



Case Report

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

Hakan AK, Varol AYDIN

Adnan Menderes Üniversitesi, Nöroşirurji Anabilimdalı, 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 regiment 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, epidural mass

Torasik Epidural Kitle ile Prezente Olan Omurganın Akciğer-Dışı Küçük Hücreli Kanseri: Olgu Sunumu

Özet

Nadir görülen akciğer dışı küçük hücreli karsinomlar akciğerin küçük hücreli karsinomu ile aynı histolojik özellikleri paylaşmaktadırlar, bununla birlikte bunlar genellikle ayrı bir antite olarak kabul edilmişlerdir. Bu tümörler daha çok erkeklerde görülmekte, genitoüriner ve gastrointestinal sistemler en sık tutulma gösteren yerlerdir. Literatürde farklı tutulum yerleri bildirilmekle birlikte omurgadan kaynaklanan küçük hücreli karsinom bildiren tek bir olgu sunumu mevcuttur. Bu yazıda 35 yaşında bir kadında omurgadan kaynaklanan ve epidural kitle kliniği ile başvuran akciğer dışı küçük hücreli karsinom vakası sunulmaktadır. Hastanın kesin tanısı ancak patolojik değerlendirilmesi ile konulabilmiş olup tanının kesinleşmesinin ardından hastaya etoposid, cisplatin ve zoledronic acid'den oluşan kemoterapi rejimi uygulanmıştır. Bu tümörlere daha erken tanı konulabilmesi ve bunların en iyi şartlarda tedavisi için daha ileri çalışmalara gereksinim vardır.

Anahtar Kelimeler: Omurga, küçük hücreli kanser, insidans, epidural kitle

INTRODUCTION

Extrapulmonary small cell carcinomas (EPSCC) are the tumors having the histological appearance of small cell carcinoma without evidence of any thoracic pathology⁽²⁾. Their incidence is very low and generally accepted as a distinct clinicopathologic entity⁽²⁾. ESCC

originating from different organs have been reported in the literature^(1,5,7,9,10), however, as far as we aware, this is the second ESCC originating from spine⁽⁹⁾.

In this report we are presenting an ESCC case in a 35 year old woman presenting with a thoracic epidural mass. Her definite diagnosis was performed after surgery and

chemotherapeutic regiment consisting of etoposide, cisplatin and zoledronic acid was given.

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 regiment consisting of etoposide, cisplatin and zoledronic acid was started.



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

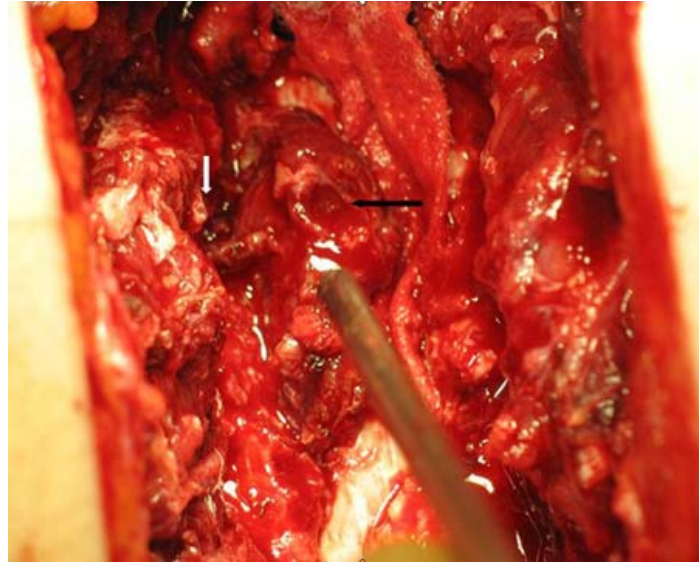


Figure 2: An intraoperative image. White arrow indicating the mass arising from the foramina of the third thoracic vertebrae. Black arrow indicating the epidural component of the mass.

