

UC Davis
Dermatology Online Journal

Title

Cutaneous metastasis of primitive neuroectodermal lung tumor

Permalink

<https://escholarship.org/uc/item/50p902r0>

Journal

Dermatology Online Journal, 19(6)

Authors

Garcia Romero, Diana
Hilara Sanchez, Yolanda
Perez Alvarez, Javier
et al.

Publication Date

2013

DOI

10.5070/D3196018567

Copyright Information

Copyright 2013 by the author(s). This work is made available under the terms of a Creative Commons Attribution-NonCommercial-NoDerivatives License, available at <https://creativecommons.org/licenses/by-nc-nd/4.0/>

Peer reviewed

Case presentation

Cutaneous metastasis of primitive neuroectodermal lung tumor

Diana Garcia Romero PhD, Yolanda Hilara Sanchez, Javier Perez Alvarez, Jose Ramon Ramirez Garcia, Maria Pilar de Pable Martin

Dermatology Online Journal 19 (6): 10

Hospital del Tajo, Aranjuez, Madrid, Spain

Correspondence:

Diana Garcia Romero PhD: dianadic78@yahoo.es

Abstract

Primary sarcomas of the chest are rare. Although primitive neuroectodermal tumor (PNET) usually develops in the chest wall, it has been described as a primary pulmonary tumor. We present an unusual case of PNET arising in the lung of an 89-year-old man.

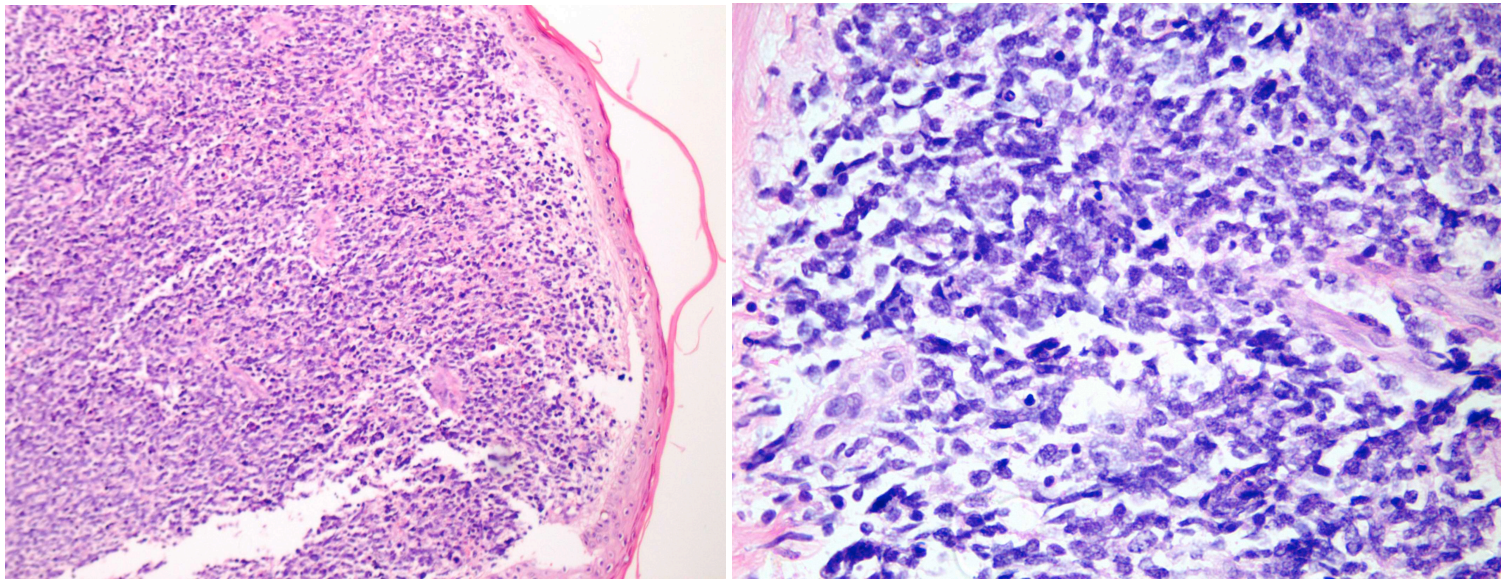
Case Synopsis

An 89-year-old man, non-smoker and well-controlled diabetic, was referred to our department because of the appearance of a mass on the scalp exhibiting progressive growth for two months. Cutaneous examination showed a large and fungating mass on the head, which had an unpleasant aroma (Figure 1). The family had noticed a gradual decline in the patient's function in recent months. Three days after presentation, he went to the emergency department because of hemoptysis and epigastric pain.



Figure 1. Fungating scalp tumor

A biopsy of the skin confirmed the diagnosis of metastatic type neuroectodermal tumor, which was enolase and CD99 positive, synaptophysin and chromogranin negative (Figures 2,3).



Figures 2, 3

The chest radiograph showed an increased density in the lower third of the right hemithorax with a well-defined nodular image. Computed tomography showed a voluminous mass of 9cm x 10cm x 12cm, compatible with primary lung cancer. Likely additional metastases were seen in the right apex and left upper lobe and in the subcutaneous tissue. Additional lesions were noted in the skull, in the right parieto-occipital region, and in multiple lymph nodes in the right hilum and subcarinal regions. There was a right adrenal nodule of 2cm and a lytic bone lesion at L4. Laboratory studies showed elevation of acute phase reactants such as platelets, fibrinogen, reactive C protein, and lactate dehydrogenase. The patient also had hypochromic anemia, hypoalbuminemia, and increased gamma-glutamyltransferase. We performed a complementary study of tumor markers, which showed significant elevation of neuron-specific enolase. Owing to the diagnosis of primitive neuroectodermal tumor of the lung, stage IV, with lymphatic, bone, adrenal, lung, and soft tissue metastasis and given the age of the patient, palliative treatment at home was chosen.

