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Case Report

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Relapsing Polychondritis Presenting with Severe Laryngotracheobronchial Involvement and a Posterior Laryngeal Web: A Case Report

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Abstract:

Background: Relapsing polychondritis (RP) is a rare inflammatory disorder that can cause severe laryngotracheobronchial complications. Its airway manifestations may resemble bronchial asthma, delaying recognition and appropriate management.

Case presentation: A 45-year-old woman presented with a four-year history of progressive dyspnea, inspiratory stridor, wheezing, and weight loss despite repeated treatment for presumed bronchial asthma. Laryngoscopy revealed subglottic stenosis and a posterior laryngeal web requiring emergency tracheostomy and surgical excision. Subsequent computed tomography and bronchoscopy demonstrated severe tracheal narrowing, carinal distortion, complete left main bronchial obstruction, and left lung collapse. Tuberculosis and autoimmune investigations were inconclusive. Relapsing polychondritis was suspected based on the clinical and airway findings, but remained unconfirmed, and endobronchial tuberculosis could not be excluded. The patient received corticosteroids and antituberculosis treatment, followed by bronchoscopic balloon dilatation for persistent tracheal stenosis. She remained clinically stable during three years of follow-up, with minimal residual stridor; however, no follow-up imaging or bronchoscopy was performed to assess anatomical resolution.

Conclusion: This case illustrates the diagnostic challenges of unexplained progressive laryngotracheobronchial stenosis and highlights the importance of considering Relapsing polychondritis while acknowledging competing diagnoses. Long-term clinical stability does not necessarily indicate anatomical resolution of major airway obstruction.

Introduction

Relapsing polychondritis (RP) is a rare immune-mediated inflammatory disorder characterized by recurrent inflammation and progressive damage to cartilaginous structures. Laryngeal and tracheobronchial involvement can cause critical airway narrowing and substantial morbidity. Breathlessness, wheezing, stridor, and persistent cough may be attributed to asthma or other respiratory disorders, potentially postponing accurate diagnosis (1,2).

Recognizing RP is challenging when characteristic features are absent or inadequately recorded and investigations do not reveal a conclusive alternative diagnosis. Progressive airway narrowing may also mimic endobronchial tuberculosis, especially in regions where tuberculosis is prevalent. Posterior laryngeal web formation accompanying extensive tracheobronchial abnormalities presents an additional diagnostic challenge.

We describe a 45-year-old woman with progressive upper and lower airway obstruction, initially managed as bronchial asthma and subsequently treated surgically for a laryngeal web. Further investigations revealed severe tracheal narrowing, carinal distortion, complete left main bronchial obstruction, and left lung collapse. RP was considered the most likely diagnosis, although confirmation was limited and endobronchial tuberculosis remained a differential diagnosis. This case highlights the importance of considering systemic inflammatory disease in unexplained progressive airway stenosis and distinguishing clinical improvement from documented anatomical recovery.

Case Report

A 45-year-old businesswoman and mother of three presented with a four-year history of progressively worsening dyspnea, inspiratory stridor, wheezing, dry cough, fatigue, generalized weakness, joint pains, and significant weight loss. She had repeatedly received treatment for presumed bronchial asthma without symptomatic improvement. She denied previous airway trauma, endotracheal intubation, or laryngeal surgery. An external radiological assessment reportedly indicated non-erosive polyarthritis; however, the original digital imaging and communications in medicine (DICOM) files were unavailable for independent review.

Initial otolaryngological assessment and subsequent deterioration

Indirect laryngoscopy revealed subglottic stenosis and a posterior laryngeal web involving approximately two-thirds of the true vocal cords and laryngeal inlet. The patient underwent

emergency tracheostomy with simultaneous laryngeal web excision, leading to marked respiratory improvement. During follow-up, she developed aphonia and severe gastroesophageal reflux symptoms. Testing identified persistent *Helicobacter pylori* infection following previous unsuccessful eradication therapy. Examination revealed posterior pharyngeal wall hyperaemia. Treatment comprised pantoprazole 40 mg once daily, clarithromycin 500 mg twice daily, and tinidazole 500 mg twice daily for 14 days.

