A Misdiagnosis of a Huge Lung Fibroleiomyomatous Hamartoma in Pleural Cavity

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Case report

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Abstract

Backgroud: Lung fibroleiomyomatous hamartoma is an extremely rare benign tumor in lung, it belongs to hamartoma. There were only five patients with solitary pulmonary fibroleiomyomatous hamartoma reported in literatures and our case has the largest size of the tumor from lung.

Case presentation: Our case is a 36-year woman with a huge mass in pleural cavity. She was once misdiagnosed mediastinal germ cell tumor with pleural effusion through percutaneous lung biopsy in other hospital. After traditional thoracotomy, a huge size of 22 x 18 x 5.5 cm mass was completely dissected and a diagnosis of pulmonary fibroleiomyomatous hamartoma was given by pathology combined with immunohistochemical staining, which showed the positive staining of SMA, Desmin, CD34, BCL2, TTF1, Napsin A, and CK7, and the negative staining of S-100 and Calretinin. The patient recovered well with 40 months following up without recurrence.

Conclusions: In our case, we had reported the largest size of lung fibroleiomyomatous hamartoma. The treatment of fibroleiomyomatous hamartoma is completely resected it and have a good prognosis.

Background

Lung fibroleiomyomatous hamartoma, a rare benign tumor, belongs to hamartoma. A coin-shaped fibroleiomyomatous hamartoma had first been resected by Cruickshank and then first defined it as “diffuse fibroleiomyomatous hamartomatosis” in 1953 [1]. It is an extremely rare type of benign tumor could be found in every sites of lung. So far, it is only five patients with solitary pulmonary fibroleiomyomatous hamartoma had been reported [2]. The histological diagnosis criteria are “a well-circumscribed nodule composed of smooth muscle cells with numerous epithelial inclusions” and “no cellular atypia or mitotic figures” [2]. While in this present clinical case we showed a huge lung fibroleiomyomatous hamartoma found in pleural cavity, which was first misdiagnosed as germ cell tumor with pleural effusion considered as pleural metastasis. Therefore, this is a very rare clinical case with such huge lung fibroleiomyomatous hamartoma.

Case Presentation

A 36-year-old woman with came to our hospital for chest tightness and suffocation. She did not have cough, hemoptysis, dyspnea, chest pain or other pulmonary symptoms. The patient also did not have the history of uterine leiomyomas and smoking. The chest CT examination showed there was a huge tumor in the left pleural cavity (Fig. 1A), and the patient was given a diagnosis of mediastinal germ cell tumor with pleural effusion through percutaneous lung biopsy in other hospital. After hospitalization, tumor markers and physical examination were both negative. The rest detections for distance metastasis with brain MRI, bone scanning, and upper abdominal enhanced CT were all shown negative.

Through the discussion of Multidisciplinary Comprehensive Treatment (MDT), we believe that there is a possibility of completely surgical resection, and the chest fluid has a complete envelope, which should be

removed completely too. Then the patient accepted a traditional thoracotomy combined with thoracoscopy. During the operation, the mass had a completed envelop with two linked parts, one is solid part and another is full bloody fluid. There are only three sites of the mass connected to the left lung and an arterial vessel from a descending aortic arch enters the tumor. The rest of the mass are free and the operation is not so hard to finish. As shown in Fig. 1A, the completely resected tumor with a huge size of 22 x 18 x 5.5 cm was rendered. Furthermore, as shown in Fig. 2, a final diagnosis of lung fibroleiomyomatous hamartoma was given by pathologist and the Immunohistochemical staining showed that the expressions of SMA, Desmin, CD34 and BCL2, TTF-1, Napsin A, and CK7 are positive, and the expressions of S-100 and Calretinin are negative, while the Ki-67 is in low expression. The patient recovered unevenly and was followed up every 6 months. After operation for 40 months, the patient lives well without local recurrence and distance metastasis.

