CASE REPORT

Association of bronchectasies and situs inversus

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Abstract Kartagener’s syndrome comprises a rare clinical triad: nasal polyposis, bronchiectasis and situs inversus. It is a hereditary disease transmitted as an autosomal recessive trait, characterized by a partial or total failure of the eyelashes vibration dampers. The diagnosis is most often made in childhood, but cases have also been described in adulthood. Initially described in 1936, Kartagener’s syndrome is a rare autosomal recessive genetic disease.

We report a case with this syndrome, and we will discuss the epidemiological, clinical, paraclinical and therapeutic characteristics of this syndrome.

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Observation

A 50-year-old with a chronic bronchorrhea for 20 years, was admitted for exacerbation exaggerating his bronchorrhea became purulent with febrile sensations for a ten days. The clinical examination revealed discrete hippocratic fingers, bilateral rales sounding and shock peak seen on the right lung. The chest radiograph (Fig. 1) recorded cystic images and thick walls of hydroaeric levels of the left parahiliar and dextrocardia. Chest computed tomography (Fig. 2) highlighted homes bronchiectasis and bilateral scattered with complete situs inversus. Otorhinolaryngology (ORL) of chest revealed no pituitary mucosa and inflammatory polyps. Echocardiography found situs inversus without vascular malformations. Bronchoscopy showed an inversion of bronchi with a diffused inflammatory appearance and purulent secretions. The search for the Koch bacillus with a culture on Sabouraud medium in liquid bronchial aspirate was negative. The sputum cytobacteriological isolated Haemophilus B lactamase negative. Thus, the patient was treated with Amoxicillin + clavulanique acid 1 g twice daily, kinesitherapy with bronchial drainage. Both an annual influenza vaccination and pneumococcal vaccination every 5 years have been advocated.

Discussion

Kartagener’s syndrome comprises a rare clinical triad: nasal polyposis, bronchiectasis and situs inversus. It is a hereditary disease transmitted as an autosomal recessive trait, characterized by a partial or total failure of the eyelashes vibration dampers. The diagnosis is most often made in childhood, but cases have also been described in adulthood.

Initially described in 1936, Kartagener’s syndrome is a rare autosomal recessive genetic disease. This syndrome is one characterized by primitive ciliary dyskinesia (PCD), which is one of a heterogeneous group of genetic respiratory diseases related to constitutional anomaly of eyelashes, the upper respiratory mucosa (ORL) and lower bronchi [1–3,11]. The prevalence is approximately 1/16,000 for DCP and about 1/32,000...
for the Kartagener syndrome [2,3]. Respiratory symptoms can start early with respiratory distress (50–70%), chronic bronchorrea in childhood, and in adults, the clinical picture shows chronic obstructive pulmonary disease with diffused bronchiectasis or an array of chronic respiratory failure [1,4–7]. At the Sphere otorhinolaryngology, you can see oedematous-purulent sinusitis with nasal polyposis and chronic otitis seromucous. In older children and adults we sometimes observe a hypo or agenesis of the frontal sinus. In principle the situs inversus is complete, but it may be only partial (isolated dextrocardia or isolated transposition of the abdominal viscera). It is frequently ignored and asymptomatic. However, heart defects can be observed in 10% of DCP and 3% of the Kartagener syndrome such as atrial septal defect, hypoplasia of the vascular tree of the right lung, and transposition of the great arteries [2.8–10]. Other abnormalities may be observed, including male infertility or subfertility by immobilizing the spermatozoa [12].

Cases of hydrocephalus and retinitis pigmentosa have also been described. The diagnosis of the Kartagener syndrome is made on the clinical triad and the identification of abnormal ciliary structures and function evidenced in electron microscopic examination and phase contrast microscopy of nasal or bronchial biopsies. The inhalation of saccharin during the test has no diagnostic value orientation [2,3,7,9]. Currently there is no specific etiological treatment. The treatment is based primarily on kinesitherapy of bronchial drainage associated with antibiotic therapy during respiratory superinfection as well as vaccination coverage and cardiac monitoring [1,2,7,8] instead of Thoracic or ORL surgery limited to failure of symptomatic treatment.

Conflicts of interest

None.

References