Scientific Research Publishing

Open Journal of Respiratory Diseases, 2021, 11, 76-83
https://www.scirp.org/journal/ojrd
ISSN Online: 2163-9418
ISSN Print: 2163-940X

# Unusual Cause of Stridor in an Adult Man, Selective Immunoglobulin A Deficiency

**Safwat A. M. Eldaboosy[1,2], Amgad Awad[1,3], Ayman Ghoneim[1,4]**

[1]Almoosa Specialist Hospital, Al-Ahsa, Saudi Arabia
[2]Department of Chest Diseases, Al Azhar Faculty of Medicine, Al-Azhar University, Cairo, Egypt
[3]Department of Internal Medicine, Al Azhar Faculty of Medicine, Al-Azhar University, Cairo, Egypt
[4]Department of Cardiothoracic Surgery, Al Azhar Faculty of Medicine, Cairo, Egypt
Email: safwatchest@gmail.com

**How to cite this paper:** Eldaboosy, S.A.M., Awad, A. and Ghoneim, A. (2021) Unusual Cause of Stridor in an Adult Man, Selective Immunoglobulin A Deficiency. *Open Journal of Respiratory Diseases*, **11**, 76-83.
https://doi.org/10.4236/ojrd.2021.112007

**Received:** March 22, 2021
**Accepted:** May 25, 2021
**Published:** May 28, 2021



## Abstract

The common causes of stridor in adults are abscesses or swelling of the upper airway, tumors, paralysis, or malfunction of vocal cords. Tracheitis due to immunoglobulin deficiency may be a rare explanation for stridor in adults, although occasionally reported in children. We report an adult man having stridor secondary to isolated immunoglobulin A deficiency. We did an in-depth review of the literature to seek out no reported cases of stridor thanks to immunoglobulin deficiency in adults. This case underlines the very fact that a standard symptom like stridor rarely occurs thanks to uncommon causes. This case is exclusive because it reports the presence of stridor associated with isolated immunoglobulin A deficiency in an adult patient. The explanation for the stridor was an intraluminal narrowing of the upper part of the extrathoracic trachea.

## Keywords

Stridor, Tracheitis, Immunoglobulin A Deficiency

## 1. Introduction

Stridor is an abnormal, high-pitched sound produced by turbulent airflow through a partially obstructed airway. Common causes of stridor in an adult include acute epiglottitis, abscesses or swelling of the upper airway, tumors, and paralysis or malfunction of the vocal cords tracheal stenosis from previous intubation or tracheostomy, angioedema, and foreign body aspiration. The explanation for stridor is typically obvious on investigations like CT scanning or flexible bronchoscopy. Stridor represents a medical emergency and stridor implies critical airway

obstruction of a minimum of 50% of the airway lumen. Patients with stridor are at high risk of respiratory failure and death and need initial stabilization to take care of ventilation and oxygenation if this is often according to the goals of care. Immunoglobulin A (IgA) is that the most abundant antibody isotype produced within the body. IgA deficiency (IgAD) is that the commonest primary immunodeficiency and therefore the pathogenesis isn't defined clearly. Selective immunoglobulin A deficiency (SIgAD) features a worldwide prevalence that differs from one region to a different one, with a lower incidence in Asian countries; like Japan, China, India, and Saudi Arabia. SIgAD occurs in Arabs at a rate of 1 case per 142 persons, in white persons at a rate of 1 case per 500 - 700 persons. SIgAD is more frequent in adult subjects with chronic lung disease than in healthy, age-matched control subjects. Most of the people with SIgAD are asymptomatic, and some may present with recurrent infections of respiratory and gastrointestinal tracts, allergic disorders, and autoimmune manifestations. We report a case with SIgAD presented with recurrent tracheitis with attacks of stridor. However, in our case, when the investigations did not reveal an etiology, an exhaustive looking for rare causes of stridor was required.