Five months later, she returned with recurrent hoarseness, noisy breathing, globus sensation, choking episodes, and dry cough. Repeat otolaryngological assessment confirmed recurrent subglottic stenosis. Her symptoms persisted despite subsequent successful *H. pylori* eradication, prompting referral to the pulmonology clinic approximately one month later.

Pulmonary investigations

Chest radiography demonstrated a small left pleural effusion, elevation of the left hemidiaphragm, and Chilaiditi syndrome **(Figure 1).** Contrast-enhanced chest computed tomography (CT) revealed severe tracheal narrowing, carinal distortion, and complete left main bronchial obstruction, associated with total left lung collapse, extensive mucus plugging, mediastinal shift, diaphragmatic elevation, and a small pleural effusion **(Figure 2A–D).**

Laboratory investigations showed a white blood cell count of $4.55 \times 10^3/\mu L$, haemoglobin of 12.24 g/dL, mean corpuscular volume of 77.45 fL, mean corpuscular haemoglobin of 23.11 pg, and platelet count of $504.8 \times 10^3/\mu L$, indicating microcytosis and thrombocytosis.

Flexible bronchoscopy demonstrated severe tracheal narrowing that prevented passage of the adult bronchoscope. A paediatric bronchoscope permitted visualization of marked carinal distortion, complete obliteration of the left main bronchus, and a normal right bronchial tree with hyperaemic mucosa **(Figure 3)**. Bronchoalveolar lavage was not feasible because the instrument was too small for adequate fluid instillation and suction.

CT-guided pleural aspiration was planned for microbiological, cytological, and biochemical analyses. However, preliminary CT imaging unexpectedly demonstrated complete left lung re-expansion and spontaneous resolution of the effusion. Mild residual left perihilar pneumonic changes and persistent gastric herniation into the thoracic cavity remained, so aspiration was abandoned **(Figure 4).**

Seven days following bronchoscopy, the patient presented with a productive cough yielding brownish sputum, evening fevers, night sweats, and repeated choking episodes. Sputum GeneXpert and bacterial cultures were negative, and oesophagogastroduodenoscopy was normal. C-reactive protein was 320 mg/L, erythrocyte sedimentation rate 110 mm/hour, and D-dimer 1,225

ng/mL. Serum adenosine deaminase, ferritin, peripheral blood smear, rheumatoid factor, anti-CCP antibodies, ANCA, and ANA profile were reported as normal or negative.

Diagnosis, treatment, and outcome

The combination of progressive laryngotracheobronchial disease and severe central airway abnormalities raised suspicion of relapsing polychondritis (RP), although endobronchial tuberculosis remained an important differential diagnosis. Dynamic expiratory CT and documented bronchoscopic expiratory collapse were unavailable; therefore, tracheobronchomalacia could not be confirmed.

Empirical corticosteroid and antituberculosis treatment was initiated. Prednisolone was administered at 60 mg daily for one week, followed by 40 mg, 20 mg, and 10 mg daily for one week each, then maintained at 5 mg daily. Azathioprine 50 mg daily was given for six months, alongside six months of antituberculosis treatment. After one month, cough resolved and weight progressively increased. However, inspiratory stridor persisted. Subsequent bronchoscopic balloon dilatation widened the tracheal lumen, but the bronchoscope could not pass beyond the completely obliterated left main bronchus.

Over three years of follow-up, the patient remained clinically stable with minimal residual stridor. No follow-up imaging or repeat bronchoscopy was performed; consequently, the anatomical status of the previously documented airway abnormalities remains unknown. Clinical stability therefore cannot be interpreted as evidence of anatomical resolution.

Discussion

Relapsing polychondritis (RP) is an uncommon immune-mediated disorder characterized by recurrent inflammation and progressive destruction of cartilaginous structures. Airway involvement in RP can be life-threatening, as laryngeal and tracheobronchial inflammation may lead to severe stenosis, permanent airway damage, and respiratory failure (3,4). Our patient exhibited extensive laryngotracheobronchial disease, characterized by subglottic stenosis, a posterior laryngeal web, severe tracheal narrowing, distortion of the carina, and total occlusion of the left main bronchus. This extensive airway involvement raised suspicion of RP, although the diagnosis remained unconfirmed.

