Radiological Diagnosis

Diagnosis of the case presented in the previous edition

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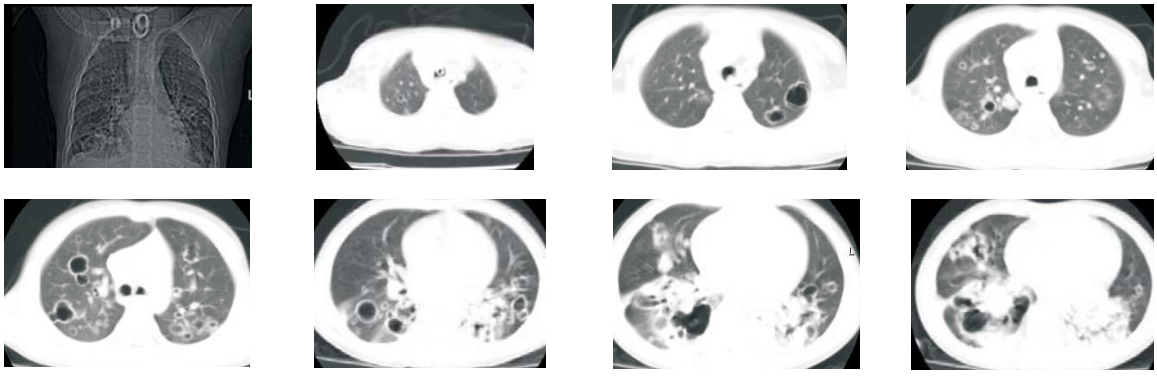
RECURRENT RESPIRATORY PAPILLOMATOSIS WITH PULMONARY INVOLVEMENT



CASE

Female, aged 4 years and 6 months. normal birth
Progressive hoarseness from 8 months of age
Four icu admissions for respiratory insufficiency, submitted to
Tracheostomy at the age of 1 year and 8 months

Recurrent respiratory papillomatosis (RRP), also known as recurrent juvenile papillomatosis, is the most common benign tumor seen in children, with approximately 1500 new cases diagnosed annually in the United States[1]. It is directly related to the human papilloma virus (HPV) subtypes 6 (from 50% to 84% of cases) and 11 (from 25% to 37% of cases). Subtype 11 is more likely to cause bronchial dissemination, relevant obstructive profiles, malignant transformation into spinocellular carcinoma and more aggressive evolution of the disease[2-3].

Dissemination of RRP to the lower airway is uncommon. Involvement of the trachea or proximal bronchi occurs in 5% of cases, and the disease extends to the lungs in only 1%. These complications usually occur from 1 to 11 years after a patient has been diagnosed with RRP, a diagnosis that is made between the ages of 6 months and 14 years[4]. Tracheostomy, which is often necessary due to restrictive respiratory insufficiency, is a significant risk factor for these complications[2].

When there is involvement of the glottic region, the most common initial symptom is hoarseness[2]. As the disease progresses, stridor, airway obstruction, fever, hemoptysis, productive cough and progressive dyspnea may occur[5].

The diagnosis of pulmonary dissemination is based on the child or adolescent having a history of various laryngeal papilloma resections and procedures in order to keep the airway open, and on chest X-rays or computed tomography (CT) scans that present images consistent with the disease[6].

In this case, the chest X-ray reveals the papilloma within the central airway. The typical manifestation in the lungs is a solid or cystic nodule with thin to moderately thick walls (2 to 3 mm), usually in the lower lobes. When detected early, most nodules are small and homogeneous but may grow, forming cavities filled with fluid or with an air-fluid level if there is co-infection. These lesions may grow slowly and may converge[4]. Obstructive atelectasis, pneumatoceles and recurrent consolidations may also be seen[5].

Chest CT scan, especially when using the high-resolution technique, is considered the test of choice for the assessment of RRP pulmonary involvement since it can detect small nodules while still in the initial phases and can better reveal the characteristics of solid or cystic nodules, lung abscesses, pneumatoceles, consolidations, etc.[2]

The main complications of pulmonary involvement by RRP are pneumatocele, which can become infected and fibrotic, and lung abscess, especially that caused by anaerobic bacteria[2].

The treatment of RRP with pulmonary involvement remains a challenge. Antibiotics, surgery, chemotherapy with cyclophosphamide, methotrexate, bleomycin and interferon have all been tested, with no success. Studies of intralesional application of Cidofovir, an antiviral nucleoside analog, seem to bring positive results, but further studies on a larger scale are necessary in order to define its true efficacy[5-6].

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