[1] Intradural extramedullary metastases (IEMs) of systemic neoplasms are extremely rare and associated with their advanced status.[2] In the majority of cases, solitary metastases are located near the nerve roots, making it difficult to differentiate them from tumors of the nerve sheaths.[3] IEMs, including those from primary cerebral tumors, resulting from cerebrospinal fluid dissemination, are only 1.6% - 6% of all spinal tumors.[1,4,5] In a retrospective autopsy study of 627 deceased patients with neoplastic disease, Costigan and Winkelman found that IEMs accounted for 8.5% of metastases in the CNS and 2.1% of all neoplastic diseases, as 84.6% originated from bronchogenic carcinoma.[6]

The purpose of this study is to present a case of IEM from lung carcinoma and to review the relevant literature.

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CASE REPORT

A 46-year-old male who complained of intense back pain between the shoulders was diagnosed with pulmonary carcinoma a year and nine months ago. Chest computed tomography (CT) scan revealed findings suggestive of lung carcinoma that invaded the paravertebral region with progressive growth to the spinal canal at the level of the second and third thoracic vertebra ***(Fig. 1)****.*

The magnetic resonance imaging (MRI) confirmed the presence of a voluminous lesion in the right lung growing to the vertebral canal at the T2-T3 level. A bigger part of the lesion was located paravertebrally, involving and partially destructing the second and third ribs on the right, the right arch and transverse process of the second thoracic vertebra, as well as the ipsilateral costovertebral joint. A smaller part of the neoplasm was located along the neural foramen, reaching medially the thecal sac without compression and dislocation of the spinal cord ***(Fig. 2)****.*

Bilobectomy and partial resection of the vertebral mass were performed. The histological examination of the tumor was consistent with moderately differentiated adenocarcinoma of the lung and bronchus, and histology of the vertebral mass revealed fibrous tissue with adenocarcinoma metastases ***(Fig. 5A)****.*

After surgery, the patient underwent palliative radiotherapy of T2 and T3 (4×3.5 Gy), as well as 6 chemotherapy courses. Eight months later, PET/CT revealed the presence of tumor metastases in the T2 and T3 vertebral bodies as well as emerging pulmo-pulmonary metastasis ipsilaterally. Additional radiosurgery was performed (3×12 Gy). One year later, the patient reported recurrent pain between the shoulders, which gradually irradiated to the legs. Progressive weakness of both legs appeared which impaired the patient's walking to a degree disabling upright standing position. Subsequently, he was admitted to the neurological clinic. Brain CT scan was negative for metastases. After consultation with a neurosurgeon, MRI of the thoracic spine was performed which

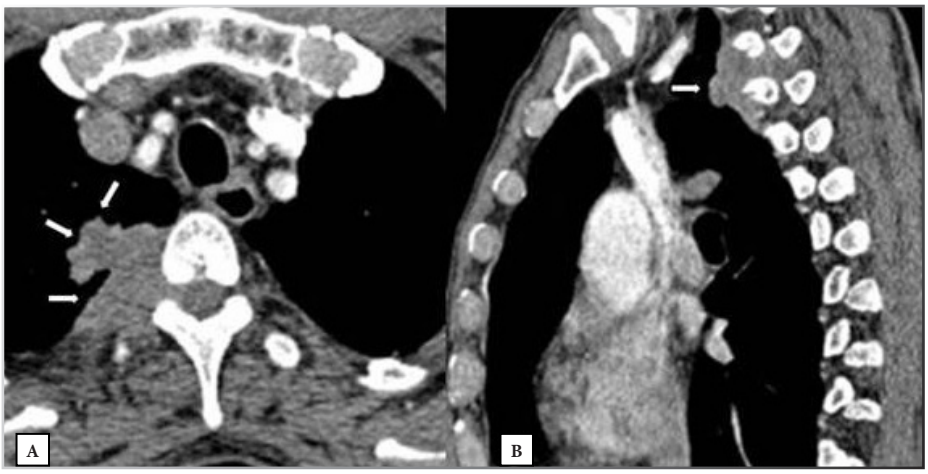

Figure 1. A. Axial CT of the thorax visualizes paravertebrally located lesion in the right lung, penetrating and dilating the T2-T3 neural foramen (arrows); **B**. CT (sagittal reconstruction) shows neoplastic formation paravertebrally to the right and the dilated neural foramen (arrow).

