

Laryngeal Heterotopic Ossification: An Atypical Etiology of Respiratory Distress

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Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to discuss the work-up, diagnosis and management for heterotopic ossification of the larynx and identify the airway risks associated with this diagnosis.

Objective: Benign bony lesions involving the larynx are rare. The objective is to review the differential diagnosis and management of laryngeal heterotopic bone formation through a case report presenting as acute respiratory distress.

Methods: Patient chart review, intraoperative imaging and radiographic studies are reported along with literature review.

Results: A 46 year old female presented in respiratory distress with a history of progressive hoarseness for one year and intermittent stridor for six months, worsened with activity. Her symptoms failed to improve with initial treatment as asthma exacerbations. Flexible transnasal laryngoscopy revealed an immobile right vocal cord. CT imaging demonstrated widespread ossification resulting in fusion of the hyoid bone, thyroid cartilage, cricoid cartilage and right arytenoid cartilage. A tracheostomy under local anesthesia was performed to secure her airway. Further workup included Tc-99m MDP SPECT-CT scanning and serum alkaline phosphatase measurement.

Conclusions: This case report presents a rare etiology for respiratory distress and highlights the principles of work-up and management of heterotopic ossification. Management for laryngeal ossification should include consideration for the more common oncologic etiologies. This benign case may represent a novel presentation of tracheopathia osteochondroplastica.

Case Report

A 45 year old female presented through the emergency room with a history of asthma and Crohn's disease for progressive hoarseness over one year and worsening stridor over 6 months. The hoarseness and stridor were worsened with prolonged talking and physical activity. Breathing treatments and rescue inhalers for her asthma did not improve her symptoms. Report from a local otolaryngology evaluation demonstrated left vocal cord paresis and right cord paralysis prompting transfer to our center. Pulmonary function testing showed a fixed extrathoracic obstruction pattern and decreased PIF. A neck CT demonstrated fusion of the hyoid to the thyroid cartilage, right cricoarytenoid fusion, bilateral cricothyroid fusion and a bony spur into the anterior commissure from the thyroid cartilage. Tracheal calcification was identified during tracheostomy done under local anesthesia. Conversion to general anesthesia permitted direct laryngoscopy which confirmed the absence of mucosal lesion. The left vocal fold was mobile to palpation, but the right fold was fixed. A preliminary diagnosis of tracheopathia osteochondroplastica was made. Rheumatology consultation was subsequently performed, ruling out relapsing polychondritis, granulomatosis with polyangiitis, and lupus. Multidisciplinary Tumor Board discussion recommended a 3-phase bone scan and SPECT-CT which confirmed active heterotopic ossification in the larynx. She has been followed for 18 months with two subsequent bone scans showing mild decline in laryngeal uptake.

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Introduction

Tracheopathia osteochondroplastica (TPO) is a rare benign disease of the trachea and bronchi, characterized by submucosal cartilaginous or osseous nodules protruding into the airway, arising from the underlying cartilage¹. The vast majority of cases are asymptomatic and are incidentally identified on bronchoscopy, radiographic exam or on autopsy². Those whose disease is severe enough to cause symptoms may present with cough, hoarseness or dyspnea with exertion. This process classically affects the distal airway but has included subglottic involvement in some reports³.

