Case Report

Extrapulmonary Small Cell Carcinoma of Spine Presenting with a Thoracic Epidural Mass: Case Report

Hakan AK, Varol AYDIN
Adnan Menderes Üniversitesi, Nöroşirurji Anabilim Dalı, Aydın, Türkiye

Abstract
Extrapulmonary small cell carcinomas have the same histological properties of small cell carcinoma of the lung which is a rare and distinct entity. They are seen more commonly in males and genitourinary and gastrointestinal tracts are the most involved systems. Cases arising from different organs have been reported in the literature but there is only one case reporting arising from vertebrae. In this report we are presenting the second extrapulmonary small cell of spine case presenting with an epidural mass in a 35 year old woman. Our definite diagnosis was performed after surgery. Patient received chemotherapy regimen consisting of etoposide, cisplatin and zoledronic acid. Further studies are still needed for the early diagnosis and the optimal management of these tumors.

Keywords: Spine, small cell carcinoma, incidence, epididual mass

INTRODUCTION
Extrapulmonary small cell carcinomas (EPSCC) are the tumors having the histological appearance of small cell carcinoma of the lung which is a rare and distinct entity. They are seen more commonly in males and genitourinary and gastrointestinal tracts are the most involved systems. Cases occurring from different organs have been reported in the literature but there is only one case reporting arising from vertebrae. In this report we are presenting the second extrapulmonary small cell of spine case presenting with an epidural mass in a 35 year old woman. Our definite diagnosis was performed after surgery and patient received chemotherapy regimen consisting of etoposide, cisplatin and zoledronic acid. Further studies are still needed for the early diagnosis and the optimal management of these tumors.

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**CASE PRESENTATION**

A 35 year old woman admitted to our clinic with the complaint of tenderness and weakness at the both legs lasting about 1 month. There was no history of smoking, trauma, infection, weight loss or any accompanying disease. Sphincter functions were normal. Her past and family history didn't reveal any pathology. In the neurological examination there was hypoesthesia below third thoracic dermatome, muscle strength was intact. Babinski sign and clonus was negative bilaterally. Dorsal MRI (magnetic resonance imaging) revealed an epidural mass between Th2-4 (Figure 1a&b). Complete blood count, liver and kidney function tests were normal. Tumor markers were negative. Chest X-ray was normal. All body bone scintigraphy revealed no other lesion. Patient underwent surgery after approval of anesthesia. In the operation, thoracic laminectomy was performed and an epidural mass originating from the left foramina of Th3 (third thoracic vertebrae) was seen (Figure 2). Mass was reddish in color and densely haemorrhagic, it was soft but not fragile. It was removed totally. Dura was intact (Figure 3a&b). Patient was discharged after 3 days of operation without any complication or complaint. Pathologic evaluation revealed small cell carcinoma with basophilic nucleus and indistinct cytoplasmic borders. Immunohistochemical staining demonstrated positive staining with cytokeratin, synaptophysin, and chromogranin. Nonspecific staining was seen with S100. Patient was consulted to the oncology clinic and chemotherapy regimen consisting of etoposide, cisplatin and zoledronic acid was started.

*Figure 1a&b: Preoperative T1 weighted sagittal and T2 weighted axial MRI images of the thoracic vertebrae demonstrating the mass.*
DISCUSSION

Patients having extrapulmonary small cell carcinoma have the histologic diagnosis of small cell carcinoma, however, their chest X-ray or computed tomography generally are normal even more their sputum cytology or bronchoscopic examination may also be normal\(^2\). This is a rare pathology and constitutes about 2.5 to 5 % of all small cell carcinomas\(^4\). It is more common in males, if cervix and gallbladder is excluded and generally affects patients older 70 years\(^6\).

Gastrointestinal and genitourinary tracts are the most common sites\(^11\). However, literature reveals a variety of distinct locations and causing so different clinical pictures. Esophagus, stomach, duodenum, liver, pancreas, common bile duct, rectum, colon, bladder, trachea, larynx, pharynx,
hypopharynx, mouth floor, sino-nasal, heart, mediastinum, pleura, prostate, ovary, bone, retroperitoneal locations, even more appendix have been reported in the literature\(^{(3,6,11)}\). Also, a different location in the external auditory canal causing facial nerve paralysis was also reported\(^{(10)}\). Although so different locations have been reported, there is only one indicating spine involvement only with back pain\(^{(9)}\). There are some differences between this case and ours. Our case was a young non-smoker woman presented with the symptoms of upper motor neuron involvement, however that case was a old woman heavy smoker presented only with back pain.

Optimal management of these lesions is still controversy\(^{(11)}\). Because of the limited data describing the optimal management and poor prognosis of these lesions, combined treatment modalities consisting of chemotherapy, radiotherapy, and/or surgery have been used\(^{(11)}\). The most common used chemotherapeutic regimen is the combination of the etoposide and cisplatin\(^{(6)}\). In management of our case, we thought to perform total excision of mass because of the evidence of progressive upper motor neuron symptoms, easy accessibility of the mass, and younger age of the patient. After learning the definite diagnosis chemotherapy regimen consisting of etoposide, cisplatin and zoledronic acid was given.

In conclusion, this case is of importance reporting the second ESCC arising from spine. Also, our case is of importance being presented with upper motor neuron involvement differing from literature. Although increasing reports of extrapulmonary small cell carcinoma cases, their management is still in debate. New diagnostic techniques will aid in the early diagnosis and the management of these tumors.

**Correspondence to:**
Hakan Ak
E-mail: nrsdrhakanak@yahoo.com

**Received by:** 08 March 2011
**Revised by:** 16 September 2011
**Accepted:** 24 September 2011

**The Online Journal of Neurological Sciences (Turkish) 1984-2012**
This e-journal is run by Ege University Faculty of Medicine, Dept. of Neurological Surgery, Bornova, Izmir-35100TR
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URL: http://www.jns.dergisi.org
Journal of Neurological Sciences (Turkish)
Abbr: J. Neurol. Sci.[Turk]
ISSN: 1302-1664

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