

CASE REPORT

Open Access



Left middle lobectomy for bronchiectasis in a patient with Kartagener syndrome: a case report

Haiping Lin, Ziang Cao, Xiaojing Zhao and Qing Ye*

Abstract

Background: Kartagener syndrome (KS) is a rare disorder characterized by the triad of chronic sinusitis, bronchiectasis, and situs inversus.

Case presentation: A 23-year-old man was admitted to our hospital because of recurrent cough with purulent expectoration, which had occurred intermittently for the past ten years. During the past 3 years, the episode frequency was 3–4 times per year. He was diagnosed with pulmonary infection and bronchiectasis of the left upper lobe, situs inversus, and KS. We concluded that the damaged left middle lobe was the source of repeat pulmonary infections. Thus the left middle lobe resection was performed to remove the source of the lung infection.

Conclusions: The post-operative course was successful and pneumonia was apparently resolved during the 6 months' follow-up period. We further describe this case in the following report.

Keywords: Kartagener syndrome, Bronchiectasis, Lobectomy

Background

Kartagener syndrome (KS) is a rare recessive autosomal disease with an incidence of approximately 1 in 32,000 live births [1, 2]. It is characterized by primary ciliary dyskinesia accompanied by sinusitis, bronchiectasis, and situs inversus [3]. Our case was diagnosed with synchronous Kartagener syndrome, pulmonary infection and bronchiectasis of the left upper lobe. Here, we reported the left middle lobectomy for a KS patient with bronchiectasis, which has been rarely reported in the literature.

Case presentation

A 23-year-old unmarried man was admitted to the Department of thoracic surgery of Renji hospital (Shanghai, China) because of recurrent cough with purulent expectoration, which had occurred intermittently for the past ten years. During the past 3 years, the episode frequency was 3–4 times per year, and an increased frequency occurred over the past 6 months. He had seen a respiratory physician many times and was diagnosed with pneumonia due to bronchiectasis. Additionally, he had received several

courses of intravenous antibiotic therapy over the past 6 months, however, frequent relapse occurred after antibiotic therapy. He had no fever, dyspnea and hemoptysis. Further questioning revealed that the patient used to suffer from sinusitis since his teenage years, presenting symptoms including repeated nasal congestion and nasal discharge. The patient acknowledged that he had been diagnosed with dextrocardia in childhood. He had no brothers or sisters, and his parents were healthy.

Yellowish discharge could be observed in the nasal cavity without obvious press-pain on paranasal sinus areas. The thoracic wall had no deformities. Chest auscultation revealed crepitations in the left infrascapular region and normal heart sounds on the right side. The apex beat was palpable over the fifth intercostal space on the right side of chest. The other physical and systemic examinations were normal.

Hemogram revealed a total leukocyte count of $11.0 \times 10^9/L$ (normal range: $3.97-9.15 \times 10^9/L$), a neutrophil percentage of 78.5% (normal range: 50–70%), and a hemoglobin of 150 g/L (normal range: 131–172 g/L). Liver functions and renal functions were normal, and the erythrocyte sedimentation rate was 40 mm/H. Sputum smear examination for *Mycobacterium tuberculosis*

* Correspondence: qingyeqyq@163.com
Department of Thoracic Surgery, Ren Ji hospital, School of Medicine, Shanghai Jiao Tong University, Shanghai 200127, China

