Ectopic Tracheal Bronchus Diagnosed on High-Resolution Computed Tomography: A Rare Entity

Awais Abbas, Qalab Abbas, Naveed ur Rehman Siddiqui and Aniqa Abdul Rasool

Department of Paaediatrics and Child Health, The Aga Khan University, Karachi, Pakistan

ABSTRACT

Ectopic tracheal bronchus, also known as a bronchial variant or accessory bronchus, is a rare congenital malformation. It refers to an abnormal bronchus originating as an additional tracheal outgrowth early in embryonic life, commonly from the lateral wall of trachea above the carina. It is often associated with some other congenital malformation. It is usually asymptomatic but may cause recurrent chest infections with right upper lobe involvement. Diagnosing a tracheal bronchus is a challenge even to an expert physician and sometimes manifests later in adulthood and leads to potential complications. We present a case of this rare congenital anomaly masked as multiple past presentations of recurrent chest infections in a critical care setting and diagnosed on a high-resolution computed tomography scan.

Key Words: *Tracheal bronchus, Ectopic bronchus, Pig bronchus, High-resolution computed tomography.*

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INTRODUCTION

Congenital malformations of the respiratory system, such as tracheoesophageal fistula (incidence 1 in 4500 births), ectopic tracheal bronchus (prevalence 0.9 - 3%), pulmonary atresia (incidence 1 in 7000 births), pulmonary hypoplasia (incidence 1.4 in 1000 births), and congenital lobar emphysema (1 in 20,000 births) cause anatomic and physiologic dysfunction with consequent respiratory distress and a significant burden of respiratory morbidity and mortality in children.[1,2] Some of these abnormalities often appear in infancy and pose diagnostic and therapeutic challenges, necessitating a multidisciplinary approach for effective management.

Tracheal bronchus is a rare congenital anomaly where an aberrant, accessory, or ectopic bronchus usually arises from the right side of the tracheal wall above the carina. The prevalence of tracheal bronchus ranges from 0.9 - 3% in the paediatric population, with most cases arising from the right side of the tracheal wall.[2] It can occur at any point in the trachea, between the cricoid cartilage to the carina, but typically presents within a 2-cm range above the main carina.

Correspondence to: Dr. Aniqa Abdul Rasool, Department of Paediatrics and Child Health, The Aga Khan University, Karachi, Pakistan
E-mail: aniqaasajwani@gmail.com

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There are two theories regarding its origin; either tracheal buds are present in utero during development and fail to regress, or there is some disturbance in embryogenesis, for instance, bronchial mesenchyme gets relocated into the trachea, that could result in the formation of a tracheal bronchus. Patients are usually asymptomatic as it generally does not interfere with normal physiologic processes. However, some children with tracheal bronchus have been shown to present with recurrent chest infections, such as pneumonia, bronchitis, and bronchiectasis. Studies have also linked this rare occurrence to other congenital malformations.[2]

Here, we present a case of an aberrant tracheal bronchus with recurrent chest infections who presented in a critical care setting, and diagnosed on high-resolution computed tomography (HRCT) scan.

CASE REPORT

An 8-month breastfed, partially vaccinated male infant weighing 9.8 kg, presented to the emergency department with complaints of bouts of cough, respiratory distress, and fever for two days. His medical history was remarkable for five episodes of respiratory illness since birth, resulting in multiple hospital admissions. The elder sibling had a history of asthma.

The physical examination revealed an irritable, excessively crying infant with a heart rate of 170/minute, respiratory rate of 60/minute, oxygen saturation of 97% on room air, and signs of respiratory distress, including tachypnea, sub-costal, and intercostal retractions with bilateral crepitations and mild wheezes on auscultation. He was fully awake with a Glasgow coma scale (GCS) of 15, capillary refill time (CRT) of <2 seconds, good volume central and peripheral pulses, and a soft abdomen with no abnormal findings.