Discussion

Pulmonary Hamartoma includes typical and atypical types. Typical hamartoma is a common tumor that contains smooth muscle tissue with numerous mesenchynal elements in lung, which is distinguished from atypical hamaroma with a predominant smooth-muscle component and many kinds of epithelial components[3]. Herein, we presented an atypical pulmonary hamartoma named fibroleiomyomatous hamartoma since it has a well-circumscribed nodule composed of smooth-muscle cells, more than one kind of epithelial inclusions and no cellular atypia or mitotic figures. Spencer had sum up three forms of hamartomatous musculofibrotic changes in the lung[4]. The first one is the smooth muscle tissue have single or numerous focal proliferation; The second one is pulmonary lymphangioleiomyomatosis and the third one is tuberous sclerosis with pulmonary changes. In the presented cases once misdiagnosed mediastinal germ cell tumor with pleural effusion through percutaneous lung biopsy in other hospital The Immunohistochemical staining results showed in the smooth muscle tissue and fibrous tissue smooth muscle actin (SMA), Desmin, CD34 and BCL2 are positive, S-100 negative, Ki-67 is in low expression. In cubic epithelium thyroid transcription factor-1 (TTF1), Napsin A, and CK7 are positive, Calretinin negative. Summarized the above results of Immunohistochemical staining, we thought diagnosis it as fibroleiomyomatous hamartoma is perfect.

In the present case, the yong woman was first misdiagnosed mediastinal germ cell tumor with pleural effusion through percutaneous lung biopsy in other hospital. The reason we think is that, first the lung fibroleiomyomatous hamartoma is rare,. To date, only six cases (including this case) have been reported and most tumor presented as incidental “coin lesions”. Due to the rare cases reported and the huge volume of this tumor, most doctor do not consider this diagnosis first. Second, lung fibroleiomyomatous hamartoma is common in women with uterine leiomyomas. Because many experts thought it is an metastasis tumor from benign leiomyoma or low-grade leiomyosarcoma especially in uterus[5]. But in this case, the patient have no history of uterine leimyomase. While according the situation, we think it should be the result of implantation and proliferation of benign smooth-muscle tissue, embolized by an intravenous leiomyomatosis or by mechanical means.[6]

The fibroleiomyomatous hamartoma thought to be a benign tumor. No other treatment should be done after surgery. The patient only should do regulation examination every half or one year. Hiroshi Itoh et al. had reported a patient had simple wedge excision had non-recurrent disease for about 10 years[5]. There are no reports of recurrence in the patients with pulmonaty fibroleiomyomatous hamartoma resection. We should emphasize that the treatment of this type of tumor called fibroleiomyomatous hamartoma had no treatment guidelines,but according previous case reports, the tumor need only remove from the patient's body and had regular review. That will be enough.

Conclusion

In this paper, we had reported a 36-year woman with a huge pulmonary fibroleiomyomatous hamartoma in pleural cavity. She was once misdiagnosed mediastinal germ cell tumor with pleural effusion through percutaneous lung biopsy in other hospital. Now the patient recovered well with 40 months following up without recurrence and distance metastasis. Though, the treatment of this type of tumor had no treatment guidelines, but according previous case reports, the tumor need only remove from the patient's body and had regular review.

Declarations

CONSENT

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. A copy of the consent form is available for review by the Editor-in-Chief of this journal.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

AUTHOR CONTRIBUTIONS

Minghui Liu, Xin Li and Jun Chen wrote this manuscript and analyzed all data. Minghui Liu, Xin Li, Fan Ren□Ming Dong, Chunqiu Xia and Hongbing Zhang provided medical care for the patients and collected the data. Jun Chen performed the operation and revised the article. All authors read and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

This case was approved by the ethics committee of our institution (Tianjin

Medical University General Hospital).

AVAILABILITY OF DATA AND MATERIALS

All available data are presented in the case.

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References

1. Cruickshank DB, Harrison GK. Diffuse fibro-leiomyomatous hamartomatosis of the lung. Thorax. 1953;8:316–8.
2. Ichiki Y, Kawasaki J, Hamatsu T, Suehiro T, Shibuya R, Matsuyama A, et al. A rare pulmonary hamartoma: fibroleiomyomatous hamartoma. Surg Case Rep. 2016;2:53. doi:10.1186/s40792-016-0184-z.
3. Ahmed S, Arshad A, Mador MJ. Endobronchial hamartoma; a rare structural cause of chronic cough. Respir Med Case Rep. 2017;22:224–7. doi:10.1016/j.rmcr.2017.08.019.
4. Spencer H. Pathology of the lung. Oxford, New York, Toronto, Sydney.
5. Paris F. Pergamon Press; 1977. p. 981–8.
6. Itoh H, Yanagi M, Setoyama T, Shirao K, Yanagi S, Kataoka H, et al. Solitary fibroleiomyomatous hamartoma of the lung in a patient without a pre-existing smooth-muscle tumor. Pathol Int. 2001;51:661–5.
7. Canzonieri V, D'Amore ESG, Bartoloni G, Piazza M, Blandamura S, Carbone A. Leiomyomatosis with vascular inva- sion. A unified pathogenesis regarding leiomyoma with vascular microinvasion, benign metastasizing leiomyoma and intra- venous leiomyomatosis. Virchows Arch. 1994;425:541–5.