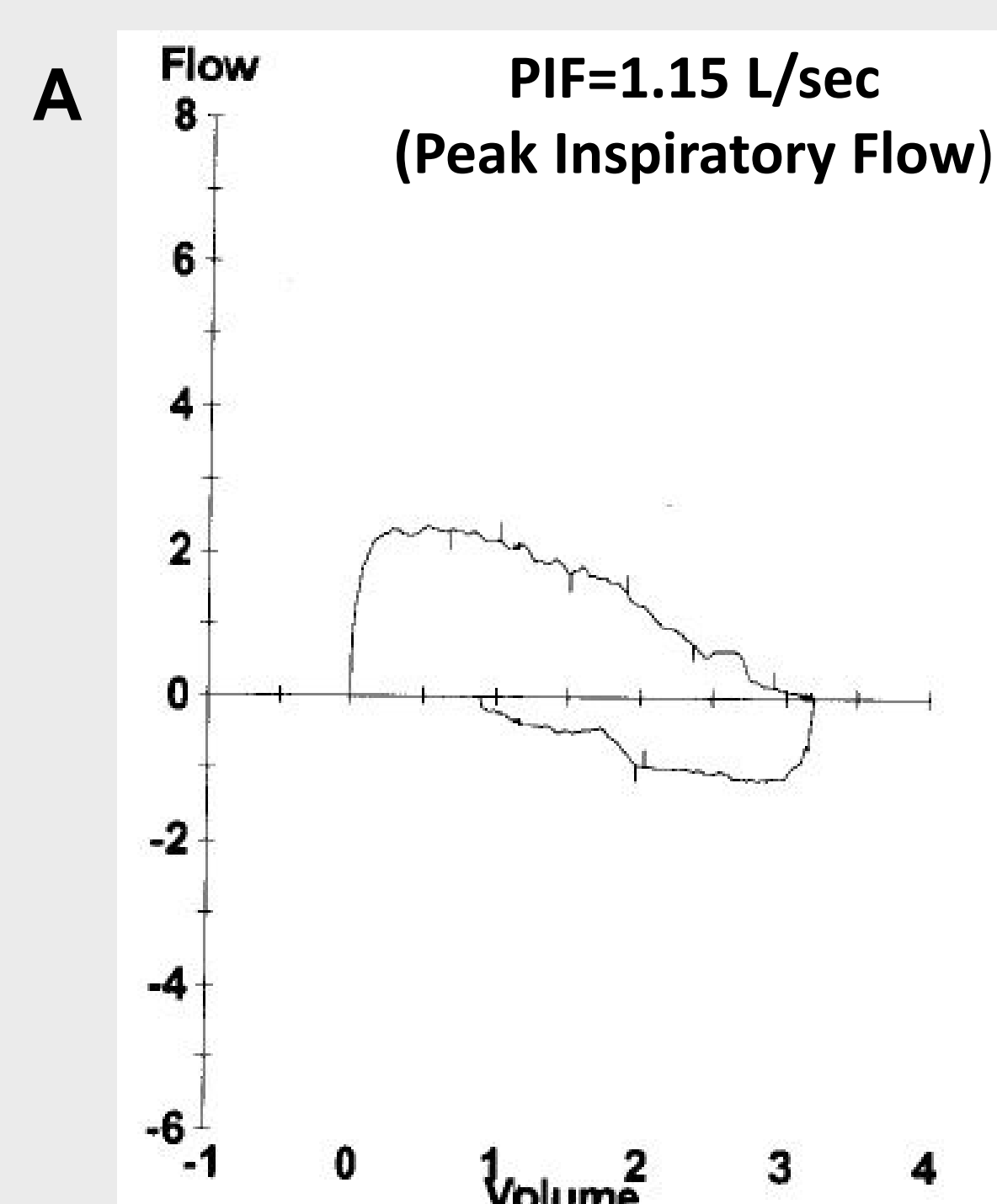


Figure 1. A) Flow volume loop on initial presentation demonstrating fixed obstructive pattern. **B)** Distal-chip flexible laryngoscopy demonstrating paralyzed right true vocal cord.

