IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 22, Issue 8 Ser.3(August. 2023), PP 25-28
www.iosrjournals.org

False Asthma Associated With Bronchiolitis Revealing Gougerot-Sjögren'sSyndrome

Wafaa EL OMARI, Hanane CHARAF, Rachida ZAHRAOUI,
Mouna SOUALHI, Jamal Eddine BOURKADI
Pneumology Department, moulayyoussef Hospital, Faculty of Medicine and Pharmacy, Med V University, Rabat, Morocco

Abstract
Bronchiolitis associated with systemic diseases is essentially found in Gougerot-Sjögren's syndrome, rheumatoid arthritis and inflammatory diseases of the digestive tract, its clinical presentation is sometimes very similar to that of asthma, making it a major differential diagnosis. Bronchiolitis can be cellular, often lymphomonocytic, sometimes associated with lymphoid follicles, as in Gougerot-Sjögren's syndrome; it can also be constrictive, with peribronchiolarfibrose, notably in rheumatoid arthritis, and which has a poorer prognosis, likely to progress to chronic respiratory failure. The diagnosis must be made in the presence of any atypical form or difficult asthma, and is based on a meticulous history, including the onset of the disease, the clinical history of symptoms and the search for extra-respiratory signs, such as polyarthralgia or clinical dry syndrome. Chest CT scans, respiratory function tests and immunological assessments are essential, although histological confirmation is rarely necessary.
Treatment combines non-specific measures (bronchodilators, vaccination, respiratory rehabilitation), possibly long-term macrolide therapy, and sometimes corticosteroids with or without immunosuppressants.
The aim of this article is to raise clinicians' awareness of this peculiar form of Gougerot-Sjögren's syndrome presentation mimicking asthma.
Key words*: False asthma, bronchiolitis, Gougerot-Sjögren's syndrome.*

Date of Submission: 29-07-2023 Date of Acceptance: 09-08-2023

I. Introduction:

bronchiolitis is one of the main causes of false asthma [1], it is the most under-reported of the respiratory disorders associated with systemic diseases [2]

The prevalence of bronchiolitis diagnosed by CT evidence of distal bronchial lesions may be as high as 35% for non-smoking patients with rheumatoid arthritis (RA) [3], and is common in Gougerot-Sjögren's syndrome (GSS), demonstrated in 48% to 68% of thoracic CT scans [4], while it is less common in other systemic diseases such as lupus erythematosus, ankylosing spondylitis and systemic sclerosis [5]. Pulmonologists should consider evoking the diagnosis of bronchiolitis in the presence of difficult-to-treat or atypical asthma, associated with extra-respiratory signs (clinical dry syndrome, arthralgia, etc.) and bronchiolar rales on pleuropulmonary auscultation [2]. The aim of this article is to draw the practitioner's attention to this clinical presentation of certain connectivitis, mimicking difficult-to-treat asthma.

We report a case of bronchiolitis during the course of Gougerot-Sjögren's syndrome (GSS), in a patient treated for false asthma.

II. Observation

Patient aged 68, with no particular exposure, no toxic habits, with a family history of atopy, two maternal uncles being treated for asthma. The patient had been treated for uncontrolled asthma for 10 years, on a fixed combination of high-dose inhaled corticosteroid and long-acting bronchodilator, with several exacerbations and emergency room visits. For several years, she had been reporting recurrent episodes of dry cough and mMRC stage II exertional dyspnea with chest wheezing, with xerostomia without xerophthalmia, and inflammatory polyarthralgia, apyrexia and preservation of general condition. Physical examination revealed bilateral sibilant rales, no Raynaud's phenomenon, no skin lesions or joint signs.