Figures

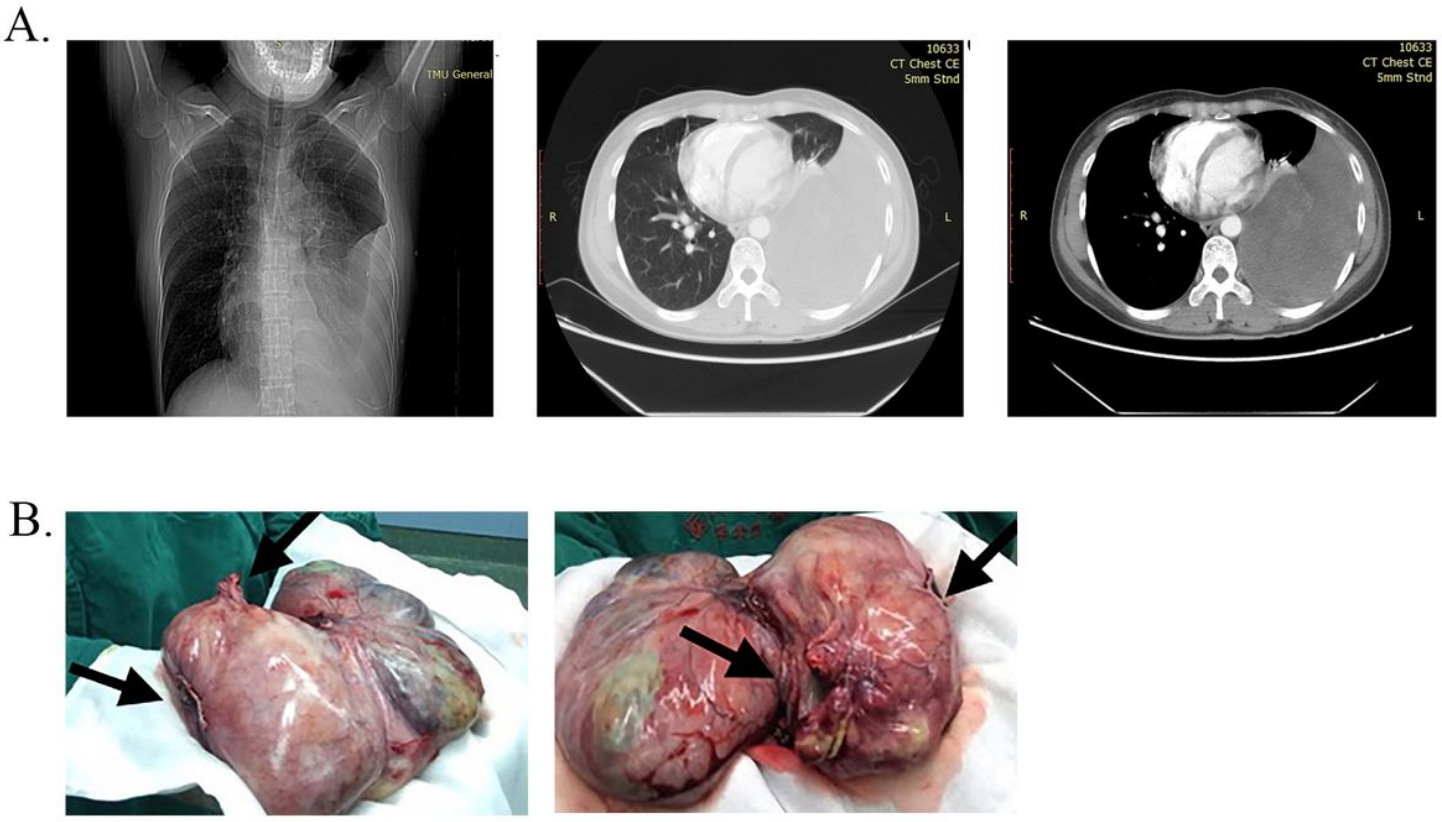

Figure 1

The patient's CT scan before surgery and solid tumor. (A) The CT scan of patient's tumor in the left pleural cavity. (B) Solid tumor after surgery. The size is 22 x 18 x 5.5 cm.