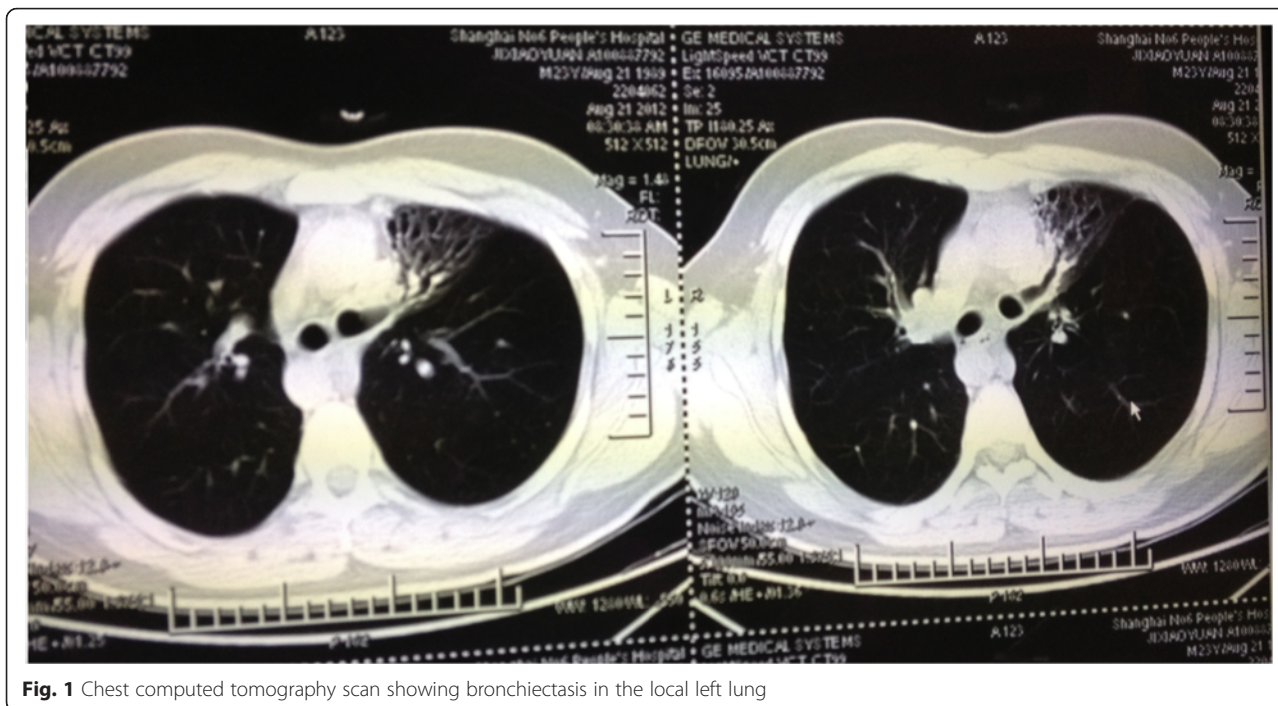


Fig. 1 Chest computed tomography scan showing bronchiectasis in the local left lung

was negative, and the blood tuberculosis antibody was also negative. Abdominal ultrasound revealed complete situs inversus and the organs were normal. Chest computed tomography (CT) scan revealed a bronchiectatic appearance in the local lung tissue of the left middle lobe (Fig. 1) and dextrocardia (Fig. 2). His admission diagnosis included pulmonary infection and bronchiectasis of the left middle lobe, situs inversus, and possible KS.

Paranasal sinus CT scan revealed that his nasal mucosa obviously thickened, besides, his left maxillary sinus and ethmoidal sinus mucosa swelled obviously (Fig. 3). The Kartagener triad of symptoms was observed in this patient, which confirmed the diagnosis of KS. His primary problem was recurrent pneumonia due to bronchiectasis, which had to be resolved effectively. After careful examination of the chest CT, we concluded that the bronchiectatic lung was destroyed and had clear boundaries. Regarding his situs inversus, the left lung was clearly composed of three lobes, with lesion in the left middle lobe. There was no bronchiectatic signs in the upper or lower left lobe or in the right lung. Thus, we concluded that the damaged left middle lobe was the source of repeat pulmonary infections. Altogether, these results suggested that this lobe had to be resected, as the infections would otherwise not be curable.