The chest X-ray was normal, apart from slight thoracic distension (Figure 1). Chest computed tomography (CT) showed a few bilateral micronodules on axial slices taken in deep inspiration and apnea (Figure 2), with slight air-trapping lesions on expiratory slices (Figure 3), and a normal blood count, blood ionogram, renal and hepatic function were normal, sedimentation rate was elevated to 50 mm at 1 hour,

immunological work-up showed positive anti-nuclear antibodies at a title of 1/1200 , anti-anti Ro52 and anti -SSA antibodies were positive, anti-mitochondria antibodies were positive at high title, and anti DNA, anti-CCP ,rheumatoid factor, anti-SSB, anti-Sm, anti-Scl70, anti-RNP, anti-jo1 antibodies were negative, with negative ANCA, total IgE is normal, aspergillary serology was negative, periungualcapillaroscopy is without abnormality, skin tests showed sensitivity to dermatophagoidespteronyssinus DP and cupressaceae. Biopsy of the accessory salivary glands (ASG) revealed a focal, medium-intensity inflammatory infiltrate of lymphocytes forming a two-point focus, the appearance of chronic sialadenitis compatible with Gougeront-Sjogren's syndrome chisholm and mason grade 4. Ophthalmological examination was normal, and joint X-rays were normal. Respiratory function tests showed obstructive ventilatory disorder, with pulmonary distension.

Diffusion capacity of carbon monoxide (DLCO) was normal, O2 desaturation was absent in the 6-minute walk test with a 360-m walking distance (WD), and trans-thoracic echocardiography was normal.
The diagnosis of Gougerot-Sjögren's syndrome (GSS), with bronchiolar involvement was retained.

The patient was put on oral corticosteroid therapy with prednisone 0.5mg/kg/d for a three-month onset phase, followed by a gradual taper of 10mg every six weeks to half the initial dose, then a taper of 5mg every six weeks to the 10mg dose, which was maintained for three months, then a taper of 2.5mg every six weeks until discontinuation.
Clinical and functional improvement was observed (Figure 4).

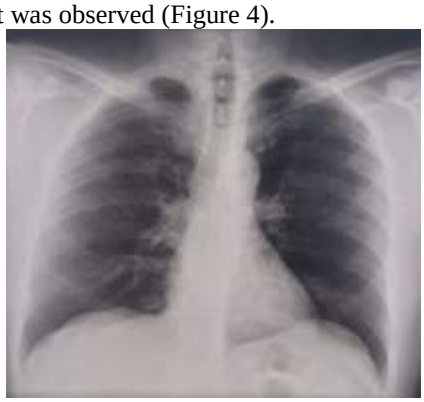


Figure 1:Chest X-ray: Thoracic distension

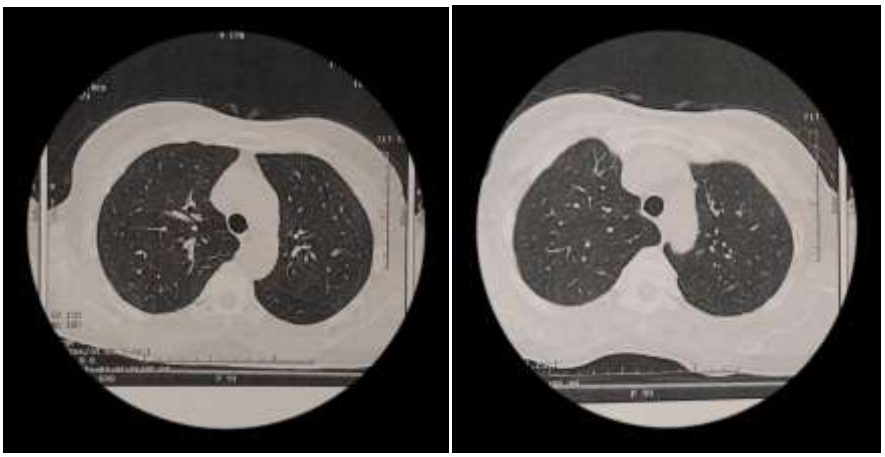


Figure 2: Chest CT in parenchymal window (inspiratory slices): A few bilateral micronodules

