

CASE REPORT

Longitudinally extensive transverse myelitis as presenting manifestation of small cell carcinoma lung

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Abstract

Longitudinally extensive transverse myelitis (LETM) is an unusual manifestation of systemic malignancy. It has been mainly reported with lung cancers and lymphoproliferative malignancy. LETM in systemic malignancy can be caused by either intramedullary metastases or paraneoplastic syndrome. We report an unusual case of small-cell carcinoma lung, who presented with LETM without having any cardinal manifestations of lung malignancy. This case report highlights the important differentiating features between intramedullary metastasis and paraneoplastic syndrome.

INTRODUCTION

Paraplegia in patients with malignancy is almost always an ominous sign associated with spinal cord metastasis; however, it can also be associated with paraneoplastic syndromes [1]. Myelitis is a rare paraneoplastic manifestation of malignancy, mostly seen with lung cancers and lymphoproliferative malignancies [2]. Other malignancies include sarcoma, carcinoma breast, prostate, skin, stomach, esophagus, thyroid, ovary, renal cell carcinoma and hepatocellular carcinoma [2, 3]. Longitudinally extensive transverse myelitis (LETM) is the inflammation of spinal cord involving three or more vertebral segments vertically [4]. LETM as a presenting feature of lung cancers is very rare.

We herein report a case of small-cell carcinoma lung in a 66-year-old male, who presented with longitudinally extensive transverse myelitis without having any cardinal features of lung malignancy.

CASE REPORT

A 66-year-old farmer, chronic smoker with 36 pack-years of exposure, presented with acute onset weakness of both lower limbs along with retention of urine for 5 days. There was no history of fever, trauma, back pain, radicular pain, diminution of vision, cough, hemoptysis, tuberculosis, diabetes and hypertension. There was history of acute onset weakness involving left upper limb 2 months back, which gradually recovered over a period of 1 month with physiotherapy. On general examination pulse rate was 70 bpm, blood pressure 130/90 mmHg, respiratory rate 18/min, and temperature was 98.6°F (37°C). Neurological examination revealed MMSE score 26/30 and detailed mental status examination including behavior, consciousness level, attention, language, memory, constructional ability, higher cognitive functions and related cortical functions was normal. Frontal assessment battery (FAB) score was 15/18. Cranial nerves including fundus examination were normal. On motor examination there

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was hypotonia in both lower limbs with motor power MRC grade 0/5. Deep tendon reflexes were absent in both lower limbs and brisk in left upper limb. Plantars were mute bilaterally. There was sensory level at T-4 spinal level with loss of sensations for all modalities. Cerebellar signs and signs of meningeal irritation were absent. Examination of other systems was normal.

Routine investigations including hematology, biochemistry and ECG were normal. X-ray chest showed a mass lesion in upper lobe of right lung.

MRI of the cervico-thoracic spine showed hyperintense signal on T₂-weighted image involving predominantly central spinal cord from C-2 to T-3 vertebral levels with no contrast enhancement, suggestive of LETM (Fig. 1a). Heterogeneous predominantly hypointense signals were also seen in spinal cord extending from T-4 to T-9 vertebral level with evidence of hemorrhage in it and showing abnormal contrast enhancement (Fig. 1b and c). MRI brain

demonstrated multiple hemorrhagic metastatic lesions involving bilateral frontal lobe, left parietal region and right cerebellar hemisphere with surrounding edema (Fig. 2a-c). In view of mass lesion in X-ray chest, contrast enhanced CT scan of thorax was done, which showed lobulated mass of size 81 × 55 × 65 mm in anterior segment of right upper lobe with infiltration of mediastinal pleura and pericardium, leading to mild right pleural and pericardial effusions suggestive of neoplastic lesion (Fig. 3). There was also enlargement of right paratracheal, aorto-pulmonary and bilateral hilar lymph nodes. CT-guided FNAC of lesion was suggestive of small cell carcinoma. CSF examination revealed leukocyte count 2 cells/mm³, protein 57 mg/dl, glucose 60 mg/dl (serum glucose 102 mg/dl) and chloride 118 meq/l. CSF microscopy and serology were negative for virus, fungi and acid-fast bacilli. CSF fungal and bacterial cultures were negative. CSF was also negative for malignant cells. Serum neuromyelitis optica antibodies titer was negative.