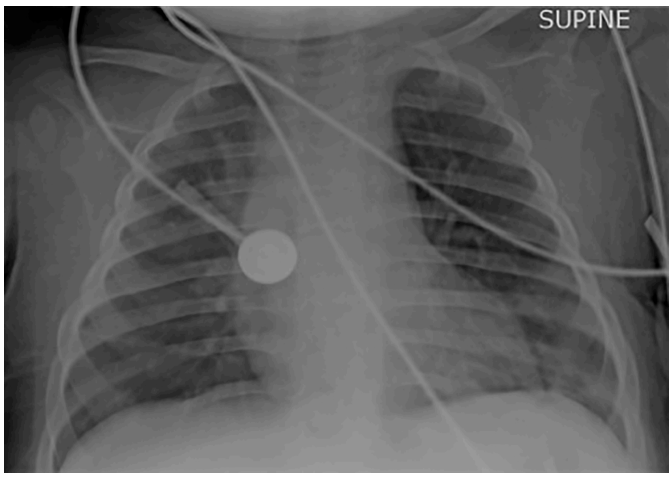

Figure 1: Chest radiograph showing peri-hilar infiltrates with inhomogeneous shadowing in the left lower zone, likely secondary to infection.

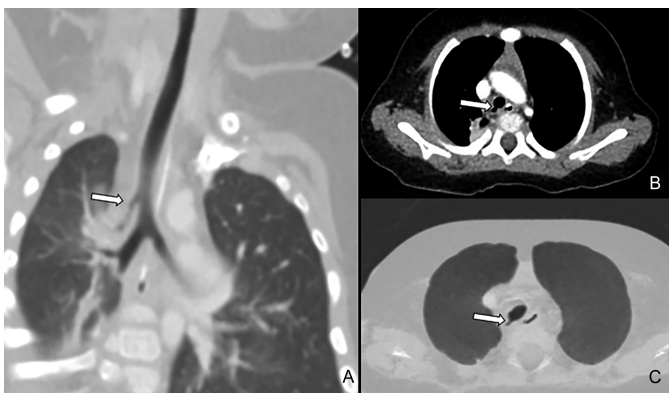

Figure 2: Right-sided aberrant bronchus arising from the supra-carinal trachea supplying posterior segment of right upper lobe. (A) Computed tomography scan, coronal view. (B) Computed tomography scan, cross sectional view. (C) Computed tomography scan, cross sectional view with lung window.

Blood investigations showed microcytic anaemia with thrombocytosis, raised serum IgE levels, and negative delta 508 mutations performed for cystic fibrosis. Nasal swab polymerase chain reaction (PCR) was positive for respiratory syncytial virus. The chest radiograph showed bilateral perihilar infiltrates with inhomogeneous shadowing in the left lower zone, suggesting infective aetiology (Figure 1). His previous workup included a swallowing study done for gastroesophageal reflux disease and trachea-esophageal fistula, which was negative. He was given nebulization, intravenous fluids, antibiotics and put on a non-invasive hi-flow oxygen device, and shifted to the peadiatric intensive care unit (ICU) to continue management and supportive care.

Given the history of repeated chest infections, we performed computed tomography (CT) scan with virtual bronchoscopy under sedation which showed a right aberrant bronchus arising from the supra-carinal trachea, supplying the posterior segment of the right upper lobe with a consolidation patch in the posterior segment of right upper lobe. This was deemed as the most likely cause of recurrent chest infections, mild atelectasis in bilateral lower lobes and sub-segmental atelectasis in left upper lobe (Figure 2).

Radiology and cardiothoracic surgery teams were taken on board to establish and address this rare finding. Surgery was offered to the family, which they refused. The patient gradually showed improvement, weaned off the oxygen support, and was discharged home after clinical improvement. The child continuously followed up with the care provider. He is still on symptomatic treatment, and his general well-being has significantly improved with symptomatic care and patient/parental education.

DISCUSSION

We report a rare case of tracheal (aberrant) bronchus originating from the right wall of the trachea in an 8-month infant admitted to our hospital who presented with recurrent chest infections and had been repeatedly treated as pneumonia.

Tracheal bronchus, sometimes referred to as "pig bronchus" is more prevalent in sheep, swine, and cattle, with a diameter ranging from 0.5 to 1.0 cm. It typically appears as supernumerary single segmental bronchus or a whole lobar bronchus, supplied by a distinct blood supply than the remainder of the lung. It is more prevalent in males and is often incidentally discovered through bronchoscopy or radiologic examination.[2] An obstructed tracheal bronchus can impede lung ventilation, resulting in recurrent atelectasis, pneumonia, chronic bronchitis, and bronchiectasis. It may also occasionally manifest during troubled intubation or intraoperative hypoxemia. This patient also had recurrent episodes of pneumonia with sub-segmental atelectasis in the left upper lobe, necessitating multiple hospital admissions.

