

Case Report

Rare Oncologic Emergency in Esophageal Cancer

Gannamani V^{1*}, Yegneswaran B¹ and Porcelli M²¹Department of Internal Medicine, Saint Peter's University Hospital, USA²Department of Hematology & Oncology, Saint Peter's University Hospital, USA

***Corresponding author:** Vedavyas Gannamani, Department of Internal Medicine, Saint Peter's University Hospital, 254 Easton Avenue, CARES building 4th floor, New Brunswick, New Jersey, USA

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Abstract

Less than 1% of cancer patients develop intramedullary spinal cord metastases (ISCM). They present with rapid progression of neurological symptoms and cause severe disability. With advent of imaging techniques and improved survival of patients, ISCM are increasingly recognized. To the best of our knowledge few cases of ISCM from esophageal cancer were reported from the United States. The patient we present here was diagnosed with adenocarcinoma of distal esophagus in June 2013 and later developed liver, lung and multiple brain metastases. He had stable disease for three months before this hospitalization. The reason for admission was history of multiple falls in three weeks. Examination showed reduced motor strength in both legs, left greater than right. Also absent ankle and knee reflexes bilaterally and reduced position sense in right leg was noted. An initial MRI of brain showed multiple metastatic lesions including left parietal lobe (30 mm in size) and right occipital lobe (23 mm in size). The physical exam findings were difficult to explain by MRI brain. Further evaluation by MRI spine showed focal intramedullary mass at T11 level with surrounding edema. Patient was treated with Decadron® and palliative radiation. At the end of radiotherapy, the improvement in neurological status was only minimal. Three months after the diagnosis, he died on hospice care.

Keywords: Intramedullary tumors; Esophageal cancer; Spinal cord tumors

Abbreviations

ISCM: Intramedullary Spinal Cord Metastases; BUN: Blood Urea Nitrogen; MRI: Magnetic Resonance Imaging; CNS: Central Nervous System

Introduction

Less than 1% of cancer patients develop intramedullary spinal cord metastases (ISCM). These tumors constitute 2-8.5% of all central nervous system metastases and are less than 5% of spine metastases [1,2]. Lung and breast cancers are commonly associated with ISCM [1-3]. Esophageal cancer rarely metastasizes to brain and even less rarely to spinal cord [4]. With improvement in imaging studies, ISCM are being increasingly recognized. Understanding of their management is necessary to reduce the disability. Worldwide, to our knowledge three cases of esophageal cancer with ISCM involving cervical region have been described and reported [4-6]. We here present a case of ISCM with involvement of lower thoracic region from esophageal cancer.

Case Presentation

An 82-year-old male with past medical history of hypertension, hyperlipidemia, tongue cancer, chronic kidney disease, benign prostate hyperplasia and metastatic esophageal cancer was admitted with the history of multiple falls in three weeks. Patient was diagnosed with moderately differentiated adenocarcinoma involving distal third of esophagus in June 2013 and received definitive chemotherapy and radiation. A follow up PET scan in February 2014 showed new metastatic lesion in liver, treated with radiofrequency ablation in May 2014. Therapy was delayed as patient was hospitalized with metastatic disease of brain and had fractionated brain radiation. Follow up

imaging revealed metastatic lung disease for which patient received chemotherapy. In early 2015 the disease progressed manifesting as multiple brain metastases and whole brain radiation was given. Patient then had stable disease with Kornafsky's score of 60-70 until the current admission in July 2015.

At admission patient complained that his legs were "giving away". His symptoms worsened gradually over three weeks. Initially he was unable to stand which progressed to the need of a wheel chair for ambulation. He had no history of incontinence, back pain or trauma. Patient did not have weakness of upper limbs, speech or swallowing difficulty.

His vital signs on admission included blood pressure of 120/71 mmHg, heart rate of 51/min, regular, temperature of 97.4F, respiratory rate of 16/min. Examination showed motor strength of 2/5 in left

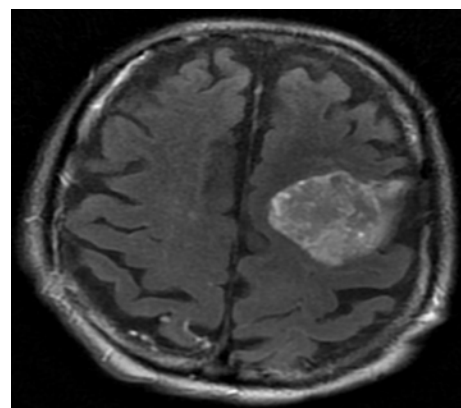


Figure 1: MRI of brain (T2 Flair) showing large left parietal metastatic lesion.

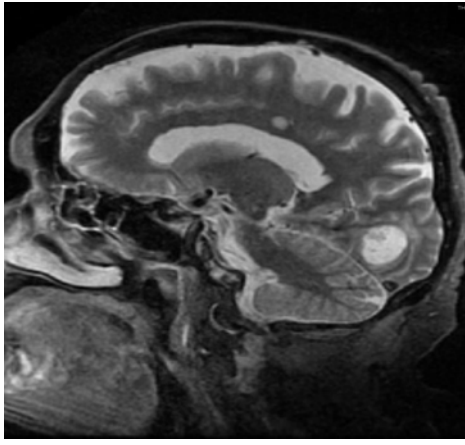


Figure 2: MRI of brain Sagittal section, STIR (Short T1 Inversion Recovery) showing multiple metastatic lesions.