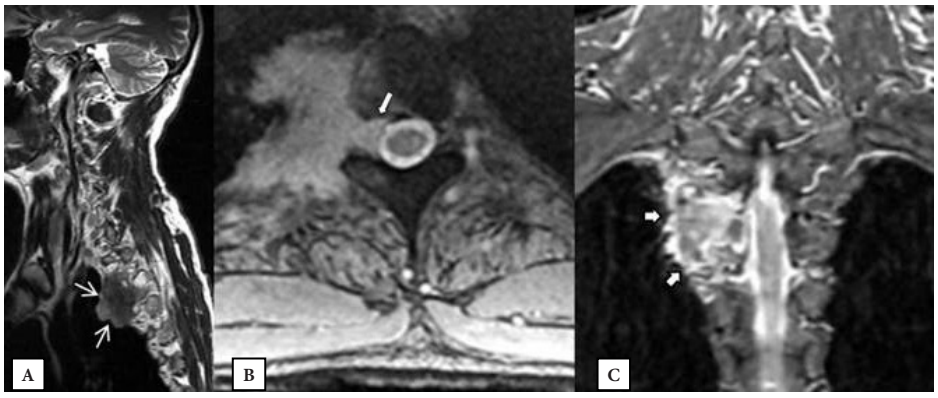

Figure 2. MRT T2. **A**. Sagittal view - tumor formation located on the right at T2-T3 level (arrows); **B**. Axial view - medial penetration of the lesion into the nerve root canal reaching the dura mater without spinal cord compression; **C**. Coronal view - location of the lesion and its relationship with the thecal sac.

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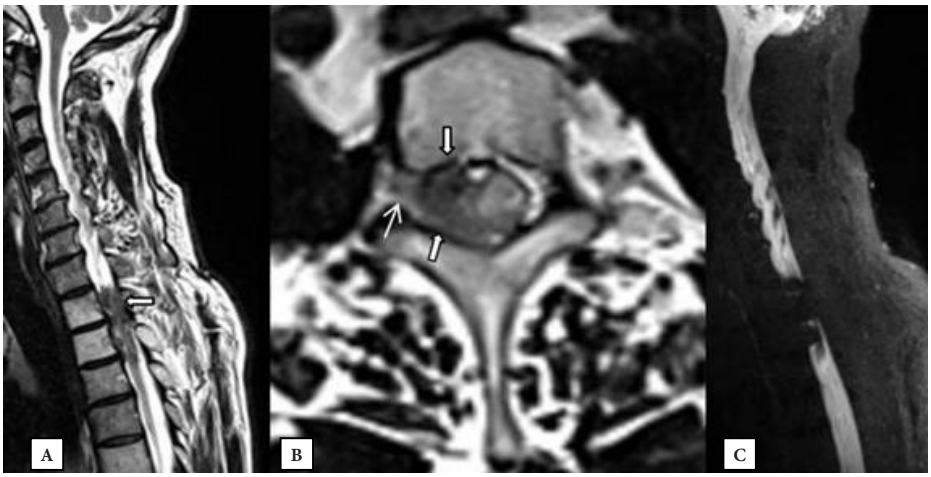

Figure 3. T2-weighted MRI prior to second surgery: **A**. Sagittal view shows the presence of intradural lesion at T2 -T3 level (arrow) and hyperintense signal from the T2, T3 and T6 vertebral bodies; **B**. Axial view - an intradural extramedullary lesion located around the right half of the medulla spinalis (black and white arrows) and extension to the right nerve root canal (white arrow); **C.** MRI myelogram - compressed subarachnoid space at T2-T3 level.

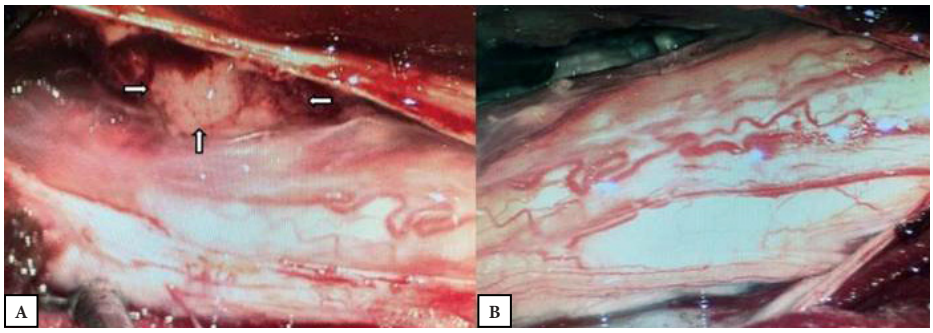

Figure 4. Intraoperative images: **A**. After opening the dura mater a tumor formation located intradurally extramedullary was disclosed (arrows); **B**. after tumor removal.