Studies indicate that a tracheal bronchus often coexists with various congenital malformations, including rib and vertebral anomalies, tracheal stenosis, duodenal and laryngeal webs, pectus excavatum, oesophageal atresia, VATER complex, hypoplastic lungs, or congenital heart disease.[3] Notably, these patients frequently exhibit cardiac manifestations in conjunction with other anomalies. Kairamkonda *et al*. reported a 5-month infant with VACTERL anomaly and patent ductus arteriosus (PDA) incidentally diagnosed with a tracheal bronchus due to persistent hyperinflation of the right upper lobe on chest radiographs.[3] Similarly, Al-Naimi *et al*. showed that up to 80% of their 20-patient cohort with tracheal bronchus had an associated congenital malformation and 40% exhibited cardiac manifestations.[4] Fortunately, this patient showed no associated congenital or cardiac anomalies and had a normal left main bronchus.

Additionally, a few cases in the literature suggest a potential link between accessory bronchus and lung cancer. However, with only a handful of case reports, there is a lack of sufficient evidence to prove a causal relationship between a tracheal bronchus and malignancy.[2,5]

Identifying a tracheal bronchus is challenging for any clinician due to its rarity. Until a diagnosis is made, most complications have occurred, indicating that tracheal bronchus should always be on the list of differential diagnoses when a patient present

with recurrent chest infections. A high-quality chest x-ray or CT scan can sometimes identify tracheal bronchus and help aid in timely diagnosis. CT scan has been shown to be beneficial in clearly identifying airway abnormalities with good results.[6] Both two-dimensional (2-D) and three-dimensional (3-D) CT scans can accurately identify the location and extent of pathologies in the central airway. A 2-D multi-detector CT scan (MDCT) is a gold standard for diagnosing congenital tracheobronchial abnormalities. The CT scan's 3-D reconstruction technique is dependable for comprehensive pathology evaluation. In this case, we opted for a CT scan to rule out any anatomical and structural anomalies or pathologies in lung parenchyma, such as cystic adenoid malformations, interstitial lung disease, or congenital anomalies in the tracheobronchial tree as a cause of recurrent infections.

The management of tracheal bronchus is challenging and requires a multidisciplinary approach. It is usually based on the severity of the symptoms. Symptomatic management is currently the mainstay of treatment. Most symptomatic cases are managed with bronchodilators, inhaled corticosteroids, anti-muscarinic agents, and antibiotics.[2] In asymptomatic cases, identified early and not compromising patient's quality of life, observation, patient, and parental education play a crucial role in maintaining pulmonary health and preventing chronic inflammation. Early and active management of pulmonary infections, appropriate vaccination, and chest physiotherapy help improve airway-clearing mechanisms and prove an efficient therapeutic approach.[7] Extra care should be exercised when intubating these patients. For a patient with known tracheal bronchus, anaesthesia and intubation should be undertaken carefully with a short endotracheal tube (ETT) and keeping the tip of ETT in the mid to proximal trachea. Thus far, quality data in the form of observational studies on managing tracheal bronchus is limited in the literature due to rare presentation and restricted sample size. When a child presents with symptoms that are refractory to medical management or with bronchiectasis or bronchial stenosis, surgical intervention, in the form of resection of anomalous bronchus or lobe, should be offered to patients. However, the extent of surgical management is not well established in the literature due to the lack of evidence in the form of quality randomised control trials (RCTs). Although lobectomy and segmentectomy have proven to be curative, data to compare surgical outcomes for tracheal bronchus in children is scarce, with only a few case series and case reports describing surgery performed in adults.[6-9]

In conclusion, tracheal bronchus, though rare, can be a critical pathology leading to potential consequences if not promptly diagnosed in children. The recurrent and chronic chest infections, especially with right lobe involvement, should raise suspicion for an ectopic bronchus. Clinicians should not hesitate to resort to a CT scan or bronchoscopy to confirm the diagnosis where necessary, as these modalities could aid in a definitive diagnosis and prove to be lifesaving.

PATIENT'S CONSENT:
Informed consent was obtained from the parents of the patient to publish this case report.

COMPETING INTEREST:
All authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
AA: Conception and drafting of the manuscript.
AAR: Drafting and revising the manuscript.
AA, QA, NUR: Drafting manuscript, critical review, and revision.
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