After three days of antibiotic treatment (2 g cefotiam through an intravenous drip), left thoracotomy was performed via left posterolateral incision under the condition of general anesthesia. The fifth intercostal muscles were incised layer by layer, and sixth rib was cut off and

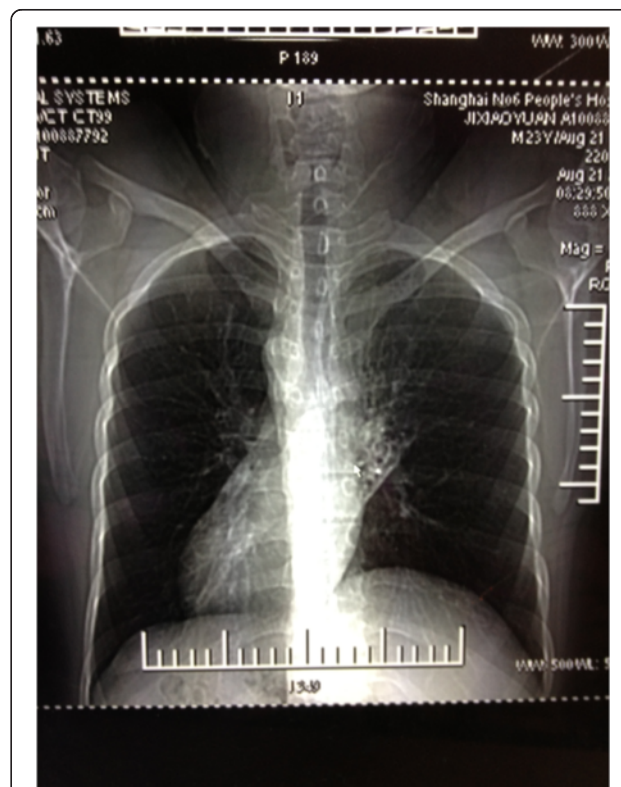


Fig. 2 Chest computed tomography scan showing dextrocardia



Fig. 3 Chest computed tomography scan showing left ethmoidal and maxillary sinusitis

opened with a rib retractor. The double lumen endotracheal catheter (Covidien Mallinckrodt Endobronchial Tube 37Fr left; Covidien, USA) was intubated into the left main bronchus. It was confirmed that the patient's left lung had three lobes, and the azygos vein and

superior vein cava could be exposed in the left thoracic cavity, confirming situs inversus. The middle lobe was collapsed and consolidated (Fig. 4), although the upper and lower lobes seemed normal and the fissures had completely developed. Before the left middle lobe was removed, the middle lobe branch of the pulmonary artery and vein, and the bronchus of the middle lobe were excised and sutured individually with staples (Covidien, USA). After lobe was resected, wound surface was stanching and the chest was washed with physiological saline water for 3 times. The flushing fluid was removed and lung wound surface was sutured with 3-0 Prolene. Rib incision was sutured with No. 10 silk; muscle layers were sutured with 2-0 Johnson ETHICON Vicryl; skin was sutured with 3-0 Johnson ETHICON Vicryl. In addition, a 28F chest tube was placed post-operatively and was removed 3 days later. The patient-controlled intravenous analgesia (PCIA) with sufentanil 0.2 mg combined with granisetron 3 mg was provided for pain relief for 48 h. The operation was completed without complication. The final pathology examination (Fig. 5) revealed the occurrence of bronchiectasis and hyperemia in this lobe, as well as the proliferation of

