Airway-Dominant Relapsing Polychondritis Mimicking a Subglottic Tumor in an Adolescent: A Diagnostic Pitfall

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Relapsing polychondritis (RP) is a rare systemic inflammatory disorder characterized by recurrent chondritis of cartilaginous and proteoglycan-rich structures, most notably the auricular and nasal cartilage and the tracheobronchial tree. Although it is predominantly described in middle-aged adults, onset in children or adolescents is uncommon.[1] Airway involvement represents the most severe phenotype and may lead to laryngotracheal stenosis, respiratory failure, and the need for surgical intervention. When the disease presents with non-specific otolaryngological features—such as hoarseness, aphonia, stridor, or subglottic narrowing—diagnosis is often delayed and may be mistaken for laryngitis, asthma, vocal fold dysfunction, post-intubation stenosis, vasculitis, or laryngeal neoplasm.[1-4] This case highlights the diagnostic challenges of airway-dominant RP in a young patient, particularly when subglottic involvement mimics a space-occupying lesion and transient auricular chondritis is not identified during earlier evaluations.

We report a 16-year-old girl with no known autoimmune history who was evaluated at three healthcare facilities before RP was diagnosed. Her illness began with a 2-month history of dry cough, followed by progressive hoarseness, aphonia, and ocular redness. At the first hospital, the presentation was attributed to a common otolaryngological or respiratory condition; no active auricular chondritis was documented. Flexible laryngoscopy revealed bilateral vocal fold edema without a tumor-like lesion (Figure 1a). Neck computed tomography (CT) demonstrated diffuse bilateral laryngeal wall thickening with luminal narrowing to approximately 4 mm, without an intraluminal mass (Figure 1b, c). She was treated empirically with antibiotics and corticosteroids, with partial improvement but no definitive diagnosis.

Several weeks later, she was admitted to a second hospital with acute laryngeal dyspnea requiring difficult intubation, bag-mask ventilation, and intensive monitoring. Procalcitonin was low, and chest radiography showed no consolidation; she was managed for tracheobronchitis with antibiotics and methylprednisolone before transfer to a tertiary referral center. Airway assessment confirmed diffuse laryngeal edema, vocal fold thickening with restricted abduction, and consequent glottic-subglottic stenosis (Figure 1d-f). She underwent tracheostomy. Laryngeal culture isolated *Staphylococcus aureus*. Although concurrent antibiotic therapy was administered, the partial and non-sustained response suggested secondary colonization rather than primary infection. Following combined corticosteroid and antibiotic therapy, she stabilized and was discharged with a tracheostomy tube in situ.

Persistent airway dysfunction was noted after discharge. Flexible laryngoscopy demonstrated severely restricted bilateral vocal fold abduction with a patent trachea below the tracheostomy tube. Paradoxical vocal fold motion and myasthenia gravis were considered; however, repetitive nerve stimulation showed no decremental response, and myasthenia serology was negative, effectively excluding a neuromuscular etiology. Follow-up neck CT incidentally identified a supraglottic-to-subglottic space-occupying lesion, raising concern for a subglottic tumor. Suspension laryngoscopy confirmed glottic-subglottic edema and stenosis (Figure 1g-i), and biopsy of the subglottic region showed only fibrotic tissue with no malignancy.

In this context, granulomatosis with polyangiitis (GPA) was considered. Both cytoplasmic anti-neutrophil cytoplasmic antibodies (ANCAs) and perinuclear ANCA were negative, there was no pulmonary or renal involvement, and histology showed no necrotizing granulomatous

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inflammation, making GPA unlikely. The diagnostic turning point came during the rheumatology assessment. Examination revealed bilateral auricular cartilage erythema and tenderness with bilateral conjunctivitis (Figure 1j, k), whereas nasal cartilage showed no tenderness or deformity (Figure 1l). Targeted history-taking elicited multiple prior self-resolving episodes of bilateral auricular pain and erythema that had not been reported during previous encounters. The combination of recurrent auricular chondritis, bilateral conjunctivitis, and persistent laryngeal-subglottic disease fulfilled three of the six McAdam criteria for RP.[5,6] The RP disease activity index was 24, driven by airway involvement, auricular chondritis, and elevated inflammatory markers.[7] At diagnosis, C-reactive protein was 117.3 mg/L, and the erythrocyte sedimentation rate was elevated. Treatment was initiated with prednisolone (1 mg/kg/day) and azathioprine (50 mg twice daily). After 1 month, auricular inflammation had subsided, respiratory symptoms had improved, and the patient was able to phonate with digital occlusion of the tracheostomy tube; she remains under surveillance for potential decannulation. The diagnostic and therapeutic course is summarized in Supplementary Figure 1.