RP commonly affects individuals between 40 and 50 years of age, with men and women affected similarly (3,5). Our patient's symptom onset at approximately 41 years was consistent with this epidemiological pattern. Although the underlying cause remains uncertain, immune-mediated mechanisms are thought to contribute to cartilage inflammation in susceptible individuals (3). No specific precipitating factor was identified in this case.

Airway manifestations of RP include laryngeal inflammation, subglottic stenosis, tracheal narrowing, and bronchial involvement, which may progress to fixed obstruction or dynamic airway collapse (3,4). In our patient, imaging and bronchoscopy established severe tracheal narrowing, carinal distortion, and complete left main bronchial obliteration associated with left lung collapse. However, dynamic expiratory imaging or documented bronchoscopic airway collapse was unavailable. Accordingly, tracheobronchomalacia could not be confirmed, and the findings are more accurately characterized as severe structural airway abnormalities. This distinction is important because fixed stenosis and dynamic airway collapse are related but different manifestations requiring appropriate assessment.

The initial presentation illustrates the diagnostic difficulty of airway-predominant RP. The patient was treated for suspected bronchial asthma for four years with little clinical benefit, until laryngoscopy identified subglottic narrowing and a posterior laryngeal web. Similar diagnostic delays have been reported because RP may resemble asthma, laryngitis, or recurrent respiratory infection (6). Laryngeal involvement can cause hoarseness, vocal cord dysfunction, stridor, and progressive airway compromise, sometimes necessitating tracheostomy (3,4). Although laryngeal manifestations are recognized features of RP, posterior laryngeal web formation appears to be an unusual presentation. The previously cited report (7) should be interpreted as evidence that this manifestation has been described, rather than sufficient evidence to establish its frequency or absolute rarity. In our patient, the web required emergency surgical excision, but its association with RP remains presumptive because the diagnosis itself was not definitive.

Systemic manifestations of RP may include fatigue, fever, weight loss, and inflammatory joint symptoms (3,5). Our patient experienced fatigue, generalized weakness, weight loss, and joint pains. An external radiological assessment reportedly identified non-erosive polyarthritis, but the original images were unavailable for independent review. Furthermore, characteristic auricular, nasal, and ocular inflammation was not documented. The reported none erosive arthritis suggests possible RP; however, the absence of additional characteristic manifestations leave the diagnosis uncertain.

EBTB was also considered because of the systemic symptoms and marked bronchial obstruction. Negative sputum GeneXpert testing and bacterial cultures did not definitively exclude EBTB. Bronchoalveolar lavage was not feasible, and direct sampling of the completely obstructed left main bronchus was not obtained. Consequently, microbiological confirmation or definitive exclusion of EBTB was not possible. The markedly elevated inflammatory markers supported an inflammatory or infectious process but were not specific for RP. Autoimmune serological investigations were negative, which helped assess alternative diagnoses but did not independently establish RP (3,5). The diagnosis should therefore be regarded as suspected rather than confirmed.

RP is primarily diagnosed clinically using established criteria, including those proposed by McAdam and subsequently modified by Damiani and Levine and by Michet (5). Although the extensive airway abnormalities and reported non-erosive polyarthritis supported the possibility of RP, the unavailable external imaging, lack of documented characteristic cartilage inflammation at other sites, and uncertainty The diagnosis remains uncertain because the response to treatment is

insufficiently documented to conclusively satisfy these criteria. Improvement after treatment is supportive only when interpreted alongside the other clinical findings.

Management of severe airway-predominant RP generally requires individualized medical treatment and, when indicated, airway intervention (3,5). Our patient received prednisolone and azathioprine alongside empirical antituberculosis therapy. Cough and weight improved after treatment, but stridor persisted, prompting bronchoscopic balloon dilatation. Because the medical treatments were administered concurrently and airway intervention followed, the improvement cannot be attributed to any single treatment. Moreover, the inability to traverse the left main bronchus indicated substantial residual obstruction at the time of the procedure.

An important outcome was clinical stability over three years, with minimal residual stridor. However, no follow-up imaging or repeat bronchoscopy was performed. Therefore, the subsequent anatomical status of the airway abnormalities remains unknown. Persistent major airway obstruction may impair ventilation and secretion clearance and increase the risk of recurrent infection or respiratory deterioration. This case emphasizes the importance of continued clinical surveillance and objective airway reassessment where feasible, while distinguishing symptomatic improvement from confirmed anatomical resolution.