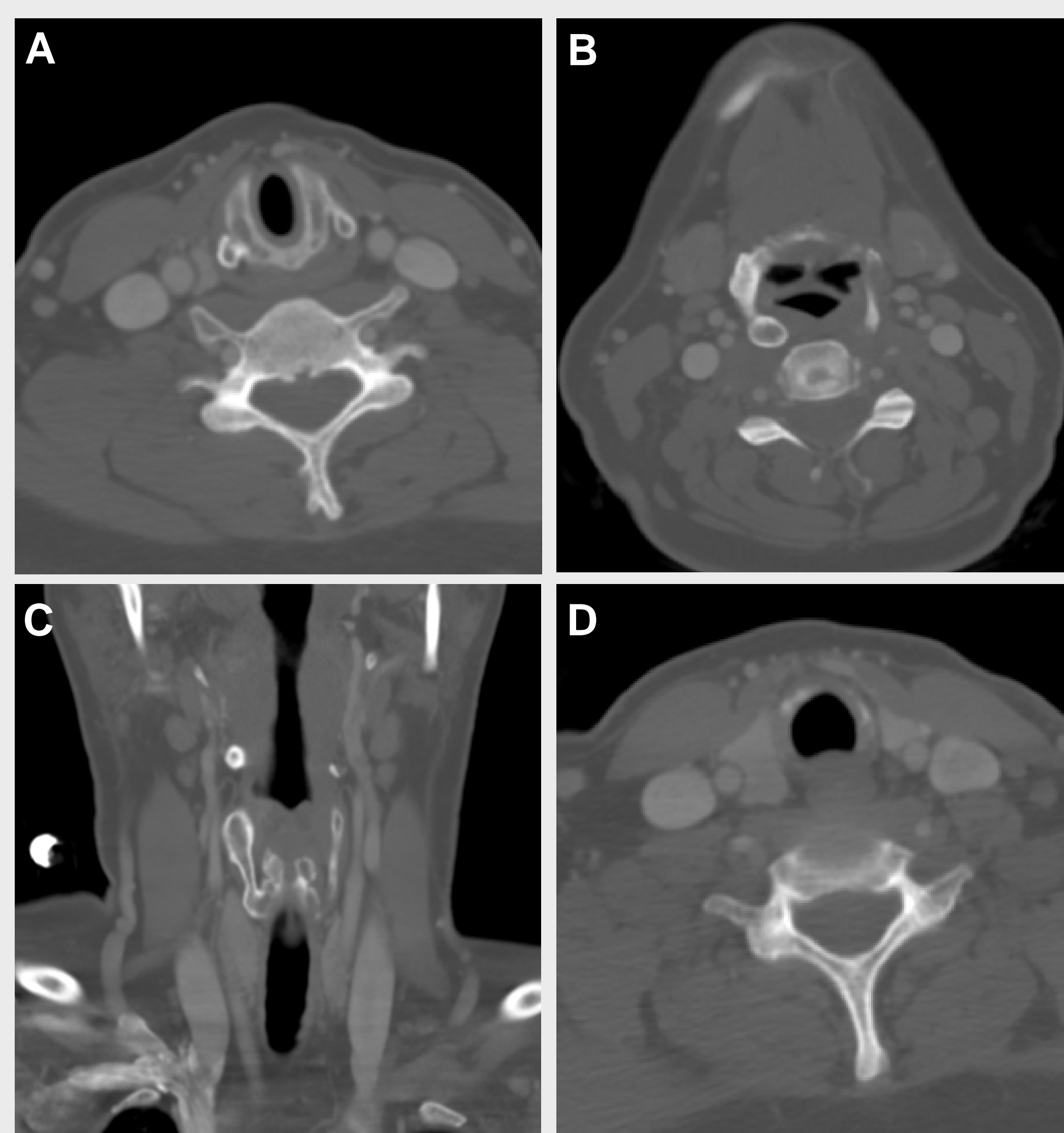


Figure 2. CT neck/larynx demonstrating ossification at the cricothyroid joint (A), from the greater cornu of the thyroid to the hyoid (B), at the thyroarytenoid joint (C), and along the anterior tracheal wall (D).

Discussion

Heterotopic ossification is a concerning finding when identified in the patient with airway symptoms. Not only could its presence act as a potential obstruction to securing the airway, but it may be a sign of an underlying malignancy – particularly chondrosarcoma⁴. The heterotopic ossification characterized by tracheopathia osteochondroplastica is not considered malignant but may be extensive enough compromise the airway – most commonly due to nodular incursion into the tracheal lumen and not due to obstruction in the endolarynx. Extratracheal presentation of this rare disease has been documented by Nielsen et al (2015) who noted a mixed osseous and cartilaginous projection from the thyroid cartilage into the laryngeal vestibule¹. Although it was not specifically identified as TPO, another report of an “ossified spur” from the thyroid cartilage into the anterior commissure described by Cantarella et al (2013) may represent an additional extratracheal presentation⁵. We propose that our case presentation represents a severe case of extratracheal TPO, with cartilaginous and osseous changes arising from several subsites of the thyroid cartilage including the thyroarytenoid joint, the cricothyroid joint and the internal surface of the thyroid lamina.

Work-up of this lesion included tracheostomy to secure the airway given the patient's respiratory distress on physical exam and documented by peak inspiratory flow⁶. Serial 3-phase bone scans with SPECT-CT were performed to follow the metabolic activity of the heterotopic ossification in accordance with published guidelines⁴. Additionally, serum alkaline phosphatase (ALP) was obtained at the time of our second bone scan after identifying this lab as a potential marker of heterotopic ossification progression^{4,7}. ALP was within normal limits and calcium was only mildly elevated in this case. Elevated ALP has been previously documented in active heterotopic ossification, but cases have been reported with normal ALP⁸. A rheumatology excluded other atypical causes of laryngeal pathology including relapsing polychondritis and granulomatosis with polyangiitis.

