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Images of Interest / Imagens de Interesse

Fatal Tracheal Stenosis in Relapsing Polychondritis

Estenose Traqueal Fatal em Policondrite Recidivante

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Abstract

Relapsing polychondritis is a rare autoimmune disorder characterized by recurrent episodes of cartilaginous inflammation with subsequent degeneration, structural loss, and fibrosis. We present the case of a 33-year-old female patient diagnosed with relapsing polychondritis, who developed hoarseness, dry cough, and dyspnea. Bronchoscopy revealed stenosis in the subglottic region and reduced vocal cord mobility. The patient developed severe glottic and subglottic edema, making orotracheal intubation impossible. An emergency cricothyroidotomy was performed, followed by a tracheostomy. A chest computed tomography revealed extensive thickening and calcification of the thyroid and cricoid cartilages, in addition to subglottic stenosis. Ultimately, the patient succumbed to her illness.

Keywords

Tracheal stenosis; Relapsing polychondritis; Computed tomography.

Resumo

A policondrite recidivante é uma doença autoimune rara, caracterizada por episódios recorrentes de inflamação cartilaginosa, com subsequente degeneração, perda estrutural e fibrose. Apresentamos o caso de uma paciente de 33 anos diagnosticada com policondrite recidivante, que desenvolveu rouquidão, tosse seca e dispneia. A broncoscopia revelou estenose na região subglótica e redução da mobilidade das cordas vocais. A paciente desenvolveu edema glótico e subglótico grave, impossibilitando a intubação orotraqueal. Foi realizada uma cricotireoidotomia de emergência, seguida de uma traqueostomia. Tomografia computadorizada do tórax revelou extenso espessamento e calcificação das cartilagens tireoide e cricoide, além de estenose subglótica. Por fim, a paciente sucumbiu à doença.

Palavras-chave

Estenose traqueal; Policondrite recidivante; Tomografia computadorizada.

Fatal Tracheal Stenosis in Relapsing Polychondritis

A 33-year-old female patient was diagnosed 15 years earlier with relapsing polychondritis (RP), initially presenting with hoarseness, dysphonia, dry cough, and exertional dyspnea. She also had retroauricular bulging on the left and right vocal fold paresis. A biopsy of the left auricle was diagnostic. The patient received treatment and was advised to follow up in an outpatient setting, where she maintained good symptom control. In a recent evaluation, however, she reported new symptoms of hoarseness, dry cough, and dyspnea. She also had stridor on auscultation and glottic and subglottic stenosis. Her medical team interpreted these symptoms as signs of disease reactivation, posing a risk of glottal and subglottic edema. Given the potential severity of the condition, the patient was referred to the emergency department and began pulse therapy with methylprednisolone + maintenance corticosteroids after pulse therapy. Etanercept and methotrexate were also used, which led to significant improvement.

Bronchoscopy revealed severe concentric stenosis in the subglottic region and reduced vocal cord mobility; the distal trachea and carina appeared normal. Laryngeal cartilage biopsy confirmed the diagnosis. Immediately after the procedure, the patient developed severe glottic and subglottic edema accompanied by stridor and respiratory distress. Despite high-dose hydrocortisone administration, the edema persisted, making orotracheal intubation impossible despite multiple attempts by the anesthesia team. The patient was in hypoxic cardiopulmonary arrest for 8 minutes. Given the critical situation, an emergency cricothyroidotomy was performed, followed by a tracheostomy in the operating room due to evidence of tracheal thickening and narrowing.
A chest computed tomography (CT) examination conducted after clinical stabilization revealed extensive thickening and calcification of the thyroid and cricoid cartilages, anterior tracheal wall, and main bronchi, in addition to subglottic stenosis above the tracheostomy site, consistent with the bronchoscopy findings. The patient subsequently developed multiple episodes of ventilator-associated pneumonia, culminating in septic shock. Despite the early initiation of broad-spectrum antibiotic therapy, she had labile blood pressure and remained dependent on vasoactive agents. Acute renal and hepatic dysfunction developed, likely secondary to septic shock and tissue hypoperfusion. Ultimately, the patient succumbed to her illness.
RP is a rare autoimmune disorder characterized by recurrent episodes of cartilaginous inflammation with subsequent degeneration, structural loss, and fibrosis. It results in the destruction of cartilage in the ears, nose, joints, and upper airways, including the larynx and subglottic trachea. The diagnosis of RP is based on clinical evidence, imaging findings, and, rarely, the biopsy of involved cartilage, as no specific laboratory test is diagnostic. Clinically, RP is

diagnosed when three or more of the following features are present: bilateral recurrent auricular chondritis, non-erosive seronegative inflammatory polyarthritis, nasal chondritis, ocular structure inflammation, respiratory tract chondritis, and cochlear or vestibular damage.[1,2]