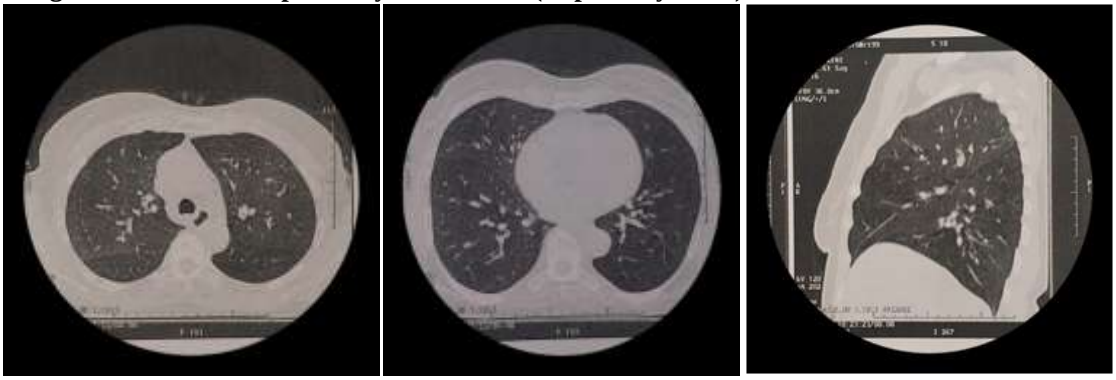
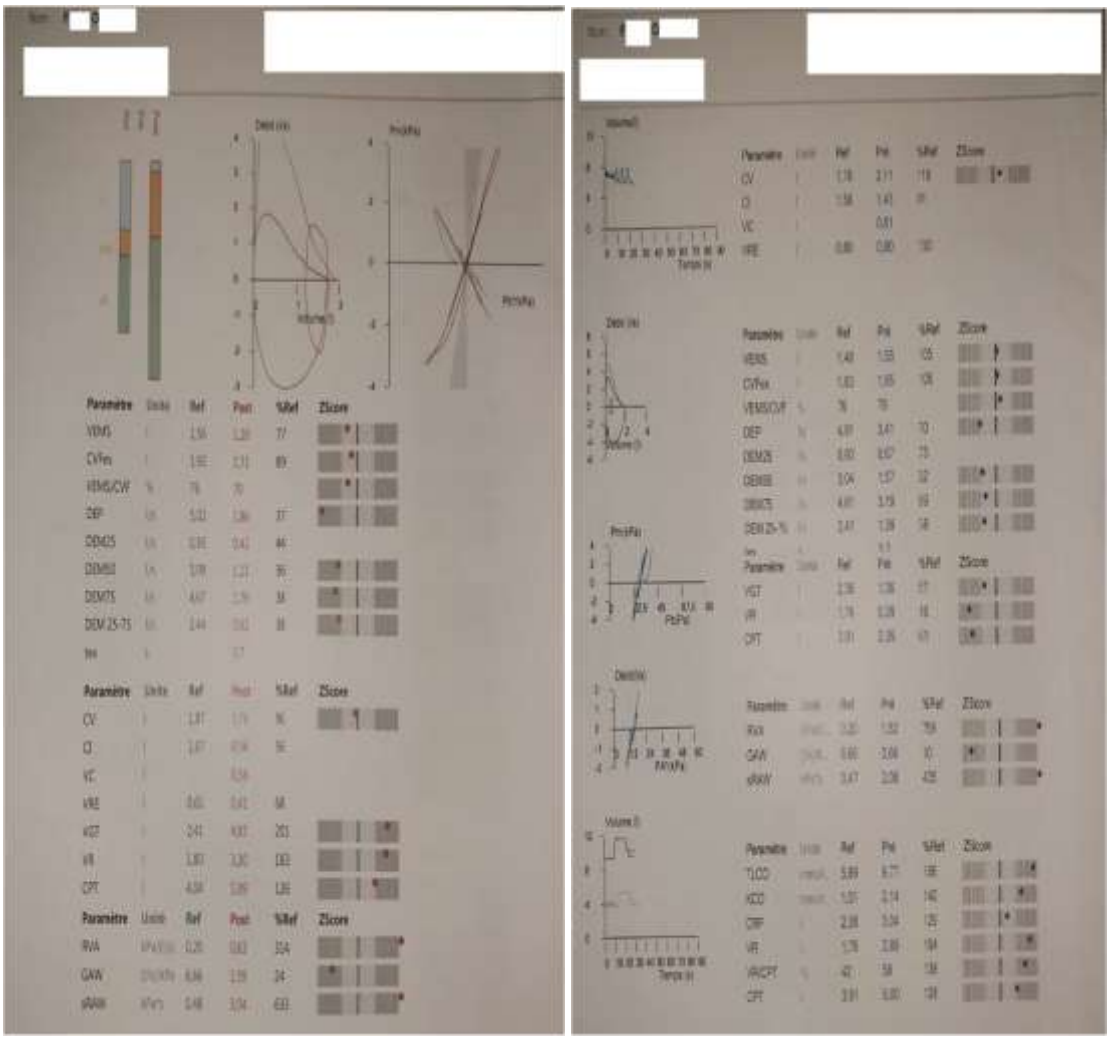