Conclusions

Heterotopic ossification involving the larynx is an atypical etiology for airway distress. It is important to rule out malignancy as the underlying cause of the ossification.

This case report failed to identify an associated pathologic process that would contribute to the presence of heterotopic ossification. TPO, with multiple sites of involvement, was left as the diagnosis of exclusion. Whereas TPO typically involves the lower trachea, this case and selected reports demonstrate extratracheal presentation of this pathology in rare occurrences.

Serial bone scans presented in our case showed gradual decline of radioisotope uptake. This finding generally correlates with maturing of ossification. As of the most recent July 2015 scan, maturation was not yet complete. The patient presented is currently at 18 months following diagnosis and has another 3-phase bone scan planned.

References

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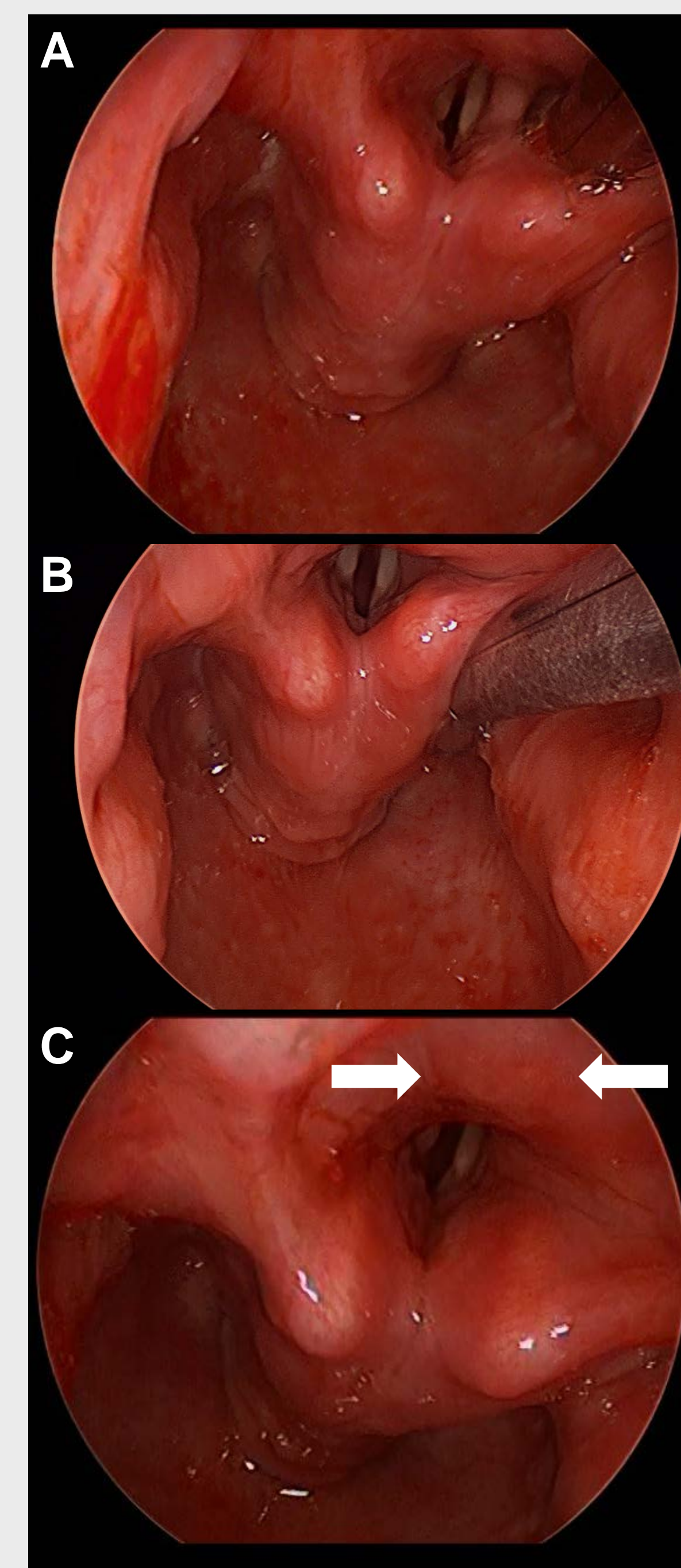


Figure 3. A,B) Direct laryngoscopy confirming fixation of the right cricoarytenoid joint by palpation. **C)** Supraglottic anterior commissure fullness from underlying ossification (arrow).