The best non-invasive method used to diagnose RP is computed tomography (CT), with end-inspiratory and dynamic expiratory scans. Post-processing of CT images with 3D may facilitate the visualization of stenosis and size reduction for interventional endoscopists, but their added value for the detection of bronchial abnormalities has not been demonstrated. Other imaging modalities have been described including magnetic resonance imaging and positron emission tomography scan. However, they have not been widely used on a routine basis. Bronchoscopy may be performed in RP patients who experience respiratory symptoms to assess mucosal inflammation as a sign of active disease, define the severity of airway involvement, and allow dynamic assessment of potential airway collapse. CT scan of our patient revealed tracheobronchomalacia and thickening of the anterior tracheal wall and/or bronchi, with or without calcification, and airway stenosis.[1,2] In cases of subglottic involvement, CT may show thickening, edema, and destruction of the laryngeal cartilages. Laryngotracheal involvement is among the most common causes of death among patients with RP, with pulmonary infection with airway stenosis or collapse being the main contributing factor. Airway obstruction may occur via several mechanisms, including inflammatory swelling, fibrous mass formation with cicatricial narrowing, and laryngotracheal cartilage destruction with subsequent airway collapse during respiration.(Fig. 1)[1,2,3]

Most of the therapies currently used are derived from research and recommendations for other autoimmune disorders and include the use of non-steroidal anti-inflammatory drugs, glucocorticoids, disease-modifying antirheumatic drugs, and biologic monoclonal antibodies, such as TNF-alpha and IL-6. The prognosis for relapsing polychondritis is highly variable, ranging from mild, self-limited chondritis to severe life-threatening forms. The differential diagnosis of RP is broad and includes several conditions that can mimic its clinical or radiological presentation. In the present case, with thickening of the tracheal wall with calcifications, the main differential diagnoses were amyloidosis and osteochondroplastic tracheopathy.

In the present case, the outcome was fatal due to severe airway involvement compounded by multiple secondary infections.

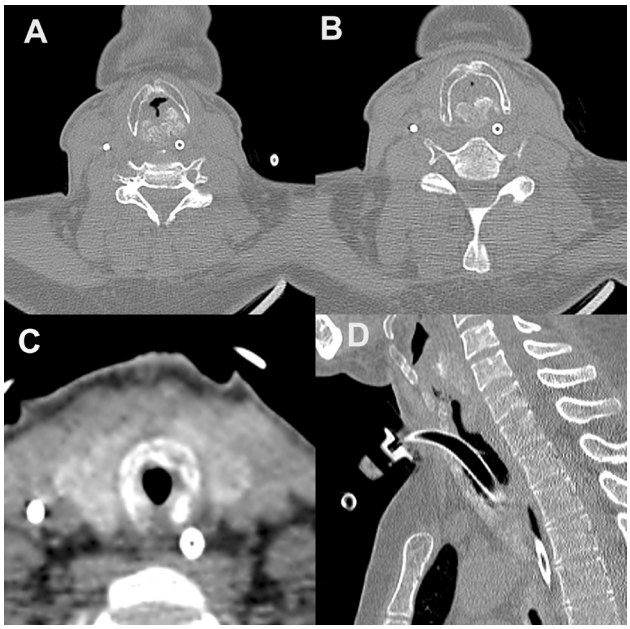

Figure 1 – (A,B,C) axial CT images of the neck demonstrating extensive thickening and calcification of the thyroid and cricoid cartilages, with marked subglottic stenosis. (D) a sagittal reformatted CT image of the upper chest showing calcification along the anterior tracheal wall in addition to the stenosis caused by laryngeal cartilage thickening. The posterior tracheal wall is preserved, as seen in figures 1C and D.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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