Poland Syndrome Associated with Lung Cancer: A Case Report and Literature Review

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Case report

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Abstract

Background: Poland syndrome is a rare congenital disease consisting a spectrum of defects occupied by unilateral chest wall hypoplasia and ipsilateral hand abnormalities. It is also closely associated with some malignant diseases and some benign diseases, such as breast cancer, lymphoma, recurrent spontaneous pneumothorax, perforating dermatosis, hemangioma, gastric cancer and lung cancer. The low incidence of Poland syndrome has hindered investigators' attempts to develop randomized prospective trials, so the disease is poorly understood. Now We find out a case of Poland's syndrome associated with adenocarcinoma of right lung, in which before this article there was only article depicting a case of lung squamous cancer with poland syndrome, this is an interesting topic. To the best of our knowledge, this is the second case of poland syndrome closely related to lung cancer.

Case presentation: A 61-year-old man came to the hospital accompanied by intermittent dry cough and hemoptysis that lasted for 2 months. Chest CT revealed showed a large lobulated mass about 5.5cm × 4.0 cm × 3.0 cm in the right lower lung field and the normal right pectoralis major muscle and pectoralis minor were both absent. we diagnosed the patient with stage IIIa lung adenocarcinoma. After two cycles of preoperative platinum-doublet chemotherapy, we have performed a regular right lower lobectomy and lymph node dissection. Until now, after 2.5 years of follow up, he did not relapse.

Conclusions: In addition to some other reported types of malignancies, there is increasing evidence that an association between these two clinical entities. Although we do not understand the underlying mechanisms, we assume there exists some common gene pathways that regulate these two clinical entities. Because these patients who got Poland syndrome are susceptibility to different kinds of malignancies, longitudinal epidemiological studies may be needed.

Background

Poland Syndrome is an uncommon congenital disorder which often occurs sporadically but few occasional familial cases have been founded in china. It has an incidence of approximately 1:7000 to 1:100000 live births and associated features may be greatly variable from slight to severe[1, 2]. It tends to occur with a higher frequency the right hemithorax compared with the left cases described. This syndrome is usually characterized by the absence of the pectoralis major and minor muscles, upper limb anomalies, hand anomalies, costal cartilage defects and some other abnormalities in the chest area[3]. A thorough history is important in establishing the specific diagnosis of Poland Syndrome. Clinical features of Poland Syndrome are often variable. Simultaneously, mild or even undiscovered functional impairment manifestations in children are always undiagnosed or misdiagnosed[4]. The pathogenic mechanisms underlying Poland's syndrome are still unknown. The most commonly accepted causal mechanism is that vascular developmental abnormality happens in the sixth week of embryogenesis, this situation leads to a reduced amount of blood supply delivered to nourish the developing tissues. Surgical correction is always available in those persons whose significant functions are severely impaired [5, 6].

However, various malignancies closely related to Poland Syndrome may also be the major focus of this disease. Up to date, Some malignancies and benign diseases coupled with Poland's syndrome have been screened in English-language literature. In 2000, Ahn has reported a right lung squamous cancer patient accompanied with Poland's syndrome because of intermittent cough and blood-tinged sputum[7]. Now we introduce a case of lung adenocarcinoma associated with Poland syndrome, which has not yet clearly been reported, and demonstrate the treatment course.

Case Presentation

A 61-year-old man came to the hospital accompanied by intermittent dry cough and hemoptysis that lasted for 2 months. He had smoked about 10 cigarettes per day for about 10 years, but had quit smoking 10 years ago. He was a factory worker and had been living in the suburbs. He had no family history of lung cancer. This patient had complained that absence of his right chest wall muscles as well as involvement of his fingers on the same side since his childhood, but he never went to see the doctor. On physical examination, his chest wall appeared apparently asymmetric. He had no anterior axillary fold, no right pectoralis major muscle and deficiency of the two fingers on his right hand. Chest CT revealed (Fig. a) showed a large lobulated mass in the right lower lung field. We also can see the normal right pectoralis major muscle and pectoralis minor muscle were both absent, accompanied by an unusually position of the right axillary line. Right ribs were also a little slightly hypoplastic. Bronchoscopic examination revealed a tumor in the right upper bronchus. Based on the result of percutaneous needle biopsy, we diagnosed the patient with stage IIIa lung adenocarcinoma mainly because of large size. Then after two cycles of preoperative platinum-doublet chemotherapy, this tumor had shrunked obviously, meanwhile metastatic lymph nodes(N1 lymph nodes) had gone missing(Fig.b).In consideration of excellent chemotherapy result, we have performed a regular right lower lobectomy and lymph node dissection. The pathological specimen confirmed lung adenocarcinoma, with metastasis to only one lymph node of the subcarinal lymph node (1 of 15).Then he received two-cycles adjuvant chemotherapy after the operation. After 2.5 years of follow up, he did not relapse. This patient referred to thoracic oncology clinic for treatment of malignant neoplasm was tested for their immediate recall of information presented and signed on informed consent document.