Conclusion

Relapsing polychondritis (RP) should be considered in patients presenting with progressive, unexplained airway stenosis, particularly when respiratory manifestations coexist with features suggestive of systemic inflammatory disease. Early recognition and appropriate management may help prevent irreversible airway damage and potentially life-threatening complications. In this patient, the clinical presentation and subsequent course raised suspicion for RP; however, the absence of definitive diagnostic investigations limits the certainty of this diagnosis. This case highlights the importance of maintaining a high index of suspicion for RP in patients with atypical and progressive laryngeal or tracheal abnormalities, while acknowledging the diagnostic uncertainty and the need for careful clinical assessment.

Limitation

This case has several limitations. RP remained a presumptive diagnosis because characteristic auricular, nasal, or ocular chondritis was not documented, and the external joint imaging could not be independently reviewed. Furthermore, negative sputum GeneXpert testing did not definitively exclude endobronchial tuberculosis. Bronchoalveolar lavage and endobronchial sampling were unavailable because of severe airway obstruction and equipment limitations. As corticosteroids and antituberculosis therapy were administered together, the improvement could not be attributed to either treatment alone, and the presence of a coexisting condition could not be ruled out. Finally, the absence of repeat imaging or bronchoscopic evaluation during the three-year follow-up limited

objective assessment of the airway changes over time and prevented confirmation of whether the stenosis had progressed.

.DECLARATION

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Availability of data and materials

No datasets were generated or analyzed during the preparation of this case report. All relevant clinical information is included in the article.

Informed consent

The authors declare that, written informed consent was obtained from the patient for the publication of this manuscript and accompanying images and it will be available upon request.

Declaration of conflicting interests

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Authors' contributions

All authors have contributed. Some take care of the patient and do follow up, some wrote the manuscript, some critically reviewed and proof read the manuscript, some prepare images and figures and all approve the manuscript to be published.

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Author 1: **Dr. Hedwiga Swai:** Drafted the manuscript, managed the patient, conceived the case report, collected and interpreted the clinical data.

Author 2: **Dr. Sibtain Moledina:** Conceptualization, Collected clinical data, participated in patient follow-up, critically revised the manuscript, and approved the final version.

Author 3: **Dr. Pauline Chale:** Conceptualization, Collected clinical data, participated in patient follow-up, critically revised the manuscript, and approved the final version.

Author 4: **Gerald Mpemba:** Prepared and processed the clinical images, critically reviewed the manuscript, and approved the final version.

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Abreviation

1.	**RP**	Relapsing Polychondritis
2.	EBTB	**Endobronchial tuberculosis**
3.	ENT	Ear nose and throat
4.	ADA	adenosine deaminase

5. LDH Lactate dehydrogenase
6. Ant CCP Anti-Cyclic Citrullinated Peptide Antibodies
7. **C-ANCA** Cytoplasmic Antineutrophil Cytoplasmic Antibodies.
8. P-ANCA Perinuclear Antineutrophil Cytoplasmic Antibodies.
9. ANA profile Antinuclear Antibodies Profile
10. SGS Subglottic stenosis
11. TBM Tracheobronchomalacia

Figures

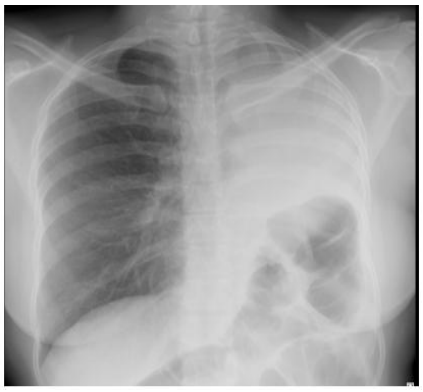


Figure 1: Chest xray showing Left sided pleural effusion and Elevation of LT hemi diaphragm