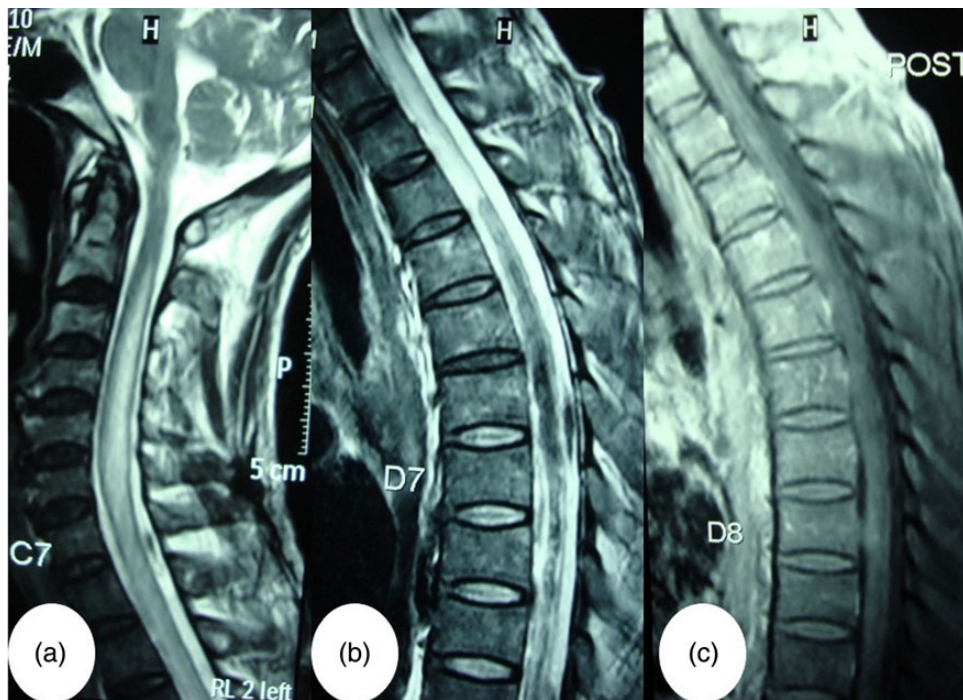


Figure 1: Sagittal T₂-weighted and post-contrast MR images of the spine showing hyperintense signal in spinal cord from C-2 to T-3 vertebral level (a), heterogeneous predominantly hypointense signal from T-4 to T-9 level (b) with abnormal contrast enhancement (c).

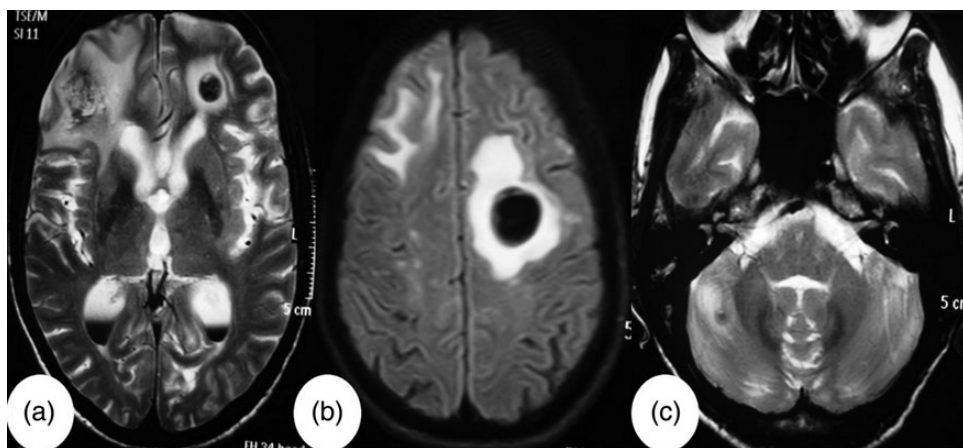


Figure 2: T₂-weighted axial MR images of brain showing hemorrhagic intra-axial lesion associated with moderate to severe edema in bilateral frontal lobes (a), left parietal region (b) and right cerebellar hemisphere (c).