Discussion

Cutaneous metastasis occurs in 0.7% to 9% of cancer patients and the incidence of cutaneous metastasis in patients with lung cancer varies between 2.8% and 8.7%. Lung cancer metastases are most frequently found on the head and neck, most commonly in the form of nodules [1]. This is generally considered a rare and delayed phenomenon in the course of most tumors, although in some cases metastases may be the first signs of internal malignancy. Clinically, they manifest as nodules, ulcers, cellulitis-like lesions, bullae, or fibrotic processes [2]. The differential diagnoses considered clinically, along with metastatic lung cancer, were squamous-cell carcinoma, basal-cell carcinoma, amelanotic melanoma, carcinoid tumour, Merkel cell carcinoma, neuroendocrine carcinoma, malignant histiocytoma, atypical fibroxanthoma, and dermatofibrosarcoma protuberans [2, 3]. Cutaneous metastases may occur through three mechanisms: lymphatic spread, intravascular dissemination, and direct extension. The high vascularity of the scalp and lack of valves in the vertebral venous system may predispose to scalp metastases [3].

PNET is very rare neoplasm with neuronal differentiation that affects principally the deep soft tissues. This tumor is extremely aggressive, which frequently results in metastasis to various organs [4]. To the best of our knowledge, about ten cases of primary pulmonary PNET have been reported in the literature. The PNET family shares common histological features of closely packed small primitive round cell tumors. It most often arises in the soft tissues and bones but has rarely been reported in other sites, such as the ovary, testis, uterus, kidney, palate, pancreas, myocardium, and lung [5-11]. No evidence of metastasis has been described in any of them. Morphological features of intrapulmonary tumors are similar to those of the PNET in a variety of other locations.

In our case the patient was 89 years old, which was an unusual age compared with previously reported PNET cases. Most of them were found in children and younger adults, a range of 15 to 64 years, with a mean age of 33 years [8-10]. One case described in the literature was 67 years of age [11]. The treatment of PNET is aggressive; the most effective treatment is surgical resection with combination chemotherapy and high-dose radiation therapy [9]. One of these cases of PNET died after surgery because of pneumonectomy [9].

References

1. Nikolaos Barbetakis, Georgios Samanidis, Dimitrios Paliouras, Elpida Samanidou, Zoi Tzimirota, Christos Asteriou, Persefoni Xirou and Christodoulos Tsilikas. Facial skin metastasis due to small-cell lung cancer: a case report. *Journal of Medical Case Reports* 2009, 3:32; PMID: 19178730. <http://www.jmedicalcasereports.com/content/3/1/32>
2. Marcoval J, Gallego MI and Moreno A. Inflammatory Cutaneous Metastasis as a First Sign of Recurrence of Squamous Cell Carcinoma of the Lung. *Actas Dermosifiliogr.* 2008;99:157-69. PMID: 18346442
3. Tadahiko Kubo, Shoji Shimose, Toshihiro Matsuo, Koji Arihiro and Mitsuo Ochi. Scalp metastasis from malignant fibrous histiocytoma of bone. *J Am Acad Dermatol* 2008;59:S88-91. PMID: 19119134
4. Leila Antonangelo, Adriana Gonçalves Rosa, Aline Pivetta Corá, Milena Marques Pagliarelli Acencio, Luís César Moreira, Francisco Vargas Suso. Uncommon pleural effusion: pleuropulmonary metastasis from primitive neuroectodermal tumor. *J Bras Pneumol.* 2009;35(6):606-609. PMID: 19618040
5. Koo HL, Jun SY, Choi G, Ro JY, Ahn H, Cho KJ. Primary Primitive neuroectodermal tumor of the kidney: report of two cases. *Korean. J Pathol* 2003; 37: 145-9.
6. Kang MS, Yoon HK, Choi JB, Eum JW. Extraskkeletal Ewing's sarcoma of the hard palate. *J Korean Med Sci* 2005; 20: 687-90. PMID:16100468
7. Danner DB, Hruban RH, Pitt HA, Hayashi R, Griffin CA, Perlman EJ. Primitive neuroectodermal tumor arising in the pancreas. *Pathol* 1994; 7: 200-4. PMID: 7911997
8. Imamura F, Funakoshi T, Nakamura S, Mano M, Kodama K, Horai T Primary primitive neuroectodermal tumor of the lung: report of two cases. *Lung Cancer* 2000; 27: 55-60. PMID: 10672784
9. Kahn AG, Avagnina A, Nazar J, Elsner B. Primitive neuroectodermal tumor of the lung. *Arch Pathol Lab Med* 2001; 125: 397-9. PMID: 11231490
10. Tsuji S, Hisaoka M, Morimitsu Y, Hashimoto H, Jimi A, Watanabe J, Eguchi H, Kaneko Y. Peripheral primitive neuroectodermal tumour of the lung: report of two cases. *Histopathology* 1998; 33: 369-74. PMID: 9822928
11. Yoon Young Lee, Do Hoon Kim, Ji Hye Lee, Jong Sang Choi, Kwang Ho In, Yu Whan Oh, Kyung Hwan Cho, Yong Kyun Roh. Primary Pulmonary Ewing's Sarcoma/Primitive. Neuroectodermal Tumor in a 67-year-old Man. *Korean Med Sci* 2007; 22 (Suppl): S159-63. PMID: 17923745