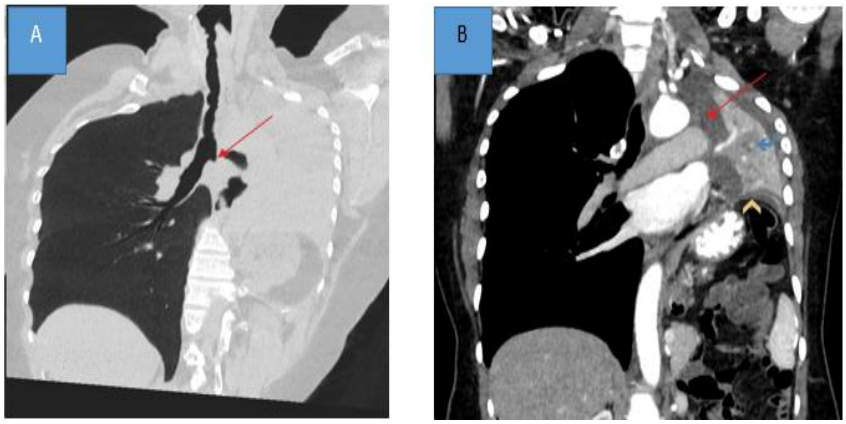


Figure 2: (A) CT chest Coronal reformatted minimum intensity projection showing irregular and narrowed trachea and abrupt termination (stenosis) of the left main bronchus (arrow). **(B)** Coronal reformatted CT showing collapsed lung (curved arrow), Loculated effusion (long arrow) and elevated left hemidiaphragm (arrow head)

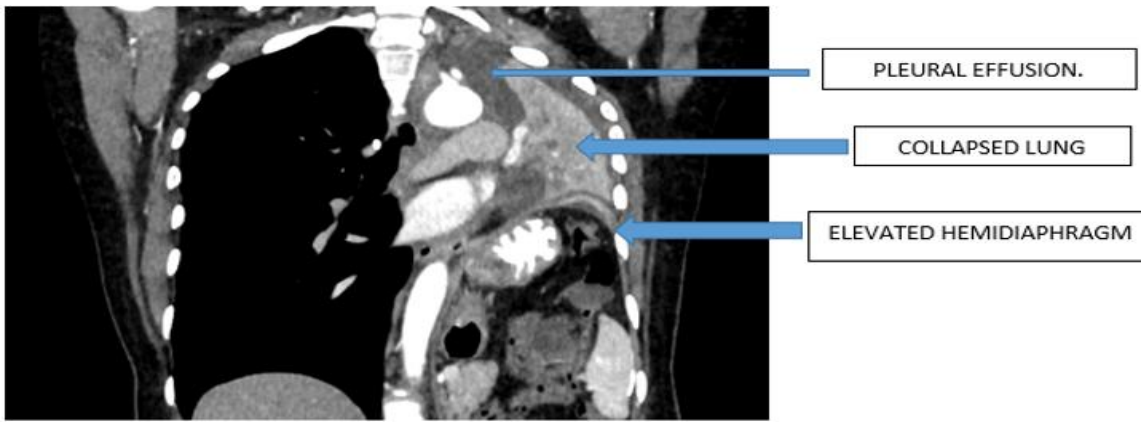


Figure 2c: Coronal contrast-enhanced chest CT shows left pleural effusion with complete passive atelectasis of the left lung, marked elevation of the left hemidiaphragm, and significant volume loss of the left hemithorax.