Discussion

Up to now, there are several English literatures clearly depicting Poland's syndrome are closely related to some malignancy disease, including breast cancer, gastric cancer, lymphoma, lung cancer, some rare lymphoproliferative and hematopoietic malignancies[8].

In 2000, Ahn has firstly reported a right lung squamous cancer patient accompanied with Poland syndrome, he thought Poland syndrome may represent an example of a genetic disorder with increased risk of lung cancer[7]. Unlike his case, this patient had got lung adenocarcinoma. As we all know, adenocarcinoma accounts for approximately 60% and squamous cell carcinoma for approximately 20%

of all NSCLC cases. They also share totally different molecular profiles. We carried out a systematic review on Pubmed and Embase databases, our case is the first lung adenocarcinoma to be reported.

Although there have been some scientific updates in the treatment, molecular subtyping of Poland syndrome has almost not changed much since Alfred Poland first described the syndrome in 1841[9]. Poland syndrome is believed to be caused by some genetic changes that can be passed down in families, however no related genes have been identified. Michele Torre hypothesized that a large deletion of chromosome 11q12.3 or 6q21-q22.1 is closely related to Poland syndrome[10]. Cohen firmly believed that there are some postzygotic mutations in b-actin[11].

This case presented here reinforce the hypothesis of a connection between the two clinical entities. The most commonly accepted causal mechanism is mainly due to the disruption in the blood supply of the brachial arch during embryogenesis[12]. This case is right side lung cancer coinciding with right pectoralis defects on the same side, they are mainly due to the disruption in the blood supply, then lacking of the blood supply gradually affects the development of the lung, thus predisposing the risk of lung cancer[3, 13, 14]. Meanwhile, cigarette smoking maybe an additional risk factor for lung cancer, although this person had already quited smoking habits for 10 years.

We postulated thatthere existed an undiscovered mutant gene which regulates the occurence of lung cancer and Poland syndrome. Under some specific circumstances,

they are susceptibility to cancer. This case supports the theory that the morphological changes observed in Poland syndrome can also extend to the pulmonary parenchyma. with the help of some adverse habits such as smoking, they are more vulnerable to lung cancer. Hence these persons may benefit from greater surveillance.

Conclusions

Hence, due to the existence of a spectrum of defects and possibilities of suffering from multiple related diseases, surgeons should pay special attention to these patients. We need more lung cancer cases with Poland's syndrome to interpret the possible altered genes between them. Also, long time epidemiological studies may be beneficial.

Declarations

Ethics approval and consent to participate

Our case involved human, who had signed informed consent. This study was designed in accordance with the Declaration of Helsinki and approved by the ethics committee of Huadong Hospital affiliated to FuDan university.

Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the consent is available with editorial office.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Yuanshan Yao and Wen Gao are surgeons of the Surgical Oncology and have performed the operation. All authors read and approved the final manuscript.

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Availability of data and material

All data analyzed during this study are included in this published article.

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Figures

Figure 1

A 61-year-old man with Poland's syndrome associated with lung adenocarcinoma. Chest radiograph shows the absence of the right pectoralis major muscle and pectoralis minor muscle, Right ribs were a little slightly hypoplastic. A large tumor (lung adenocarcinoma) was located in the middle of right lower lobe and enlarged lymphnodes can be seen in the surrounding of the bronchus of right lower lobe.

Figure 2

CT at the same level of the right inferior pulmonary vein demonstrates the mass had dramatically shrunk and enlarged lymphnodes had already become invisible(nearly complete remission).After a right lower lobectomy and lymph node dissection,this patient recovered well and he did not relapse until now. The pathological specimen found a little residual lung adenocarcinoma cell, with metastasis to only one lymph node of the subcarinal lymph node(yoT3N1M0).