The diagnostic challenge arose not from a lack of clues but from their asynchronous and fragmented presentation across successive encounters in different specialties. Ocular redness, self-limiting auricular chondritis, and progressive glottic-subglottic stenosis

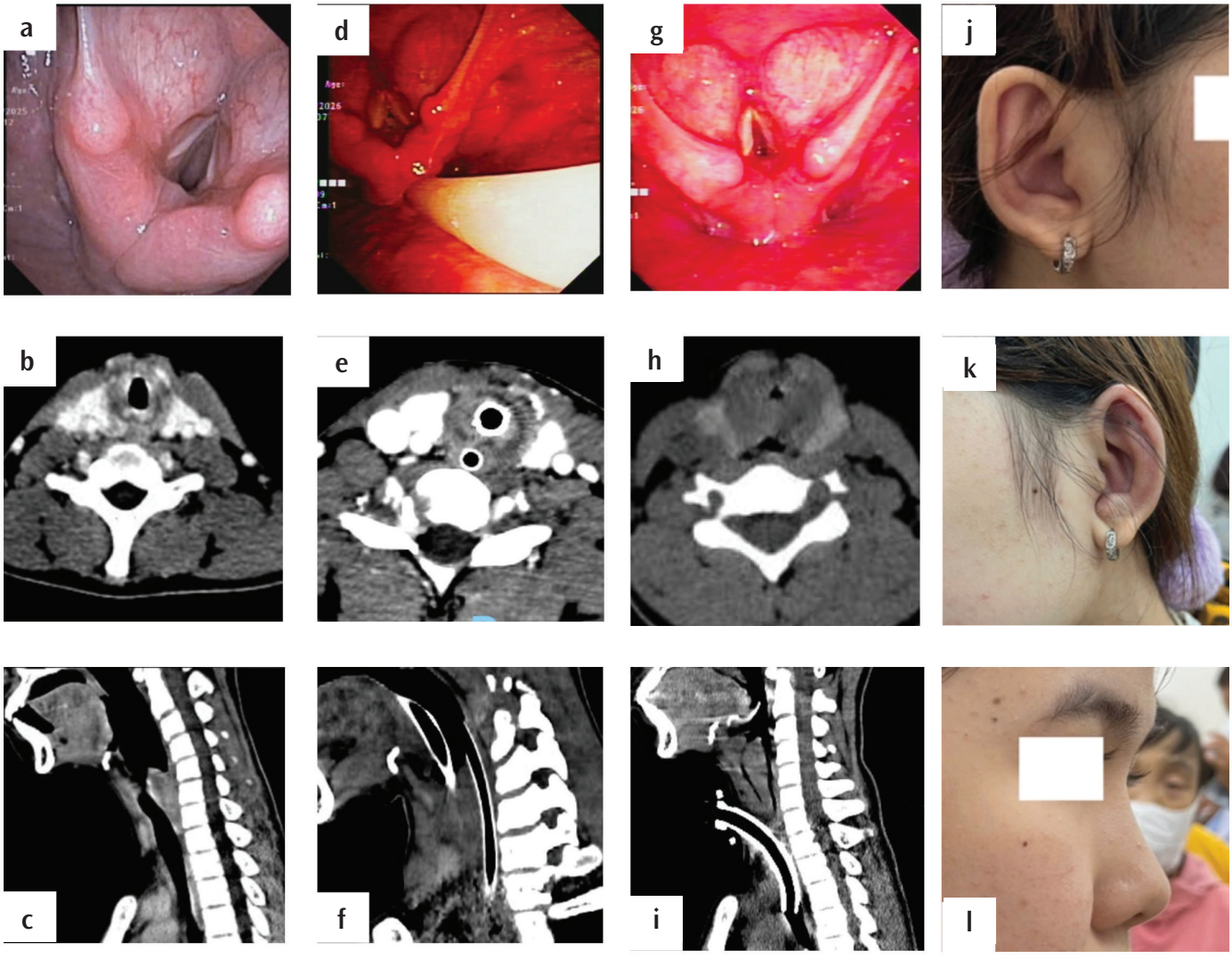

FIG. 1. Sequential endoscopic, radiological and clinical findings during the diagnostic course. (a) Initial laryngoscopy at the first hospital showing diffuse vocal fold edema without an apparent tumor. (b, c) Initial neck computed tomography (CT) at the first hospital showing laryngeal wall thickening and marked airway narrowing without an intraluminal mass. (d) Laryngoscopy during the acute airway episode following difficult intubation and tracheostomy, showing severe laryngeal edema and glottic narrowing. (e, f) Neck CT during the same acute episode showing laryngeal wall thickening and airway narrowing. (g) Follow-up laryngoscopy when a subglottic tumor was suspected, showing persistent glottic-subglottic edema and stenosis. (h, i) Neck CT at the tumor-mimic stage showing a supraglottic-subglottic mass-like stenotic lesion. (j, k) Bilateral auricular chondritis detected at rheumatology assessment. (l) Normal nasal cartilage appearance.

were initially interpreted in isolation. Integrating the ocular, cartilaginous, and airway findings reframed the diagnosis from a localized laryngeal disorder to airway-dominant RP. Airway involvement occurs in approximately 30-60% of patients with RP and is associated with poor prognosis.[1,3,4,8] In children and adolescents, RP remains rare; stridor, hoarseness, or subglottic stenosis may precede recognition of auricular or nasal chondritis.[9,10]

The most distinctive feature was a tumor-like inflammatory-fibrotic subglottic lesion. The clinicoradiological mismatch between a mass-like laryngeal lesion suggestive of neoplasm (Figure 1g-i) and a histopathologically non-malignant biopsy should prompt reconsideration of a chronic inflammatory airway process. In a series of 173 patients, laryngeal CT abnormalities were present in 41.1% of assessed patients, and 66% had subclinical airway involvement on CT.[2] RP is well recognized as a mimic of tumor-like or malignant airway lesions.[11] Fibrosis alone is not diagnostic of RP; however, in the appropriate clinical context—recurrent auricular chondritis, persistent glottic-subglottic disease, non-malignant biopsy findings, and exclusion of key differentials—this finding is more consistent with chronic inflammatory–fibrotic airway disease than with neoplasm.