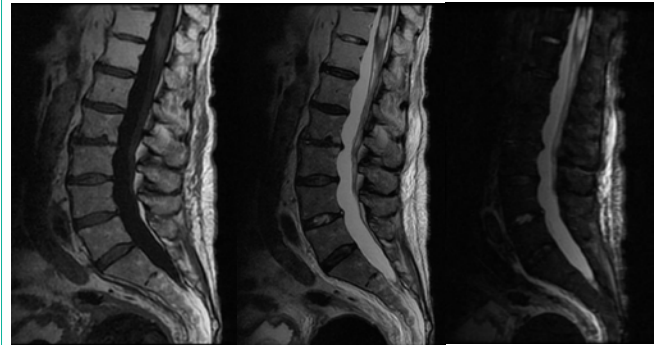
leg with no ability to move distal part of left leg and 3/5 strength in right leg; reduced position sensation in right foot, absent knee and ankle reflexes bilaterally. The Babinski's test was negative bilaterally. Rest of the examination was unremarkable. His admission labs included WBC $7.0 \times 10^9/L$, hemoglobin 100 gm/L, platelets count of $134 \times 10^9/L$, serum sodium 134 mmol/L, potassium 4.5 mmol/L, BUN 17.5 mmol/L, creatinine 162 $\mu\text{mol/L}$ and calcium 2.28 mmol/L. An initial MRI of the brain showed a large 30 mm left parietal metastatic lesion with edema, a 23mm right occipital lesion and multiple small metastases in both hemispheres (Figure 1 and 2). His symptoms and physical findings were not completely explained by the MRI findings.

The new onset paraplegia with loss of position sense and considerable reduced strength in left leg was difficult to explain by findings on MRI of brain leading to further evaluation by MRI Spine. It showed a focal intramedullary mass at level of T11 with surrounding edema. Increased T1 and T2 intensity was noticed in lower thoracic region consistent with history of prior radiation. There was no evidence of osseous metastatic disease or epidural cord compression (Figure 3,4 and 5). An intramedullary biopsy is a complex procedure and given patient's overall condition at the time of presentation, the metastasis was presumed to be from esophageal cancer and no biopsy was contemplated. Patient was treated with both Decadron and radiation. Strength in left leg improved after first three cycles of radiation but by the end of 10 cycles of treatment, the change was minimal. Three months after the diagnosis, he died on hospice.

Discussion

ISCM are very rare tumors seen in patients with wide spread metastatic disease. With improved survival of patients, uncommon tumors like ISCM are being seen in clinical practice more frequently [1]. In contrast to primary spinal cord tumors, ISCM have rapid progression of symptoms within days to few weeks. Most commonly these tumors present with weakness of limb(s), sensory deficits, pain or incontinence and rarely as Brown-Sequard Syndrome [1,2]. These tumors are reported to involve every level of spinal cord from cervical region to lumbar region [2,7].

The primary cancers frequently associated include lung and breast cancers [2,3]. Other cancers that have been reported are renal



Left - Right

Figure 3: Sagittal T1 Flair. MRI Thoracolumbar Spine showing intramedullary lesion at level of T11 vertebrae.

Figure 4: Sagittal T2 FRFSE (Fast Relaxation Fast Spin Echo).

Figure 5: Sagittal STIR (Short T1 Inversion Recovery).

cell cancer, lymphoma and melanoma. One or more cases of ISCM have been reported with gastric cancer, colon cancer, rectal cancer, cholangiocarcinoma and others [8-10]. In most situations ISCM are seen in patients with widely metastatic disease. Pulmonary and brain or other CNS metastases are common associations [1,2,7].

Other reported cases of ISCM with esophageal cancer showed involvement of cervical region of spinal cord. The type of esophageal cancers described in these reports were squamous carcinoma, neuroendocrine carcinoma and adenocarcinoma [4-6]. The patient in this discussion had adenocarcinoma with involvement of spinal cord at T11 level. Studies done on Western population with brain metastases from esophageal cancer showed adenocarcinoma as commonly associated type [11]. However very few studies are available now to comment on likeliness of a specific pathological type of esophageal cancer causing ISCM.

The treatment for ISCM is not yet standardized. Including corticosteroid administration, therapy is aimed at improving neurological status and thus functionality. Various treatment strategies are being practiced. Treatment options include conservative approach with radiation and steroids and/or resection. Biopsy with chemotherapy based on histological diagnosis of primary malignancy also been proposed [12,13]. Surgical treatments include partial or total resection, which can be combined with adjuvant radiation. Surgery offers better outcomes compared to radiation [1,2,7,12,13]. Of the three cases of ISCM with esophageal cancer we reviewed, two patients received palliative radiotherapy for spinal cord lesions [4-6]. Patients with ISCM have poor prognosis and survival is usually few months. Among the above reviewed cases with esophageal cancer, one patient died in 20 days and one survived for 6 months [4-6].

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