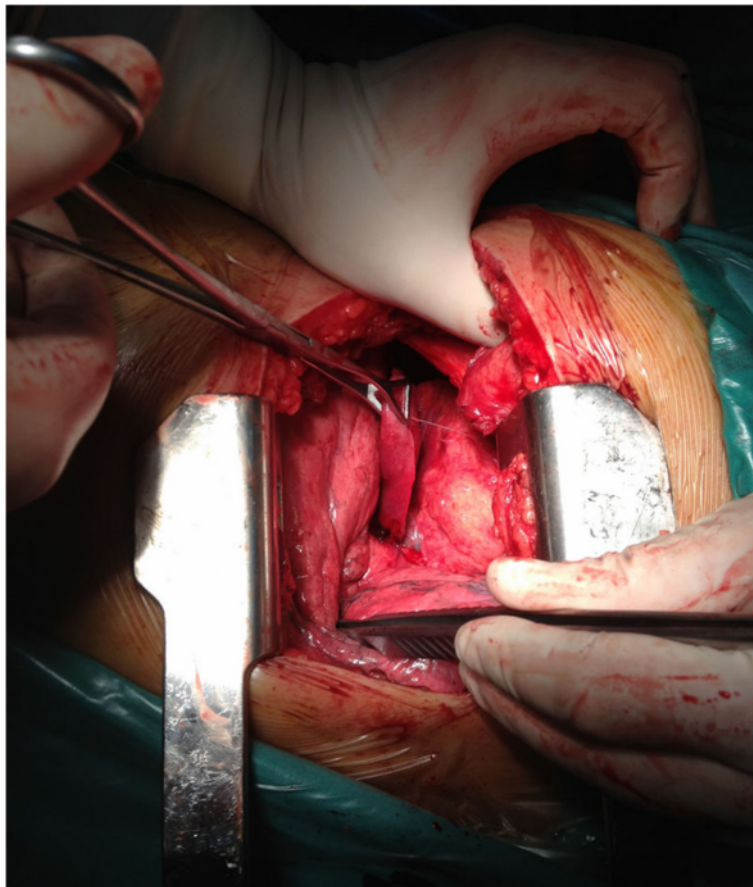


Fig. 4 Consolidation of the left middle lobe (held by lung-grasping forceps)

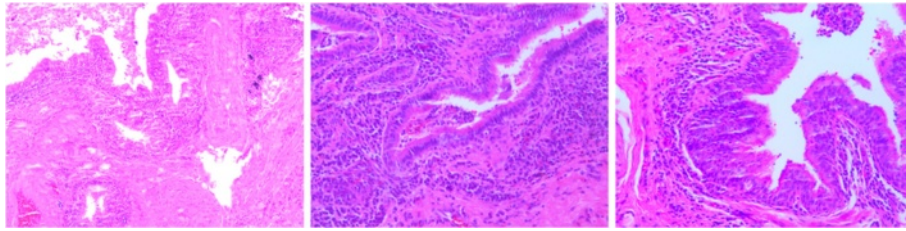


Fig. 5 The histologic images of the resected left middle lobe. A: 100 ×; B and C: 200×

lymphocytes and plasmocytes around the bronchus. The post-operative antibiotic therapy (2 g cefotiam through an intravenous drip) lasted 7 days, and chest CT indicated left lung recovered well without pulmonary infection. On postoperative day 8, the patient was discharged. Regular follow-up was conducted by the ear-nose-throat (ENT) specialist and thoracic surgeons. Mucolytics, such as myrtol standardized capsules (Pohl-Boskamp GmbH & Co. KG, Germany; 0.3 g three times a day), was taken to treat his remaining sinusitis, as administered by the ENT doctors. After 6 months of follow-up, his symptoms of cough and purulent sputum were relieved, and his nasal congestion and discharge were alleviated. Follow-up chest X-rays confirmed no further issues (Fig. 6).

Discussion

KS patients generally present with recurrent lower respiratory tract infection and nasal sinusitis due to ineffective mucociliary clearance [4]. Additionally, male patients are usually sterile, as sperm motility is dependent on ciliary function [5]. In the present study, testing in primary ciliary dyskinesia was not performed because of the technical condition constraints in our hospital, which was a limitation of our study. Importantly, the triad of symptoms in KS was observed in the present case, although no male infertility was noted because sperm motility was not assessed.

The left middle lobectomy in KS has rarely been reported due to the rarity of this disorder. The present case recovered well after the left middle lobectomy. We therefore summarized the treatment method for this disease.

The primary symptom of the patient was recurrent respiratory tract infection owing to bronchiectasis in the left middle lobe. The damaged lung was the source of the irreversible infection, if untreated, the range of the lesion would expand to further destroy the lung tissue. It has been reported that pulmonary heart disease may occur if lung transplantation is not performed in an end-stage KS patient [6–8]. Therefore, the left lobe resection was necessary in this case. Specially, the advantage of pulmonary resection for treating chronic bronchiectasis has been reported [9]. During the follow-up, the respiratory tract infection did not occur any further, although the patient suffered from occasional nasal congestion and discharge due to the ineffective mucociliary clearance. The results of the left lobectomy were desirable for him. However, the patient should continue to see his ENT specialist for the clinical treatment of sinusitis. Importantly, myrtol could help alleviate his symptoms.

