Cauda Equina Syndrome Secondary to Leptomeningeal Carcinomatosis of Gastroesophageal Junction Cancer

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Key Words
Cauda equina · Leptomeningeal carcinomatosis · Gastroesophageal cancer

Abstract
Leptomeningeal carcinomatosis (LMC) is a diffuse or multifocal malignant infiltration of the pia matter and arachnoid membrane. The most commonly reported cancers associated with LMC are breast, lung, and hematological malignancies. Patients with LMC commonly present with multifocal neurological symptoms. We report a case of LMC secondary to gastroesophageal junction cancer present initially with cauda equina syndrome. A 51-year-old male patient with treated adenocarcinoma of the gastroesophageal junction presented with left leg pain, mild weakness, and saddle area numbness. Initial radiological examinations were unremarkable. Subsequently, he had worsening of his leg weakness, fecal incontinence, and urine retention. Two days later, he developed rapidly progressive cranial neuropathies including facial diplegia, sensorineural hearing loss, dysarthria, and dysphagia. MRI with and without contrast showed diffuse enhancement of leptomeninges surrounding the brain, spinal cord, and cauda equina extending to the nerve roots. Cerebrospinal fluid cytology was positive for malignant cells. The patient died within 10 days from the second presentation. In cancer patients with cauda equina syndrome and absence of structural lesion on imaging, LMC should be considered. To our knowledge, this is the first case of LMC secondary to gastroesophageal cancer presenting with cauda equina syndrome.
Introduction

Leptomeningeal carcinomatosis (LMC) is defined as a diffuse or multifocal malignant infiltration of the pia mater and arachnoid membrane. It is clinically diagnosed in 5–10% of all cancer patients [1, 2]. The most commonly reported cancers associated with LMC are breast, lung, and hematological malignancies. Patients with LMC commonly present with multifocal neurological symptoms. Symptoms are related to increased intracranial pressure, hemispheric dysfunction, cranial neuropathies, and spinal roots dysfunction. We report a case of LMC secondary to gastroesophageal junction cancer initially presenting with cauda equine syndrome.

Case Presentation

We present the case of a 51-year-old right-handed male patient diagnosed with signet ring adenocarcinoma of the gastroesophageal junction. He had surgical resection followed by chemotherapy and radiation therapy. Follow-up showed no evidence of disease recurrence.

Nine months later, he complained of left hip pain followed by mild left leg weakness. Subsequently, the patient was admitted with intestinal obstruction. During admission, he had gradual worsening of the left leg weakness and the development of right leg weakness. It associated with lower limb numbness and saddle area numbness. Initial spine magnetic resonance imaging (MRI) was normal. Nerve conduction study revealed mild motor axonal neuropathy. The patient was discharged from hospital after conservative treatment of the bowel obstruction.

Eight days following discharge, he presented to the emergency department with fecal incontinence and urinary retention requiring Foley catheter insertion and worsening of his lower limb weakness. Repeat spine MRI with contrast was normal. Two days later, he developed progressive bilateral facial weakness, slurring of speech, and swallowing difficulties. He had no headache, double vision, and no alteration of mental status. His review of systems revealed significant weight loss.

Examination of the patient during admission to neurology revealed dysarthric speech. His cranial nerve examination revealed facial diplegia, bilateral sensorineural hearing loss, and weak gag reflex. He had mild asymmetrical upper limb weakness and normal upper limb reflexes. He showed hypotonia of lower limbs, severe asymmetrical weakness, and absent lower limb reflexes. He had length-dependent sensory loss pinprick and normal vibratory and proprioceptive sensation. Furthermore, the patient had absent rectal tone. General examination was unremarkable.

The patient had repeat spine MRI with contrast (fig. 1) which showed diffuse leptomeningeal enhancement surrounding the spinal cord and cauda equina and associated with extension of the enhancement of the nerve roots. MRI brain with contrast (fig. 2) showed diffuse leptomeningeal enhancement most prominent at basal CSF cistern and cerebellum with extension into the cerebellar sulci and surround the brain stem. Cerebrospinal fluid (CSF) examination showed: protein = 648.3 mg/dl (15–45 mg/dl), glucose = 7 mg/dl (40–75 mg/dl), WBC = 39 cells/dl (lymphocytes 59–70%), and RBC = 11–24 cells/dl.

Initial CSF cytology was negative. Repeat CSF cytology showed malignant cells which is large atypical cells with enlarged nucleus and prominent nucleoli (fig. 3). Immunohistochemistry the atypical cells are weakly positive for CK AE1/AE3 and negative for CD68.

The patient was diagnosed with LMC. He had progressive worsening of his cranial neuropathies and died 10 days after hospital admission.
Discussion

LMC is a rare devastating complication of systemic cancer. The most common encountered malignancies are breast 35%, lung 24%, and hematological malignancies 16% [1]. The development of LMC with gastrointestinal tumors is rare and most commonly reported with gastric cancer. Few case reports described the development of LMC with esophageal cancer and gastroesophageal junction tumors [3–9]. Adenocarcinoma is the most commonly reported esophageal and gastric cancer associated with LMC.

The tumor cells disseminated to the leptomeninges occur via direct invasion, hematogenous spread, or perineural spread especially in head and neck cancer [10].

Symptoms related to LMC are typically multifocal in nature and resulted from different mechanisms. Patients may present with features of increased intracranial pressure related to CSF flow obstruction, neuronal dysfunction from metabolic competition for nutrients, and vascular alteration due to tumor growth [11]. Symptoms can be divided into those resulting from hemispheric dysfunction, cerebellar dysfunction, cranial neuropathies, and spinal radiculopathies. In a study of 118 breast cancer patients with LMC, headache was the most common reported symptom (54%), followed by cranial neuropathies (42%), cerebellar signs (35%), paresis (26%), mental changes (19%), meningism (11%), seizure (9%), and radicular pain (7%) [12].

The development of cauda equina syndrome secondary to LMC has been previously in a 66-year-old female patient with advanced ovarian cancer [13]. Her MRI was normal. Her CSF cytology was positive for malignant cells. Our patient normal imaging initially but subsequently it showed an evidence of diffuse leptomeningeal enhancement. High clinical suspicion should be considered in any cancer patient presenting with radicular symptoms and features of cauda equina syndrome. The presence of normal imaging studies does not rule out the presence of LMC. In the breast cancer LMC study by Niwinska et al. [12], 8% of patients had normal neuroimaging initially. CSF examination remained the gold standard for diagnosing LMC. Initial CSF examination will be positive only in 50% of cases, but the yield will increase up to 90% after the third exam [11].

In cancer patients with cauda equina syndrome and absence of structural lesions on imaging, LMC should be considered and further testing with study of the CSF is required. To our knowledge, this is the first case of LMC secondary to gastroesophageal cancer presenting with cauda equina syndrome.

Statement of Ethics

The authors have no ethical conflicts to declare.

Disclosure Statement

The authors have no conflicts of interest to declare.

References

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Fig. 1. MRI of the spine with and without contrast showing diffuse leptomeningeal enhancement surrounding the spinal cord and cauda equina and associated with extension of the enhancement of the nerve roots.
Fig. 2. MRI of the brain with and without contrast showing diffuse leptomeningeal enhancement most prominent at basal CSF cistern and cerebellum with extension into the cerebellar sulci and surround the brain stem.

Fig. 3. CSF cytology showing malignant cells, i.e. large atypical cells with enlarged nucleus and prominent nucleoli.