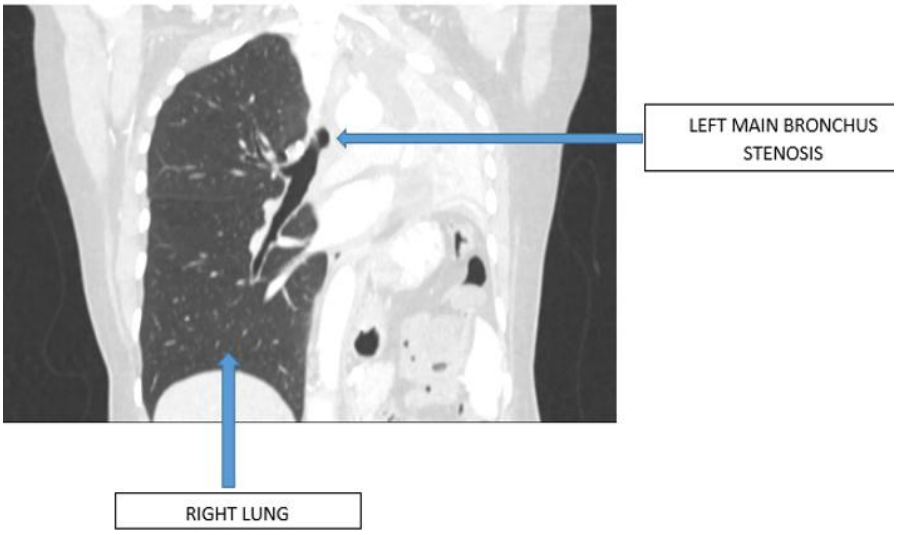


Figure 2d: Coronal chest CT demonstrating stenosis of the left main bronchus (arrow), with preserved aeration of the right lung.

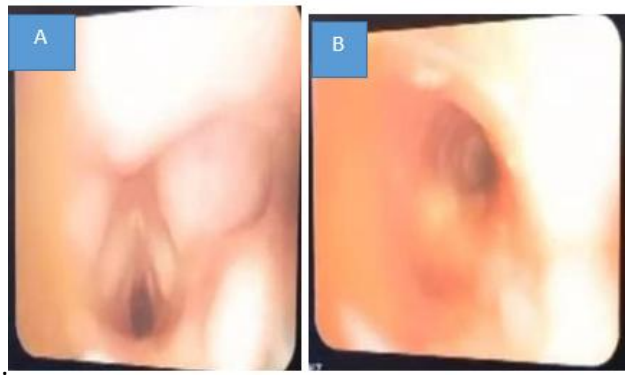


Figure 3: **Image A:** Shows proximal tracheal narrowing (tracheal orifice) with relatively intact surrounding mucosa. **Image B:** Demonstrates marked narrowing of the deeper tracheal lumen, compromising airway patency.

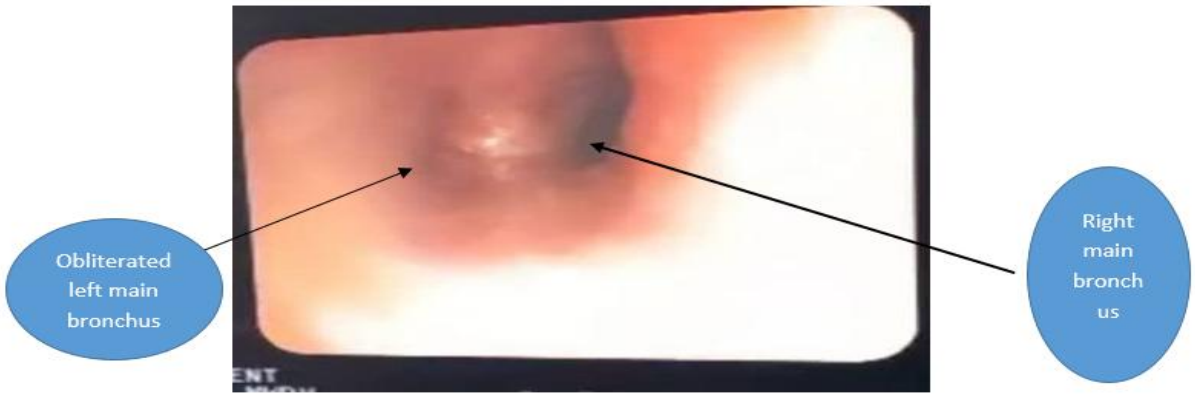


Figure 3 c: The carinal ridge is markedly distorted and blunted. Complete occlusion of the left main bronchus, with a patent right main bronchus.

