SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FOR TALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

SPONTANEOUS PNEUMOMEDIASTINUM IN AMYLOPATHIC DERMATOMYOSITIS - A RARE MANIFESTATION

LAÍS DE VICO (UNESP, BOTUCATU, SP, Brasil), JOÃO PAULO CANAAN (UNESP, BOTUCATU, SP, Brasil), SEAN HIDEO SHIRATA LANÇAS (UNESP, BOTUCATU, SP, Brasil), MATHEUS ZANATA BRUFATTO (UNESP, BOTUCATU, SP, Brasil), JOELY CRISTINA DA SILVA TOLEDO (UNESP, BOTUCATU, SP, Brasil), LÍVIA CÂMARA OLIVEIRA (UNESP, BOTUCATU, SP, Brasil), SULA GLAUCIA LAGE DRUMOND PACHECO (UNESP, BOTUCATU, SP, Brasil), ANDREA DE ALMEIDA PEDUTI BATISTA (UNESP, BOTUCATU, SP, Brasil), LUIZ EDUARDO VALENTE (UNESP, BOTUCATU, SP, Brasil), FÁBIO VICENTE LEITE (UNESP, BOTUCATU, SP, Brasil), HENRIQUE PEREIRA SAMPAIO (UNESP, BOTUCATU, SP, Brasil)

BACKGROUND

Amyopathic dermatomyositis (AMD) is characterized by dermatomyositis (Gottron papules and heliotrope) cutaneous lesions in the absence of myositis. It accounts for 20% of cases of dermatomyositis. Interstitial lung disease may occur in 30%. Pneumomediastinum, pneumothorax, and subcutaneous emphysema are rare complications with few cases described in the literature. Hamman's syndrome was discribed a case of AMD with complicated ILD by spontaneous pneumomediastinum.

CASE REPORT

MB, male, 41 years old, previously health, rural worker, initiated a polyarticular inflammatory arthralgia associated with erythematous lesions on metacarpophalangeal, bilateral proximal and distal interphalangeal (gottron papules), elbow extensor erythema (gottron sign) and erythematous-violaceous upper bilateral (heliotrope) lesions associated with loss gain amount 30 kg in 6 months, in addition to dyspnea on moderate exertion. Absence of objective muscle weakness, normal muscle enzymes, electromyography without alterations and skin biopsy compatible with dermatomyositis, characterizing AMD. Negative paraneoplastic screening was performed. Autoimmune profile with ANA 1/80 nuclear fine dotted, Anti- Ro, La, Rheumatoid Factor, Anti citrulline antibodies, serologies for hepatitis B and C, HIV and Syphilis are negative. Chest tomography with interstitial infiltration suggestive of pulmonary fibrosis on discrete bases. We adoptted conservative treatment with prednisone 1mg / kg / day and azathioprine 2mg / kg / day. After 2 months, the patient was admitted to the ER with progressive worsening of dyspnea, severe retrosternal pain, dry cough, hoarseness, dysphagia, and a recent and progressive increase in the volume of the cervical region.

Eupneic, Haemodinamic stable Conscious and oriented. Pulmonar auscult with crepitations on 2/3 lower of the bilateral thorax. Emphysema subctunaeous was notted ate cervical region. New Tomography confirmmed: emphysema subcutaneous, Pneumomediastinum and progression of lung infiltrated. The hypothesis of ILD secondary to AMD and Hamman Syndrome has been explored. Evaluated by toracic surgery without indications of an invasive approach. He was pulsed with methylprednisolone 1g for 3 days, followed by cyclophosphamide 0.6mg / m2 / month.

After 30 days of hospital discharge, he was admitted to other service for sudden chest pain, evolving to death.

CONCLUSION

Spontaneous pneumomediastinum is an infrequent manifestation in systemic autoimmune myopathies and can be predisposed by ILD or previous corticosteroid therapy. We considered it important to report

this case because of the rarity of the finding and to highlight it as a differential diagnosis of sudden dyspnea in these group of patients requiring rapid recognition, since it may be fatal if left untreated.