## 2. Case Presentation

A 33-year-old, Saudi male, a non-smoker, presented to the chest clinic with chronic cough and shortness of breath with cough sometimes progressive to shortness of breath with minimal effort related to a way of suffocation for a period of three months. He had experienced multiple emergency department visits due to breathing difficulties. The patient received a diagnosis of asthma and was treated with short courses of systemic steroids, multiple inhalers, and courses of antibiotics; leading to only mild and temporary improvement in his symptoms. The patient had infrequently experienced expectoration of huge size viscid secretions. He denied fever, pain, or hemoptysis. He denied rash or arthralgia. He had no history of pneumonia or gastroenteritis. Within the past, he had chronic sinusitis, and seasonal rhinitis, and that he received antihistamines, analgesics, and antibiotics. Case history was irrelevant. The patient appeared well with a vital signs of blood pressure 120/80 mmHg, pulse of 90 beats/min and regular, rate of respiration of 20/min. His saturation on room air was at 96%. Head and neck examination didn't demonstrate lymphadenopathy or signs of chondral inflammation. His cardiovascular examination revealed a traditional vena jugularis pulse with normal heart sounds and no pedal edema. Respiratory examination demonstrated the absence of clubbing and no evidence of wheezing or crackle. However, forced expiration produced an audible stridor sound. Laboratory tests revealed normal complete blood count, liver function test, and renal function test. CRP was 4.5 mg/L. Old computed tomographic (CT) paranasal sinuses scanning showed deviated septum to the left side, congested nasal mucosa, paranasal sinuses mucosal thickening with obliterated bilateral osteomeatal complex and left sphenoethmoidal recess. Nasal swab for culture and sensitivity revealed a mixed

growth of Klebsiella pneumoniae and Streptococcal pneumoniae. Chest X-ray was normal (Figure 1). The CT scan of the thorax demonstrated a couple of filling defects within the lumen of the upper thoracic trachea related to mild wall thickening without significant luminal narrowing, also as clear lung parenchyma without mediastinal mass (Figure 2). The 2 main differential diagnoses were granulomatous polyangiitis or tracheal papillomatosis Spirometry demonstrated a flattening of the inspiratory curve. Forced expiratory volume in 1 (FEV1) was measured as 80% and FEV1/forced capacity was measured as 99%. The mid expiratory flow of 25% - 75% was 66%. The patient underwent flexible bronchoscopy, which showed inflammation of the upper a part of the trachea with circumferential narrowing and viscid secretions. The remaining of the tracheobronchial field was clear without significant inflammation. Biopsy and aspiration from the affected part were performed and sent for microbiology and histopathology

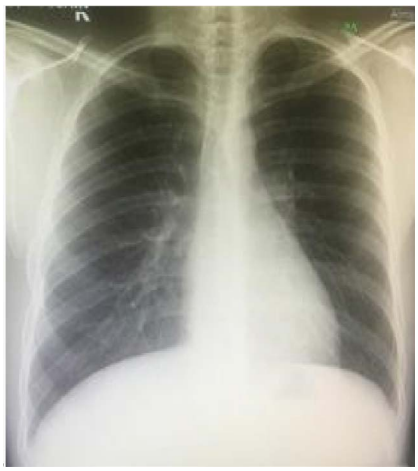

**Figure 1.** Chest X-ray: normal.

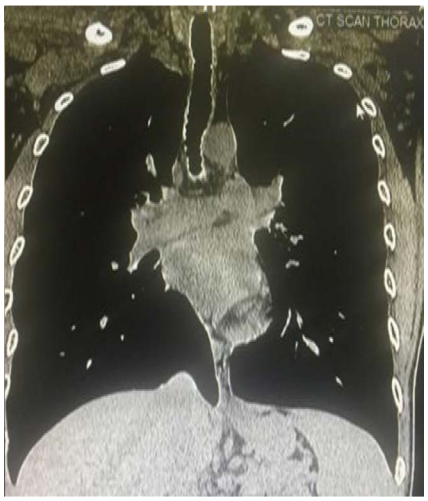


**Figure 2.** CT chest: a few filling defects in the lumen of the upper thoracic trachea associated with mild wall thickening without significant luminal narrowing.

(Figure 3 and Figure 4). The patient was given oral steroids for 10 days with intravenous piperacillin-tazobactam 4.5 gm TID for five days followed by levofloxacin 500 mg orally once daily for 15 days.