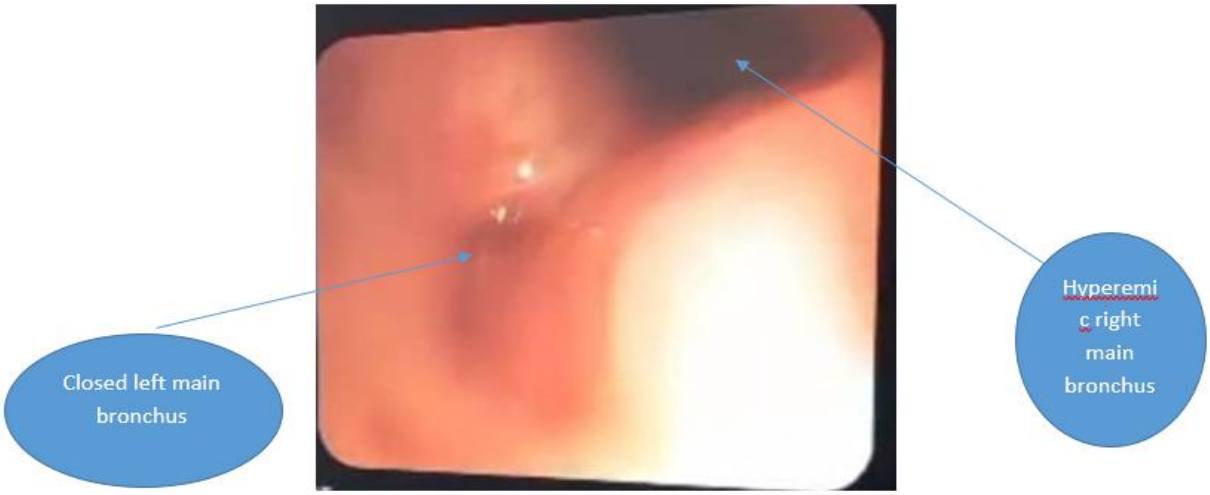


Figure 3 d: Complete obliteration of the left main bronchial orifice, with no visible patent lumen. Patent right main bronchus with visibly erythematous mucosa.

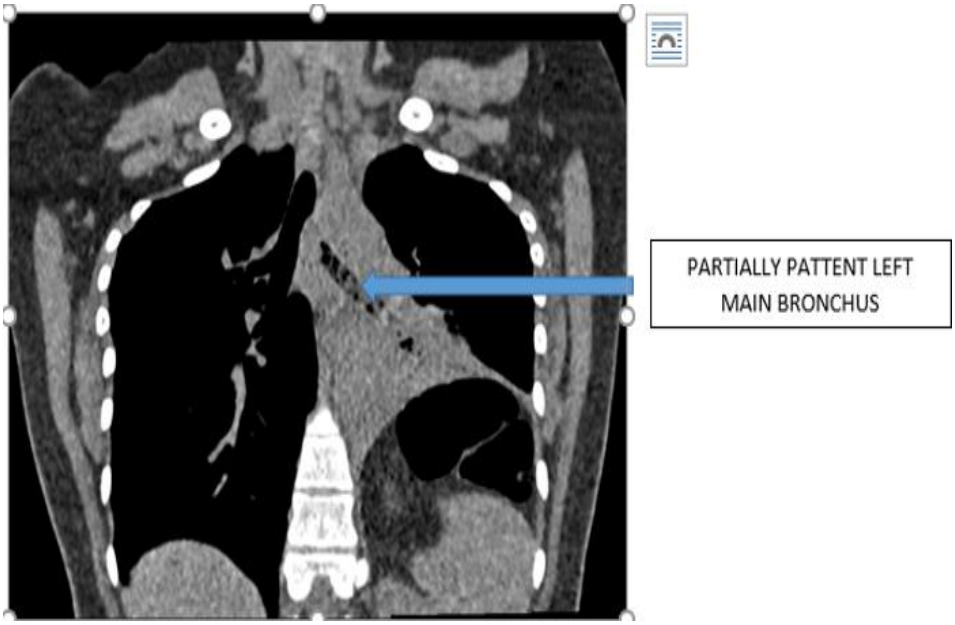


Figure 4a: CT chest: After bronchoscopy dilatation showing partially patent left main bronchus and expansion of the left lung