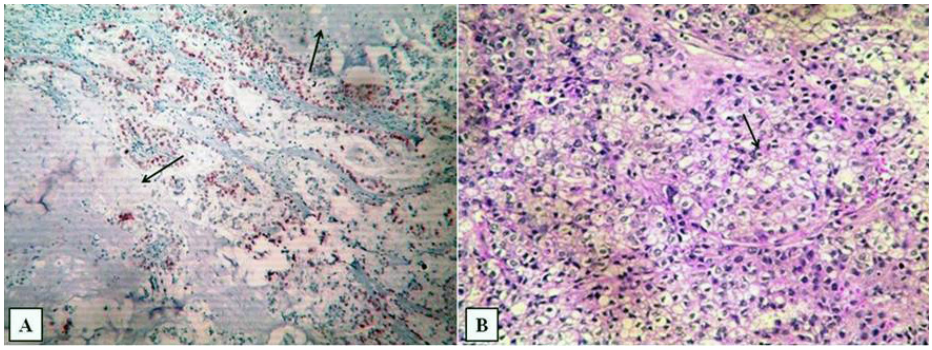

Figure 5. Histological examination from: **A**. Lung: Primary moderately differentiated light cell pulmonary adenocarcinoma with papillary and mucous sections (arrow). IHC, TTF1 positive in the tumour cells nuclei. *Napsin A* positive in cell cytoplasm. Magnification ×40. **B**. Lung adenocarcinoma intradural spinal metastasis presented only by a solid light-cell component (arrow) with mild differentiation. Evident progression of the tumor from moderately differentiated to low differentiated type. Diffuse infiltration of solid nests of light tumor cells with centrally located nuclei. TTF1 moderately positive for tumor nuclei. *Napsin A* is negative in metastatic cells. Magnification ×200 H.E.

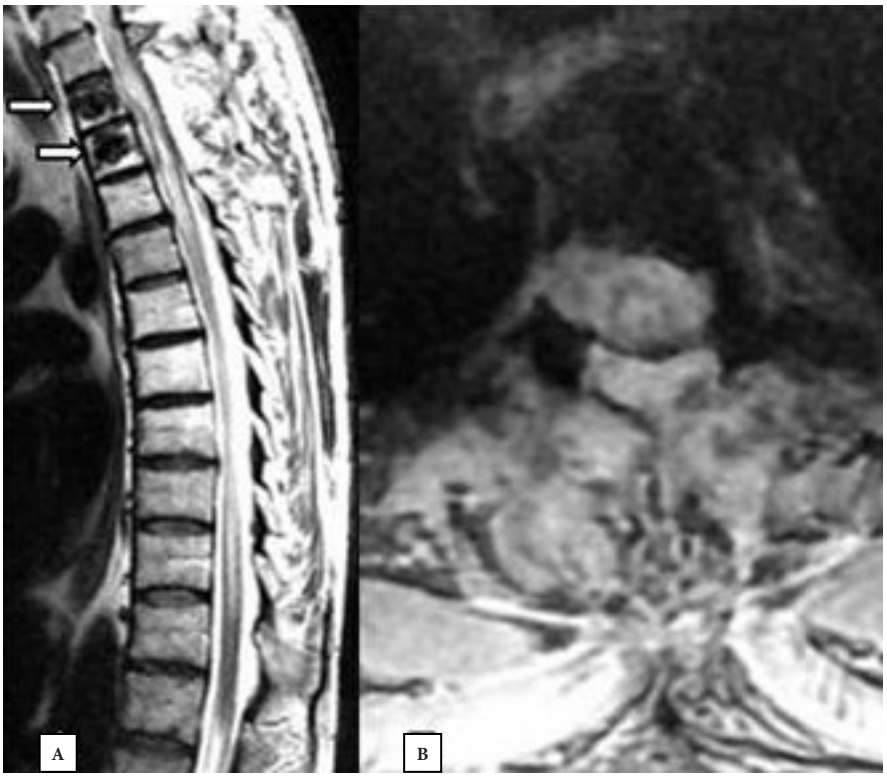

Figure 6. Postoperative T2-weighted MRI: **A**. Sagittal projection shows the performed vertebroplasties of Th2 and Th3 (arrows) and the absence of the intradural extramedullar lesion; **B**. Axial projection.

detected metastases in the T2 and T3 vertebral bodies and intradurally extramedullary lesion located at the T2-T3 level on the right, compressing the spinal cord ***(Fig. 3)****.*

Upon admission to the neurosurgical clinic, the patient had pronounced vertebralgia (Denis scale - grade P4), lower spastic paraparesis (Frankel scale - grade C), conductive hypoesthesia distal to the mamillae and retention of pelvic reservoirs.

