Case Report

Research on Chronic Diseases

Neurofibromatosis Emphysema and Interstitial Lung Disease

Abstract

Lung cancer associated with neurofibromatosis type I is considered veritably rare, and only a many case reports have been described in the literature. There's some substantiation that a inheritable relation between neurofibromatosis and carcinogenesis in the lung may live. We present a 42- time-old lady, continuance nonsmoker with a given history of neurofibromatosis type I, free of respiratory symptoms, which passed a low- cure HRCT of the lungs to probe any occult interstitial lung changes. A solitary ill- defined bump of ground- glass nebulosity was detected apropos in the middle lobe with no associated lymphadenopathy or metastatic complaint. Several thin- walled lung excrescencies were also seen in the lower lobes. Histological analysis of the bump after middle lobectomy revealed well- discerned adenocarcinoma. The case didn't admit systemic chemotherapy or radiotherapy. She was free of complaint on 18- month follow up.

Liu Yongjun*

Department of Radiology, Democritus University of Thrace, Greece

*Author for correspondence:

Yongjunlinedu123@gmail.com

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Introduction

A 42- time-old lady, continuance nonsmoker with a given history of neurofibromatosis type I presented to the Outpatient Neurology Clinic for a regular follow- up. Physical examination revealed multiple cafe au- lait spots ′ and cutaneous neurofibromas of variable size throughout her body (**Figure 1**). She also suffered from vitiligo since the age of 20 times old. Her neurologic examination was normal, and she was free of respiratory symptoms. She reported no history of environmental or occupational exposure to other implicit carcinogens for lung cancer [1]. still, as part of her thorough follow- up, she passed a low- cure HRCT of the lungs to probe any occult interstitial lung abnormalities and presence of lung excrescencies that have been described in neurofibromatosis cases. HRCT of the lungs detected an incidental ill- defined solitary pulmonary bump1.4 cm in periphery, flaunting a ground- glass nebulosity and air alveologram (**Figure 2**). No other lung nodes or areas of connection were seen in any Case donation. A 42- time-old lady, continuance nonsmoker with a given history of neurofibromatosis presented to the

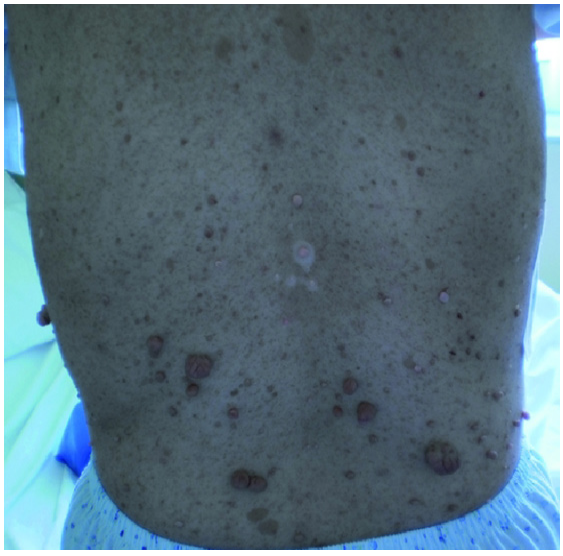

Figure 1: Multiple café-au-lait spots and cutaneous neurofibromas of variable size were noted throughout the patient's body.

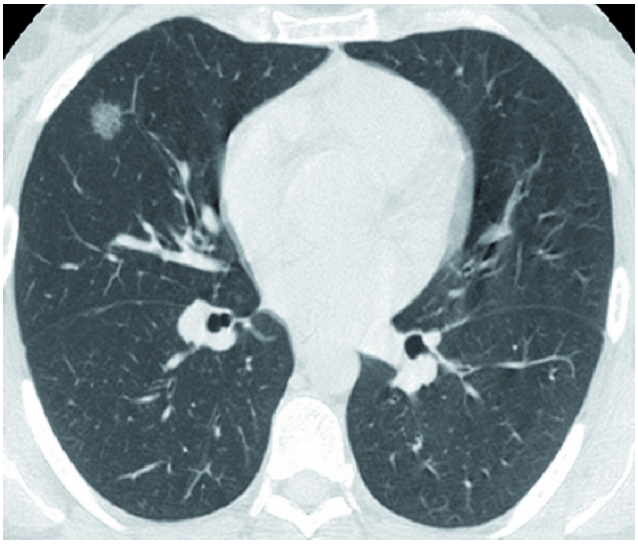

Figure 2: HRCT at the level of the middle lobe shows a 1.4 cm ill-defined nodule of a ground-glass opacity in the right middle lobe demonstrating air alveologram**.**