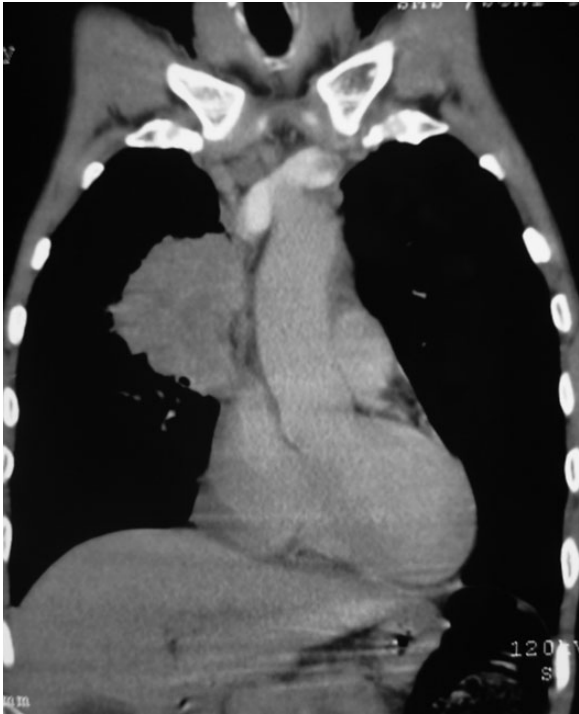


Figure 3: CT scan thorax showing large lobulated mass lesion in suprahilar location in right upper lobe.

In view of LETM, injection methylprednisolone was given intravenously for 5 days but there was no improvement. The patient was transferred under the care of the oncology team and a biopsy was recommended for confirmation of the diagnosis and to aid further management. Unfortunately, no consent was given for this and the patient self-discharged against medical advice and has not returned for outpatient follow-up appointments.

DISCUSSION

There are many etiologies for the LETM; however, the most common is inflammatory demyelinating disease [5]. Neoplastic diseases can cause long segment myelitis through intramedullary metastases, intramedullary tumor, such as ependymoma, intravascular lymphoma and paraneoplastic syndrome [5]. LETM due to intramedullary spinal cord metastases is very rare. Intramedullary spinal cord metastases is seen in only 0.1–0.4% patients of neoplastic diseases [5]. The common underlying malignancies are lung, breast, renal cell carcinomas and melanoma [5]. In a review, most of the cases of intramedullary metastases demonstrated contrast enhancement and extensive edema on T₂-weighted images above and below the metastasis on MR imaging [5]. Intramedullary metastatic lesions have predilection for thoracic spinal cord and are usually solitary. These lesions lie eccentrically and can cause expansion of the cord [6]. Paraneoplastic spinal cord lesions are very rare. They may occur even before the malignancy is detected [7]. Paraneoplastic myelopathy can manifest in two ways: one is the massive cord necrosis presenting as an ascending transverse lesion and other is the patchy multifocal necrosis involving mainly white matter of the spinal cord similar to that of multiple sclerosis [8]. It has been postulated that the underlying malignancy and part of the involved nervous system share common antigens, thereby mounting an autoimmune response [8]. Several antibodies such as CRMP-5, VGCC, amphiphysin, ganglionic AchR, VGKC, ANNA-1,2 have

been associated with paraneoplastic spinal cord lesions [7, 9]. In a systematic review of 31 cases of paraneoplastic myelopathy it was observed that disability develops rapidly and is usually severe. Only a minority of patients showed improvement with treatment [2, 10]. Dominant MRI finding was symmetric longitudinally extensive T₂-weighted signal abnormality in a tract/gray matter distribution that often showed symmetric enhancement [7, 9, 10]. Patchy gadolinium enhancement has also been seen in a patient with necrotizing paraneoplastic myelopathy [10]. CSF analysis may show increased protein concentration in paraneoplastic myelitis [7].

Our case seems to be quite unusual as he had presented with longitudinally extensive transverse myelitis without cardinal manifestations of lung malignancy. Rapid progression, no response to treatment and symmetric hyperintense signal from C-2 to T-3 vertebral levels predominantly involving central spinal cord in T₂-weighted MR imaging suggests the paraneoplastic etiology of LETM. However, in view of intracranial hemorrhagic metastasis, heterogeneous signals in spinal cord extending from T-4 to T-9 vertebral level with evidence of hemorrhage and abnormal contrast enhancement, possibility of intramedullary spinal cord metastasis as an etiology of LETM can also be considered. The diagnosis could not be confirmed between the two possibilities as the patient's family members were not willing for further investigations and treatment.

This case report highlights that LETM is a rare presenting manifestation of malignancy. It can be caused by either paraneoplastic etiology or may be due to intramedullary spinal cord metastasis. When an elderly patient comes with LETM, possibility of any underlying malignancy should always be sought with thorough work up, as the demyelinating diseases are rare in this age group.

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