The culture of the bronchoscopic samples revealed mixed growth of Klebsiella pneumoniae, Pseudomonas aeruginosa, and Acinetobacter. Serum immunoglobulin levels revealed normal ranges of IgG (15 g/dL; normal 6.5 - 16 g/dL), and IgM (0.05 g/dL; normal 0.35 - 2.5 g/dL), with very low levels of IgA (0.4 g/dL; normal 0.5 - 3 g/dL) (Isolated IgA deficiency). The patient has diagnosed with selective immunoglobulin A deficiency consistent with very low serum immunoglobulin A while normal levels of immunoglobulin G and M. The patient still had some equivalent symptoms despite antibiotic and systemic steroid so rigid bronchoscopy was performed aiming for a re-evaluation and to clear the airway. This revealed that the upper a part of the trachea was crammed with necrotizing tissue and secretions were partially obstructing the upper airway. The airways were cleaned and cleared at the top of the procedure and biopsies were taken (Figure 5 and Figure 6).

Antibiotic completed for two weeks and one dose of intravenous immunoglobulin 500 mg/kg given over 6 hours without complications. The histopathological results revealed non-specific chronic inflammation. Flexible bronchoscopy:

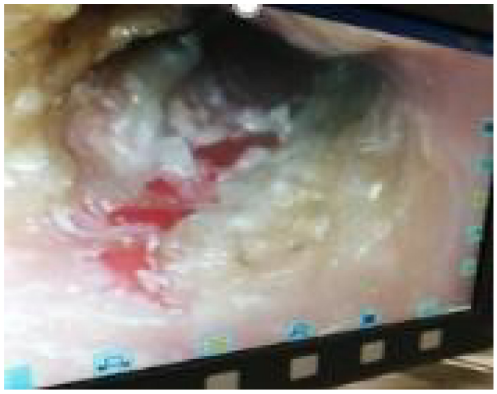

**Figure 3.** Flexible bronchoscopy: inflammation of the upper part of the trachea with circumferential narrowing and viscid secretions.

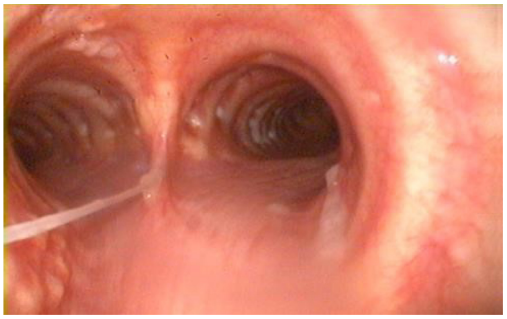

**Figure 4.** The remaining of the tracheobronchial field was clear without significant inflammation.

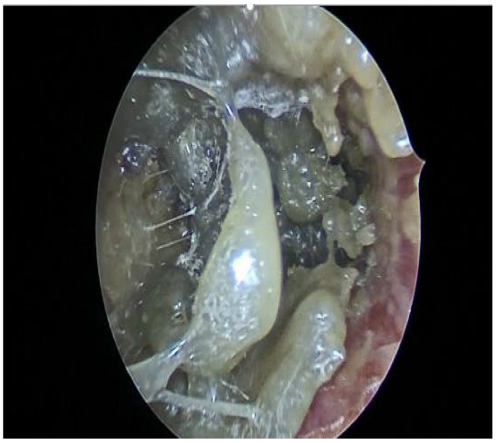

**Figure 5.** Rigid bronchoscopy was performed aiming for a second look and to clear the airway. This revealed that the upper part of the trachea was filled with necrotizing tissue and secretions were partially obstructing the upper airway.

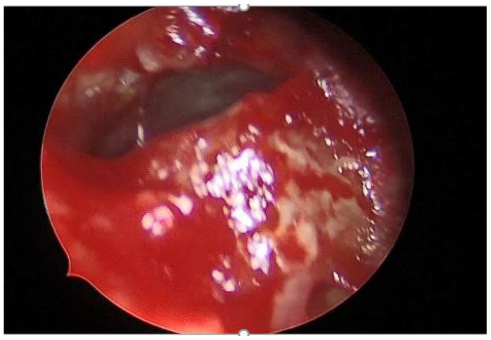

**Figure 6.** Rigid bronchoscopy finding after biopsy still show partial obstruction of the upper part of the trachea.

done after 2 months and it had been normal. Our case was managed properly with bronchoscopic interventions, intravenous antibiotics, and intravenous immunoglobulin replacement. He has been symptom-free throughout a six-month follow-up period.