GPA remains an important differential diagnosis in subglottic stenosis. Although negative ANCA serology does not definitively exclude ANCA-associated vasculitis, the absence of pulmonary-renal involvement, lack of necrotizing granulomatous inflammation, and presence of recurrent auricular chondritis collectively supported RP over GPA.[12,13] This approach aligns with recent guidelines emphasizing that RP diagnosis is based on clinical assessment, organ involvement pattern, and exclusion of mimics rather than any single diagnostic test.[1,14]

From a therapeutic standpoint, this case illustrates the complementary roles of airway intervention and immunosuppressive therapy. Intubation and tracheostomy addressed the acute life-threatening obstruction but did not modify the underlying inflammatory-fibrotic process. The subsequent response to combined corticosteroids and azathioprine, with improvement in both auricular inflammation and respiratory function, suggests that a reversible inflammatory component persisted within the stenotic airway. Given that the evidence base for RP treatment remains largely observational, early recognition of the airway phenotype is particularly important to prevent progression to fixed stenosis or bronchomalacia.[14,15]

Clinical pearl: In an adolescent with persistent laryngeal-subglottic stenosis—especially when imaging suggests a tumor-like lesion but biopsy is non-malignant—the evaluation should not be confined to localized laryngeal pathology. Diagnostic value lies in integrating apparently disparate findings: progressive airway stenosis, prior self-limiting auricular or nasal chondritis, and ocular manifestations. When these features are not temporally concurrent or are not spontaneously reported, targeted history-taking may be the decisive step in reframing the diagnosis from a local laryngeal process to a systemic inflammatory condition requiring immunosuppressive therapy.

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Supplementary Figure: https://balkanmedicaljournal.org/img/files/BalkanMedJ-2026-5-57-supple.pdf

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