Figure 3: Chest CT in parenchymal window (expiratory slices): Mild lesions of air trapping with a few bilateral micronodules



Before treatment **After treatment**

Figure 4: The patient's functional evolution on oral corticosteroids

III. Discussion

Bronchiolitis is an inflammatory, fibrosing or destructive process affecting only (or predominantly) the bronchioles, with a variety of causes and clinical consequences [7]. Bronchiolitis occurring during systemic diseases is bronchiolitis of undetermined cause in a well-characterized context [2]: cellular bronchiolitis (including follicular bronchiolitis), and constrictive bronchiolitis (also known as "obliterative" bronchiolitis) [8], Bronchiolar involvement can be observed in all connectivitis, but it is during rheumatoid arthritis (RA) and Gougerot-Sjögren syndrome (GSS) that bronchiolitis is most frequently observed [6].

Clinical manifestations associated with bronchiolitis are not very specific, and may point to false asthma with a variable combination of cough, sputum, exertional dyspnea and chest wheezing [9], as in our patient's case.

Chest X-rays are often "normal" in bronchiolitis, except in cases where there is intense cellular bronchiolar infiltration that may give rise to a miliary or infiltrative pneumonitis appearance [7]. They may show airway distension or diffuse micronodulation [6].

High-resolution chest CT with acquisition during inspiration and expirationmay, be normal or shows direct signs of bronchiolitis, such as centrilobular nodules and bud-tree opacities, whereas indirect signs of bronchiolitis include areas of attenuation on inspiratory slices (mosaic perfusion appearance) and gas trapping on expiratory slices. These abnormalities are secondary to obstruction of the bronchial lumen, particularly in constrictive bronchiolitis [10].

The functional impact of cellular/follicular bronchiolitis is less severe than that of constrictive bronchiolitis [2]. Respiratory function tests may be normal or show obstructive, restrictive or mixed ventilatory disorders. It is generally estimated that an obstruction of more than 75% of the bronchioles (objectified by a decrease in DEM50 and 25-75) is necessary before an obstructive ventilatory disorder appears on the EFR. Partial reversibility after inhalation of bronchodilators may occur, but is usually absent [6-11], Gougerot-Sjögren's syndrome (GSS) is characterized by abnormal function and destruction of exocrine glands secondary to infiltration by T and B lymphocytes. Respiratory manifestations are frequent, potentially affecting the entire respiratory system, particularly the airways [6]. Bronchiolar involvement may be isolated or associated with non-specific interstitial lung disease (NSIP), or lymphocytic interstitial lung disease (LIP), such as follicular bronchiolitis. Signs of GSS include dry cough, bronchial hyperreactivity and local dry syndrome. Lymphoid infiltration bronchiolitis may be localized, manifesting as atelectasis that may mimic middle lobe syndrome without proximal bronchial obstruction. SGS is manifested by a 25-75% fall in FEV1 and sometimes in DLCO. Chest CT shows signs of distal airway involvement (bronchial wall thickening, centrilobular nodules and expiratory trapping) [2].

The treatment and prognosis of airway involvement in GSS are not defined. Some extraglandular manifestations (lymphoid proliferations) respond to corticosteroids [12]. The indication for general corticosteroid therapy should be discussed in symptomatic and progressive forms. In severe forms of bronchiolitis, a combination of corticosteroids and immunosuppressants has been proposed, but the results do not support the efficacy of this option [13]. When these treatments (bolus corticosteroids, high-dose systemic corticosteroids and/or immunosuppressants) are proposed for severe bronchial or bronchiolar disease, it is essential to assess their efficacy on the basis of complete CT and respiratory function parameters, including assessment of lung distension, to avoid subjecting the patient to the adverse effects of an ineffective treatment [2].