Outpatient Neurology Clinic for a regular follow up [2]. Physical examination revealed multiple cafe au- lait spots ′ and cutaneous neurofibromas of variable size throughout her body (**Figure 1**). She also suffered from vitiligo since the age of 20 times old. Her neurologic examination was normal, and she was free of respiratory symptoms [3].

She reported no history of environmental or occupational exposure to other implicit carcinogens for lung cancer. still, as part of her thorough follow up, she passed a low- cure HRCT of the lungs to probe any occult interstitial lung abnormalities and presence of lung excrescencies that have been described in neurofibromatosis cases [4,5]. HRCT of the lungs detected an incidental ill- defined solitary pulmonary bump1.4 cm in periphery, flaunting a ground-glass nebulosity and air alveologram (**Figure 2**) [6]. No other lung nodes or areas of connection were seen in any Discussion Neurofibromatosis type 1 or von Recklinghausen's complaint is an autosomal dominant dysplasia of the ectoderm and mesoderm characterized substantially by the presence of neurofibromas, cafe- au- lait spots, and painted hamartomas ′ in the iris. The NF- 1 gene has been localized to chromosome 17q11 and functions as excrescence suppressor gene, and its separate gene product has been named as neurofibromin. It has been suspected that the mutation of excrescence suppressor NF- 1 gene increases the case's threat for the development of colorful malice substantially deduced from the neural crest similar as nasty schwannoma, neurofibrosarcoma, intracranial glioma, and pheochromocytoma [7,8].

Still, the association of NF- 1 with lung cancer isn't common. A review from Japan 20 times ago reported only 11 cases of NF- 1 with lung cancer, and we interestingly were suitable to find 7 further cases up to now- without including the presented case — also from the Japanese literature [9]. Adenocarcinoma was the most frequent histological opinion(72.9) in the first 11 cases reviewed, while the following 6 cases were diagnosed as no small cell cancer(3 cases), small cell cancer(1 case), inadequately discerned cancer (1 case), adenocarcinoma (1 case), and carcinosarcoma (1 case). Smoking habit wasn't always mentioned in the reported cases in the literature, but there were some cases that were no way smokers as in our case [10,11]. In our case, histology revealed a well-discerned adenocarcinoma, which is also the most common histological type of lung cancer in nonsmokers, who are by far more common in women. The case in the presented case was a continuance nonsmoker and so was her hubby. Since there's no association with smoking in the presented case, a question raises whether there's a inheritable relation between NF- 1 and the development of lung cancer [12]. NF- 1 gene has been counterplotted in the small part of chromosome 17q. p53 gene belongs to this area and has been intertwined in the development of colorful tumors including lung cancer. Also, in one study, it has been reported that the loss of heterozygosity in chromosome 17p — but not in 17q — was detected in a case with small cell melanoma with NF- 1 [13.14].

Conclusion

The authors hypothecated that the inactivation of excrescence suppressor gene on chromosome 17p — most likely p53 — might have been responsible for the development of small cell lung cancer in that case. Still, the association of lung cancer with NF- 1 might be coincidental. The bump in our case was of pure ground-glass nebulosity which is known to have a high probability of malice compared to "solid nodes" and represents the " Broncho alveolar " element of lung adenocarcinoma indicating a better prognostic of lung adenocarcinoma. It remains to be answered whether the development of lung cancer in this youthful, no way - smoker, womanish case with NF- 1 simply followed the rules of the epidemiology of lung cancer in no way - smokers or there's a inheritable link between NF- 1 and carcinogenesis in the lung. Large inheritable- grounded studies are demanded in order to probe these interesting findings. The evaluation of pulmonary involvement in NF- 1 cases with low- cure casket CT in long intervals could be proposed in order to descry early development of lung cancer or interstitial lung complaint.

Acknowledgement

None

Conflict of Interest

None

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