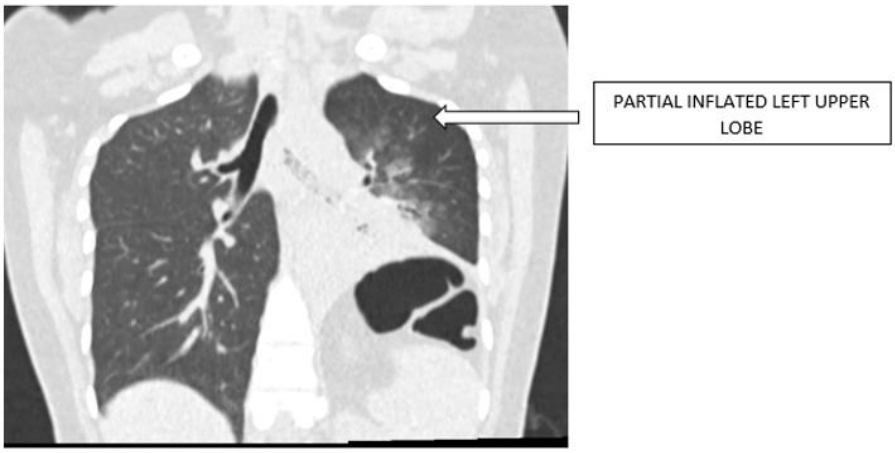


Figure 4b: CT chest: After bronchoscopy dilatation showing partially inflated left upper lobe

Clinical Timeline

Time	Event	Management and outcome
Four years before pulmonology referral	Progressive dyspnea, inspiratory stridor, wheezing, dry cough, fatigue, joint pains, and weight loss. Symptoms persisted despite treatment for presumed bronchial asthma.	Repeated asthma treatment provided no symptomatic improvement.
Initial ENT assessment	Laryngoscopy revealed subglottic stenosis and a posterior laryngeal web involving approximately two-thirds of the true vocal cords and laryngeal inlet.	Emergency tracheostomy and web excision were performed, resulting in substantial improvement in breathing.
During subsequent follow-up	Aphonia and severe gastroesophageal reflux symptoms developed. Persistent Helicobacter pylori infection was identified.	Pantoprazole 40 mg once daily, clarithromycin 500 mg twice daily, and tinidazole 500 mg twice daily were administered for 14 days.
Five months after the initial ENT procedure	Recurrent hoarseness, noisy breathing, globus sensation, choking episodes, and dry cough developed. Repeat assessment confirmed recurrent subglottic stenosis.	Symptoms persisted despite subsequent successful H. pylori eradication, prompting referral to pulmonology approximately one month later.
Initial pulmonary evaluation	Chest radiography showed a small left pleural effusion, elevated left hemidiaphragm, and Chilaiditi syndrome. Contrast-enhanced CT demonstrated severe tracheal narrowing, carinal distortion, complete left main bronchial obstruction, and total left lung collapse.	Further pulmonary investigations and bronchoscopy were undertaken to characterize the airway abnormalities.
During bronchoscopy	Severe tracheal narrowing prevented passage of the adult bronchoscope. A paediatric bronchoscope demonstrated marked carinal distortion and complete left main bronchial obliteration.	Bronchoalveolar lavage was not feasible because of equipment limitations.
During planned pleural aspiration	Preliminary CT unexpectedly showed complete left lung re-expansion and spontaneous resolution of the pleural effusion.	Pleural aspiration was abandoned. Mild residual left perihilar pneumonic changes and persistent gastric herniation remained.
One week after bronchoscopy	Productive cough with brownish sputum, evening fever, night sweats, and recurrent choking developed. Inflammatory markers were elevated; sputum GeneXpert and bacterial cultures were negative.	Further evaluation was performed. RP was suspected, while endobronchial tuberculosis remained an important differential diagnosis.

Following diagnostic assessment	Progressive laryngotracheobronchial disease and severe central airway abnormalities supported a presumptive diagnosis of RP.	Prednisolone was initiated at 60 mg daily for one week, followed by 40 mg, 20 mg, and 10 mg daily for one week each, then maintained at 5 mg daily. Azathioprine 50 mg daily was administered for six months, alongside six months of antituberculosis treatment.
One month after treatment initiation	Cough resolved and weight progressively increased, but inspiratory stridor persisted.	Medical treatment was continued; subsequent clinical improvement could not be attributed to any single treatment.
After medical treatment	Persistent airway narrowing required further intervention.	Bronchoscopic balloon dilatation widened the tracheal lumen, but the bronchoscope could not pass beyond the completely obliterated left main bronchus.
Three-year follow-up	The patient remained clinically stable with minimal residual stridor.	No repeat imaging or bronchoscopy was performed; therefore, the long-term anatomical status of the airway remained undetermined.

Supplementary Files

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