The patient was selected for surgery based on the clinical and neuroimaging findings. Vertebral body augmentation with bone cement of the second and third thoracic vertebra was performed followed by same level laminectomies and tumor resection. After dural opening, an intradural extramedullary tumor formation was found to be located ventrolaterally to the right from the spinal cord and contacting firmly to the right T3 nerve root. It was totally removed via microsurgical technique ***(Fig. 4)****.*

The histological examination demonstrated evidence of pulmonary adenocarcinoma metastasis, with tumor progression from moderately differentiated to low differentiated type ***(Fig. 5B)****.*

In the early postoperative period, pain reduction to Grade P3 on the Denis scale was reported. At the beginning of the second postoperative week, myelopathy regressed to Grade D on the Frankel scale - the patient began to walk with assistance. Postoperative MRI confirmed the removal of the tumor ***(Fig. 6)****.*

DISCUSSION

The majority of patients with neoplastic disease develop skeletal metastases and the spine is commonly affected. Great part of the spinal tumors are metastatic, occurring 20 times more often than primary spinal neoplasms.[7] Autopsy studies show that 70% of deceased patients with neoplastic disease have spinal metastases, with only 10% of them experiencing neurological symptoms.[7,8] Spinal metastases occur in all age groups but most often in the age range of 40-65 years and are slightly more common in males, also noted in our case.[7]

Diagnosed IEMs from non-neurogenic neoplasms are very rare, with a frequency of 5% of all secondary tumors of the spine. They result from advanced primary neoplastic disease and most often originate from lung or breast adenocarcinoma, similarly as in the described case.[4,5,9,10] IEMs present a significant neurological complication of systemic neoplasms ranging from 3% to 43% according to clinical and autopsy studies, with only 0.8% - 3.9% being symptomatic. [2,9-11]

Infiltration of tumor cells in the intradural space is possible via cerebrospinal fluid dissemination from brain metastases, via haematogenous pathway (arterial or via Batson venous plexus), via direct invasion of spinal cord sheaths or via the lymphatic perineural ducts.[12] In the present case, the infiltration of the metastasis in the intradural space is

possible via several of the possible pathogenetic mechanisms. The invasion of the pulmonary paravertebral lesion to the nerve root canal and its tight contact to the thecal sac suggests possible penetration into the intradural space either through direct infiltration or through the perineural lymphatic pathway. On the other hand, the presence of additional metastases in other vertebral bodies is in favour of the hematogenous pathway.

Although patients with malignancies live longer due to the contemporary development of the treatment, spinal metastases impair their quality of life and shorten survival span.[13] The localization of the metastasis in our patient confirms Chen and Tsai's statement that the thoracic spine is affected most often (70%), followed by the lumbar (20%) and the cervical (10%) spine.[8]

IEM may occur between several weeks and 17 years after diagnosis and treatment of the primary neoplasm, with frequent worsening of the clinical status and patient's prognosis.[4,12,14] Our case illustrates that, even following fully adequate treatment (radiotherapy, radiosurgery and chemotherapy), the metastatic development is possible within less than 2 years. It demonstrates that post-therapy tumor cells get modified as their receptors lose susceptibility to chemotherapeutic drugs which leads to uncontrolled proliferation of the most resistant and undifferentiated tumor branch.[16]

Upon occurrence of IEM, the average survival of patients is about 7.5 months and survivors are about 10% - 22%.[4,9,12,14]

Postoperative outcome in our patient confirm literature statement that surgical resection of IEM improves patient's quality of life by reducing the pain intensity and improving the neurological status.[12,17]

CONCLUSION

Although IEMs are rare, physicians should bear in mind that they are one possible complication of systemic cancer, especially in cases where the primary malignancy is associated with a paraneoplastic syndrome with concomitant neurological dysfunction.[18] The omission of IEM may lead to ineffective treatment and failure to improve the patient's quality of life.

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Интрадуральный экстрамедуллярный метастаз в верхнем грудном отделе позвоночника - клинический случай и обзор литературы

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Абстракт

Интрадуральные экстрамедуллярные метастазы системных новообразований встречаются крайне редко и связаны с прогрессированием скрытого заболевания.

Мы сообщаем о случае 46-летнего мужчины, оперированного по поводу слабо дифференцированной аденокарциномы лёгких. В послеоперационный период он прошёл шесть курсов химиотерапии и лучевой терапии. У него развился прогрессирующий тяжёлый парапарез нижней части тела, сопровождаемый мучительной болью в плечах через два года. ЯМР выявил метастазы в тела позвонков Т2 и Т3 с прилежащим интрадуральным экстрамедуллярным поражением, давящим на позвоночник. Пациент подвергается хирургической декомпрессии и аугментации с помощью цемента тела позвонка, что привело к облегчению боли и частичному неврологическому восстановлению. Гистологическое исследование подтвердило наличие метастазов от низкодифференцированной лёгочной аденокарциномы. Хирургическая резекция интрадурального экстрамедуллярного метастаза улучшила качество жизни пациента, уменьшив интенсивность боли и неврологический дефицит.

Ключевые слова

интрадулярный экстрамедуллярный метастаз, аденокарцинома лёгких, хирургия