Conclusions

In conclusion, serious damage of the lung may occur after bronchiectasis and pneumonia in KS patients unless a diagnosis of this rare syndrome can be confirmed and therapy is provided. Surgical resection is an acceptable method for patients whose bronchiectatic lungs are localized within one or two lobes. To the best



Fig. 6 Chest X-rays showing recovery after six months follow-up

of our knowledge, we are the first to report left middle lobectomy for treating bronchiectasis in KS patient. The progression of bronchiectasis was halted by surgery during the immediate post-operative follow-up period. For this reason, it should be emphasized that KS requires a high degree of suspicion for its early diagnosis by physicians, ENT surgeons, and thoracic surgeons.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Highlights

1. Chest CT revealed bronchiectasis in the left upper lobe and dextrocardia.
2. Paranasal sinus CT scan revealed left ethmoidal and maxillary sinusitis.
3. The left middle lobectomy was performed and the patient recovered well after surgery.

Abbreviations

CT: computed tomography; ENT: ear-nose-throat; KS: Kartagener syndrome.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

HL participated in the design of this study, and they both performed the statistical analysis. ZC carried out the study, together with XZ, collected important background information, and drafted the manuscript. QY conceived of this study, and participated in the design and helped to draft the manuscript. All authors read and approved the final manuscript.

Acknowledgements

None.

Received: 8 May 2015 Accepted: 5 February 2016

Published online: 09 March 2016

References

1. Gupta S, Handa KK, Kasliwal RR, Bajpai P. A case of Kartagener's syndrome: Importance of early diagnosis and treatment. *Indian J Hum Genet.* 2012;18:263.
2. Case R. *Anaesthetic Management of a Patient with Synchronous Kartagener Syndrome and Biliary Atresia.* 2015.
3. Pandit S, Choudhury S, Das A, Basuthakur S, Das SK. A rare case of Kartagener's syndrome. *J Nat Sci Biol Med.* 2014;5:175.
4. Boon M, Jorissen M, Proesmans M, De Boeck K. Primary ciliary dyskinesia, an orphan disease. *Eur J Pediatr.* 2013;172:151–62.
5. Montjean D, Courageot J, Altié A, Amar-Hoffet A, Rossin B, Geoffroy-Siraudin C, Tourame P, Boyer P. Normal live birth after vitrified/warmed oocytes intracytoplasmic sperm injection with immotile spermatozoa in a patient with Kartagener's syndrome. *Andrologia.* 2015;47(7):839–45.
6. Sha Y-W, Ding L, Li P. Management of primary ciliary dyskinesia/ Kartagener's syndrome in infertile male patients and current progress in defining the underlying genetic mechanism. *Asian J Androl.* 2014;16:101.
7. Shen Y-c, Wen F-q, Yi Q. Pulmonary arterial hypertension in a Kartagener syndrome patient: Treatment with beraprost sodium. *Int J Cardiol.* 2012;159:e9–10.
8. Sidney Filho LA, Machuca TN, Camargo JJ, Felicetti JC, Camargo SM, Perin FA, Sanchez LB, Schio SM. Lung transplantation without the use

- of cardiopulmonary bypass in a patient with Kartagener syndrome. *J Bras Pneumol.* 2012;38:806–9.
9. Hacıbrahimoglu G, Fazlioglu M, Olcmen A, Gurses A, Bedirhan MA. Surgical management of childhood bronchiectasis due to infectious disease. *J Thorac Cardiovasc Surg.* 2004;127:1361–5.

Submit your next manuscript to BioMed Central and we will help you at every step:

- We accept pre-submission inquiries
- Our selector tool helps you to find the most relevant journal
- We provide round the clock customer support
- Convenient online submission
- Thorough peer review
- Inclusion in PubMed and all major indexing services
- Maximum visibility for your research

Submit your manuscript at
www.biomedcentral.com/submit