IV. Conclusion

Bronchiolar involvement is common in certain connectivitis, notably GSS, and should be systematically investigated in the context of atypical or difficult asthma as a differential diagnosis, starting with an interview to look for extra-respiratory signs, a thoracic CT scan with fine inspiratory and expiratory sections, immunological and histological work-up, and Respiratory function tests.

Bibliography

[1]. Perez, T., Deroubaix, C., Derollez, M., Wallaert, B., &Tonnel, A. B. (1989). Faux Asthmes De L'adulte. Revue Française D'allergologie Et D'immunologie Clinique, 29(1), 19-27.

[2]. Tillie-Leblond, I., Crestani, B., Perez, T., &Nunes, H. (2012). Voies Aériennes D Distales Et Maladies De Système. Revue Des Maladies Respiratoires. Https://Doi.Org/10.1016/J.Rmr.2012.10.020

[3]. Munira Alisherovnakhusainova, Sanjarnizamitdinovichkhaydarov, Khanuzadavranovnamakhmudova, &Abrorshokirovichnayimov (2023). PrevalenceOf Bronchiolitis In Patients WithRheumatoidArthritis. Science And Education, 4 (5), 232-241.

[4]. Chung, A., Wilgus, M. L., Fishbein, G., &Lynch III, J. P. (2019, April). Pulmonary And Bronchiolar Involvement In Sjogren'sSyndrome. In Seminars In Respiratory And Critical Care Medicine (Vol. 40, No. 02, Pp. 235-254). Thieme Medical Publishers.

[5]. Ravaglia, C., &Poletti, V. (2020, April). Bronchiolitis And Bronchiolar Disorders. In Seminars In Respiratory And Critical Care Medicine (Vol. 41, No. 02, Pp. 311-332). Thieme Medical Publishers.

[6]. Maurac, A., Borie, R., Debray, M., Cazes, A., &Crestani, B. (2018). Atteintes Bronchiolaires De La Polyarthrite Rhumatoïde Et Du Syndrome De Gougerot-Sjögren. Revue Du Rhumatisme Monographies. Https://Doi.Org/10.1016/J.Monrhu.2018.04.002

[7]. Cordier, J. (2005). Bronchiolites. Emc- Pneumologie, 2(4), 204-218. Https://Doi.Org/10.1016/J.Emcpn.2005.08.001

[8]. Fournier, M., Dauriat, G., Thabut, G., Colombat, M., &Groussard, O. (2008). Bronchiolites De L'adulte. Encyclopédie Médico-Chirurgicale. Https://Doi.Org/10.1016/S1634-6939(08)45277-8

[9]. Devouassoux, G., Cottin, V., Lioté, H., Marchand, E., Frachon, I., Schuller, A., F, B., &Cordier, J. (2009). CharacterisationOf Severe Obliterative Bronchiolitis In Rheumatoid Arthritis. The European Respiratory Journal, 33(5), 1053-1061.

[10]. Https://Doi.Org/10.1183/09031936.00091608

[11]. Devakonda, A., Raoof, S., Sung, A., Travis, W. D., &Naidich, D. P. (2010). Bronchiolar Disorders. Chest, 137(4), 938-951. Https://Doi.Org/10.1378/Chest.09-0800

[12]. Edwards, C. A., Cayton, R., &Bryan, R. T. (1992). Chronic Transmural Bronchiolitis : A Non-Specific Lesion Of Small Airways. Journal Of Clinical Pathology, 45(11), 993-998. Https://Doi.Org/10.1136/Jcp.45.11.993

[13]. Chen HA, Lai SL, Kwang WK, Liu JC, Chen CH, Huang DF. Middle Lobe Syndrome As The Pulmonary Manifestation Of Primary Sjögren'sSyndrome. Med J Aust. 2006 Mar 20;184(6):294-5. Doi: 10.5694/J.1326-5377.2006.Tb00242.X. PMID: 16548837.

[14]. Ito I, Nagai S, Kitaichi M, Et Al. Pulmonary Manifestations Of Primary Gougerot-Sjögren's Syndrome. A Clinical Radiologic, And Pathologic Study. Am J RespirCrit Care Med 2005;171: 632—8