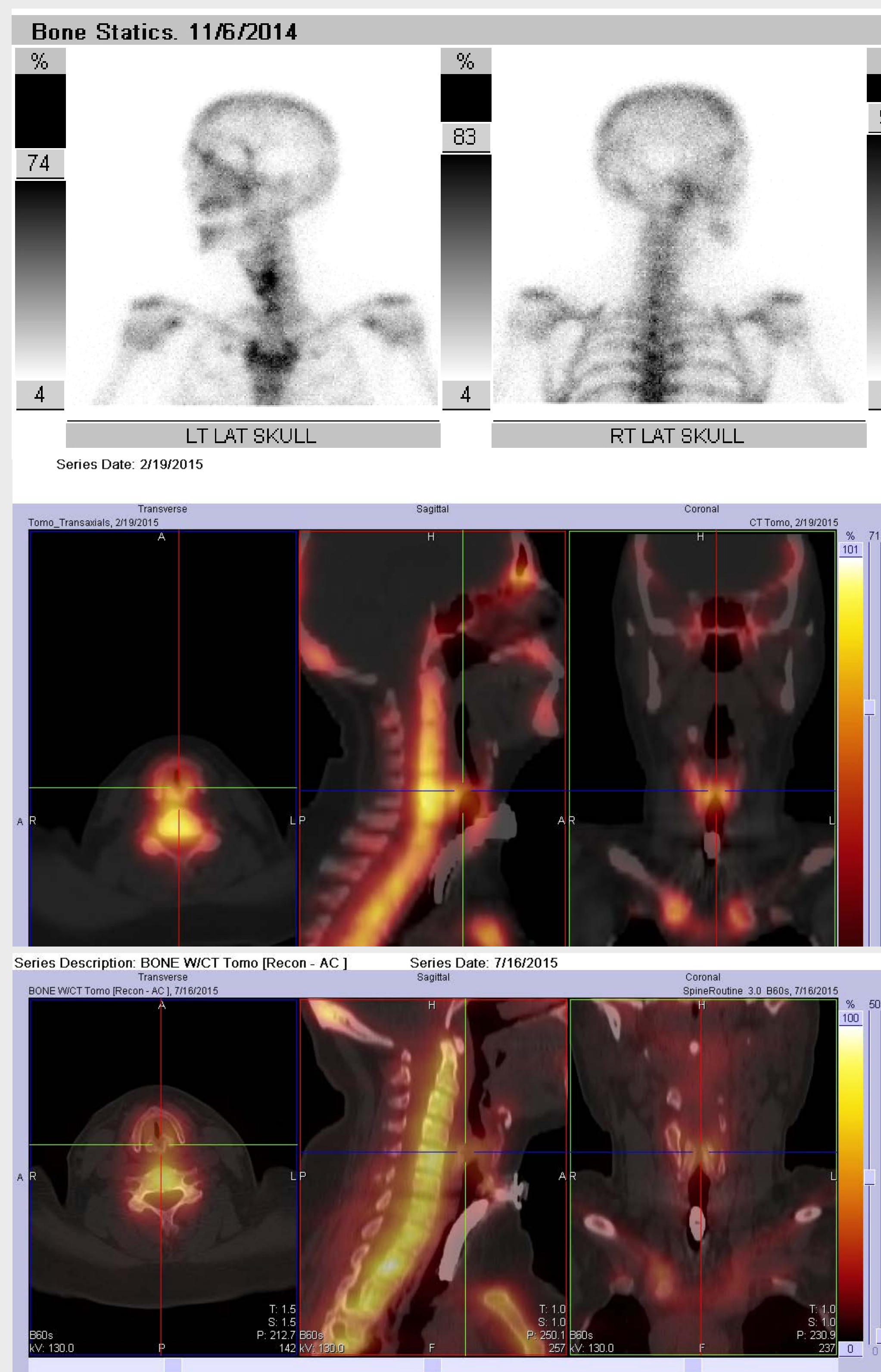


Figure 4. 3-phase bone scan (top) and serial SPECT-CT sequences (middle, bottom) demonstrating a slow decline in heterotopic ossification at multiple laryngeal subsites.