## 3. Discussion

Our patient demonstrated the syndrome of selective IgA deficiency related to stridor thanks to tracheitis. This is often the primary case within the literature reported thanks to stridor associated with immunoglobulin deficiency. Selective IgA deficiency defines as serum IgA and secretory IgA (SIgA) were absent while other immunoglobulins were present in normal quantity and performance. Recurrent sinopulmonary infections could be presented only within the sort of upper tract infection or of more severe forms that find yourself with sequels like bronchiectasis or obliterative bronchiolitis [1] [2]. Most infections are caused by extracellular encapsulated bacteria (e.g. Haemophilus influenzae, Streptococcus

pneumoniae). it had been found that 25% of patients with IgA deficiency were diagnosed during screening for allergic disorders [3].

Central airway infections causing obstruction are probably rare. Awareness of their existence and therefore the possibility of bronchoscopic intervention for rapid relief of obstructive symptoms or treatment of persistent airway obstruction is important [4]. Appropriate initial workup for a case of stridor includes visualization of the airway through bronchoscopy for intraluminal pathology and CT scans with or without virtual reconstruction for extraluminal causes. These are usually sufficient for diagnosis and treatment planning. However, in our patients, these investigations were helpful without identifying the underlying cause so we requiring out-of-the-box thinking to think about rare diagnoses. The role of bronchoscopy is invaluable for the diagnosis of CAOI, however, its role in CAOI management and post-treatment surveillance isn't well defined. Clinical presentation and imaging are essential for the diagnosis of airway obstruction but bronchoscopy is usually required to get specific diagnosis through direct airway inspection, and tissue sampling using an endobronchial biopsy, brush, fine needle aspiration, and bronchial washing [5] [6]. Immunocompromised patients may present with nonspecific respiratory symptoms, and routine bronchoscopy of those patients may help within the diagnosis of endobronchial disease. The endobronchial appearance of infection can present with wall edema, pseudomembranes, necrotizing pseudomembranous lesions, ulcerative lesions, whitish or yellowish plaques, endoluminal masses, and vegetations [7].

The airway management of CAOI is actually almost like cases of malignant airway obstructions and consists of endobronchial debulking, balloon bronchoplasty, endobronchial laser therapy, argon plasma coagulation, cryotherapy, and airway stent placement. Both flexible and rigid bronchoscopies are often wont to treat CAOI [4] [8] [9]. In our case bronchoscopy did major diagnostic, and therapeutic values. There's no recommended specific treatment for patients with SIgAD, and supported recognized condition, patients should be managed individually. However, some patients gradually develop normal levels of IgA without treatment. In contrast, a couple of patients with IgA deficiency can reach common variable immunodeficiency (CVID) [10]; this attended occur in adolescence or young adulthood.

Management of patients with SIgAD consists of various modalities including education, periodic monitoring, treatment of associated allergic or autoimmune conditions, prolonged or maybe prophylactic antibiotics, administration of polyvalent pneumococcal vaccines, and administration of intravenous or subcutaneous immunoglobulin (IVIg) replacement therapy [11]. In our case, we got 2 weeks of intravenous and oral antibiotics with oral steroids with partial improvement. Patients receiving IgG therapy should have regular monitoring of IgG trough levels, blood corpuscle counts, and serum chemistry. The adequacy of IgG replacement is detected by the trough or steady-state IgG level in association with the clinical course [12]. In most of the practice guidelines, a starting dosage of IgG between 400 and 600 mg/kg/month is suggested to realize a serum

trough IgG level of 600 - 800 mg/dL. Generally, immunoglobulin replacement should be done cautiously with a product low in IgA. During this situation, usually, IVIg therapy is often given safely [13]. In our case, the patient received intravenous immunoglobulin once under closed observation without complications.