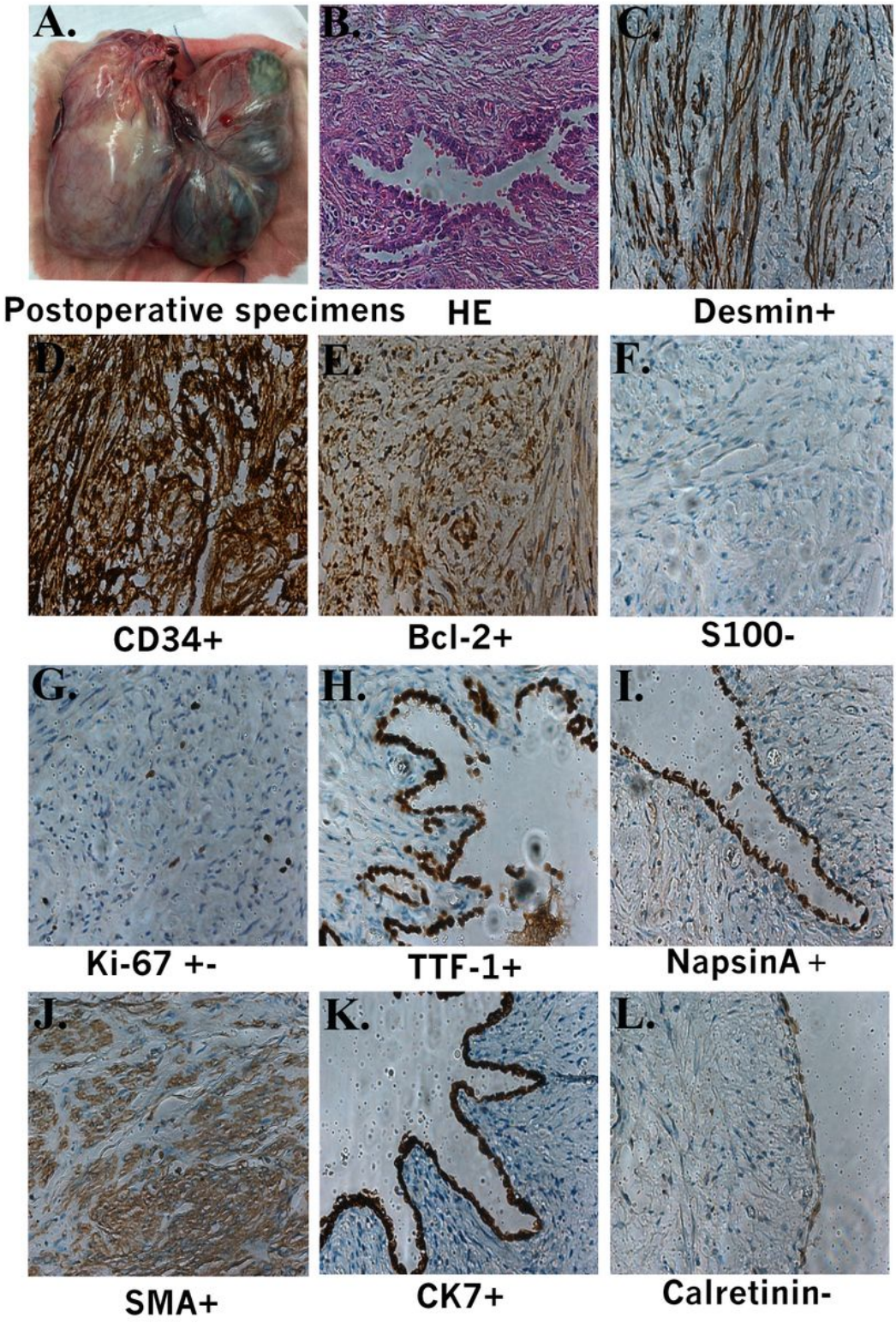

Figure 2

The IHC results of the huge tumor. Immunohistochemical staining (IHC) showed that the expressions of SMA, Desmin, CD34 and BCL2, TTF-1, Napsin A, and CK7 are positive, and the expressions of S-100 and Calretinin are negative, the Ki-67 is in low expression.