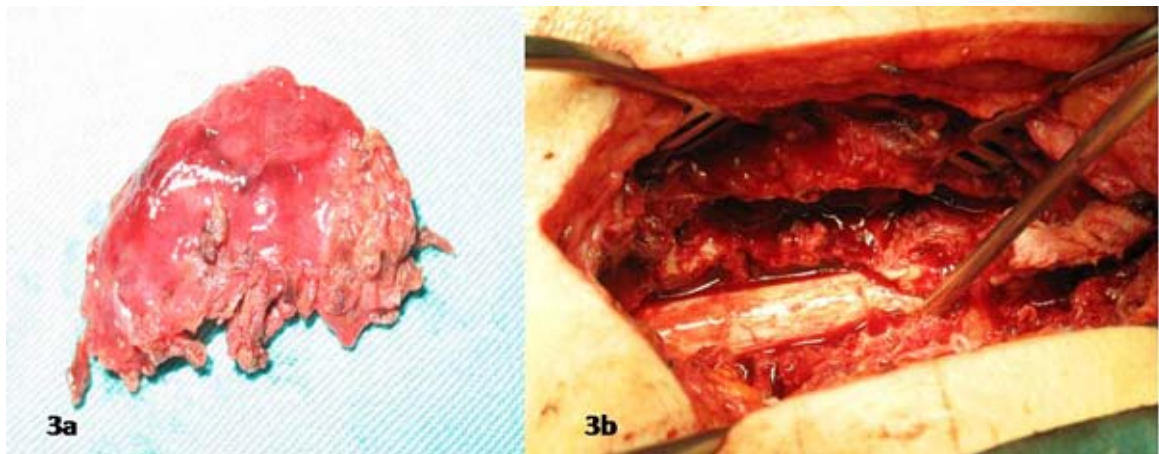


Figure 3a&b: Figure 3a demonstrating totally removed epidural mass and 3b demonstrating intact structure of the dura.

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 regiment 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 regiment 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

as part of the Ege Neurological Surgery World Wide Web service.

Comments and feedback:

E-mail: editor@jns.dergisi.org

URL: <http://www.jns.dergisi.org>

Journal of Neurological Sciences (Turkish)

Abbr: J. Neurol. Sci.[Turk]

ISSNe 1302-1664

REFERENCES

1. Gaba A, Mbaoma R, Breining D, Smith RV, Beitler JJ, Haigentz M: Unusual sites of malignancies. *Journal of Clinical Oncology*. 2005; 23; 2094-96.
2. Galanis E, Frytak S, Lloyd RV. Extrapulmonary small cell carcinoma. *Cancer* 1997; 79;1729-36.
3. Haider K, Shahid RK, Finch D, Sami A, Ahmad I, Yadav S, Alvi R, Popkin D, Ahmed S. Extrapulmonary small cell cancer: A Canadian province\'s experience. *Cancer* 2006; 107; 2262-9.
4. Henricus FM, Heijden van der, Heijdra Y. Extrapulmonary small carcinoma. *Southern Medical Journal*. 2005, 98: 345-9.
5. Iwamuro M, Tanaka S, Bessho A, Takahashi H, Ohta T, Takada R, Murakami I. Two cases of primary small cell carcinoma of the stomach. *Acta Med. Okayama* 2009; 63;293-8.
6. Lee SS, Lee JL, Ryu MH, Chang HM, Kim TW, Kim WK, Lee JS, Jang S-J, Khang SK, Kang Y-K. Extrapulmonary small cell carcinoma: Single center experience with 61 patients. *Acta Oncol*. 2007; 46; 846-51.
7. Moskal TM, Zhang PJ, Nava HR: Small cell carcinoma of the gallbladder. *Journal of Surgical Oncology*. 1999; 70;54-9.
8. O\'Kane AM, O\'Dannel M, Shah R, Carey DP, Lee J. Small cell carcinoma of the appendix. *World*

- Journal of Surgical Oncology* 6:4, 2008.
doi:10.1186/1477-7819-6-4.
9. Raina V, Milroy R, Al-Dawoud A, Dunlop D, Soukop M. Extrapulmonary small cell carcinoma of bone. *Postgrad Med J.* 1992; 68; 147-8.
 10. Rudman KL, King E, Poetker DM. Extrapulmonary small cell carcinoma metastasis to the external auditory canal with facial nerve paralysis. *American Journal of Otolaryngology-Head and Neck Medicine and Surgery* xx: xxx-xxx,2010.
doi:10.1016/j.amjoto.2010.03.007
 11. Wong YNS, Jack RH, Mak V, Henrik M, Davies EA. The epidemiology and survival of extrapulmonary small cell carcinoma in South East England. *BMC Cancer.* 2009; 9: 1970-2004.