In summary, this case is exclusive because it reports the presence of stridor associated with isolated immunoglobulin A deficiency in an adult patient. The explanation for the stridor was an intraluminal narrowing of the upper a part of the extrathoracic trachea.

## Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this report.

## Consent

Written consent was obtained from the patient and Ethical approval was obtained from the Institutional Review Board at our hospital (IRB log number: ARC-21.02.3).

## References

[1] Yazdani, R., Latif, A., Tabassomi, F., *et al*. (2015) Clinical Phenotype Classification for Selective Immunoglobulin A Deficiency. *Expert Review of Clinical Immunology*, **11**, 1245-1254. https://doi.org/10.1586/1744666X.2015.1081565

[2] Ozkan, H., Atlihan, F., Genel, F., Targan, S. and Gunvar, T. (2005) IgA and/or IgG Subclass Deficiency in Children with Recurrent Respiratory Infections and Its Relationship with Chronic Pulmonary Damage. *Journal of Investigational Allergology and Clinical Immunology*, **15**, 69-74.

[3] Cunningham, R.C. (2001) Physiology of IgA and IgA Deficiency. *Journal of Clinical Immunology*, **21**, 303-309. https://doi.org/10.1023/A:1012241117984

[4] Keshishyan, S., DeLorenzo, L., Hammoud, K., Avagyan, A., Assallum, H. and Harris, K. (2017) Infections Causing Central Airway Obstruction: Role of Bronchoscopy in Diagnosis and Management. *Journal of Thoracic Disease*, **9**, 1707-1724

[5] Qingliang, X. and Jianxin, W. (2010) Investigation of Endobronchial Tuberculosis Diagnoses in 22 Cases. *European Journal of Medical Research*, **15**, 309-313. https://doi.org/10.1186/2047-783X-15-7-309

[6] Aliyali, M., Hedayati, M.T., Habibi, M.R., *et al*. (2013) Clinical Risk Factors and Bronchoscopic Features of Invasive Aspergillosis in Intensive Care Unit Patients. *Journal of Preventive Medicine and Hygiene*, **54**, 80-82.

[7] Gupta, V., Rajagopalan, N., Patil, M., *et al*. (2014) Aspergillus and Mucormycosis Presenting with Normal Chest X-Ray in an Immunocompromised Host. *BMJ Case Reports*, **2014**. https://doi.org/10.1136/bcr-2014-204022

[8] Shah, P. (2011) Atlas of Flexible Bronchoscopy. CRC Press, London. https://doi.org/10.1201/b13458

[9] Scarlata, S., Fuso, L., Lucantoni, G., Varone, F., Magnini, D., Antonelli Incalzi, R. and Galluccio, G. (2017) The Technique of Endoscopic Airway Tumor Treatment. *Journal of Thoracic Disease*, **9**, 2619-2639. https://doi.org/10.21037/jtd.2017.07.68

[10] Aghamohammadi, A., Mohammadi, J., Parvaneh, N., *et al*. (2008) Progression of

Selective IgA Deficiency to Common Variable Immunodeficiency. *International Archives of Allergy and Immunology*, **147**, 87-92. https://doi.org/10.1159/000135694

[11] Yazdani, R., Azizi, G., Abolhassani, H. and Aghamohammadi, A. (2017) Selective IgA Deficiency: Epidemiology, Pathogenesis, Clinical Phenotype, Diagnosis, Prognosis and Management. *Scandinavian Journal of Immunology*, **85**, 3-12. https://doi.org/10.1111/sji.12499

[12] Bonilla, F.A., Khan, D.A., Ballas, Z.K., *et al*. (2015) Practice Parameter for the Diagnosis and Management of Primary Immunodeficiency. *Journal of Allergy and Clinical Immunology*, **136**, 1186-1205. https://doi.org/10.1016/j.jaci.2015.04.049

[13] Cheraghi, T., Aghamohammadi, A., Mirminachi, B., *et al*. (2014) Prediction of the Evolution of Common Variable Immunodeficiency: HLA Typing for Patients with Selective IgA Deficiency. *Journal of Investigational Allergology and Clinical Immunology*, **24**, 198-200.