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A Review of Pneumomediastinum in Dermatomyositis

Kerry Jobling [1*], Martin Lee [1], Jonathan Guckian [1].

Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK.

[*]**Corresponding Author**: Kerry Jobling, Newcastle upon Tyne Hospitals NHS Foundation Trust, Royal Victoria Infirmary, Queen Victoria Road, Newcastle upon Tyne, NE1 4LP, UK. **Email:** Kerry.jobling@nuth.nhs.uk Tel: 01912820070 .

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Abstract

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy of autoimmune aetiology, characterized by numerous systemic features including weakness, a characteristic heliotropic rash and Gottron's papules on the hands. DM may rarely be complicated by pneumomediastinum (PnM), the presence of free air within the mediastinum. PnM occurring in DM has been hypothesized to be due to distortion of lung architecture or vasculitic lesions causing alveolar necrosis. This is a significant complication of DM, with a relatively high mortality. Complications of this phenomenon include pneumothorax and surgicalemphysema.There is some debate regarding medical management of PnM in DM. Whilst the majority of reports in the literature describe oral and IV corticosteroids as being a mainstay of management, the use of numerous immunosuppressant therapies has been described, including cyclophosphamide, azathioprine, methotrexate, cyclosporine, Rituximab and IVIG. In view of the high mortality associated with this complication, more research is required to provide clinicians with evidence-based management recommendations. This review presents the current understanding based on existing evidence.

Keywords

Pneumomediastinum; Dermatomyositis; Interstitial lung disease.

Introduction

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy of autoimmune aetiology. It causes inflammation of skeletal muscle and other organs. Like most autoimmune disease, a combination of genetic and environmental factors likely predispose an individual to the development of DM. 10-20% of adults with DM have anti-Mi2 auto-antibodies [1]. Other inflammatory myopathies exist including polymyositis and inclusion body myositis.

DM is characterized by: symmetrical weakness of limb-girdle muscles/anterior neck flexors, myositis evident on muscle biopsy, elevated serum muscle enzymes (creatine kinase/CK), typical features on electromyography (EMG) and associated rash (a characteristic heliotropic rash and Gottron's papules on the hands). These features contribute to the diagnostic criteria of DM; Bohan and Peter criteria [1].

There are bimodal peaks of incidence in DM with onset occurring between years 5-15 and 45-65. The current incidence of DM is estimated to be 1/100,000 population/year [2]. There are multiple systemic complications of DM. Pharyngeal weakness may lead to difficulty swallowing and dysphonia. Dysphagia may also be complicated by aspiration pneumonia. There is a well-established association with malignancy (15-25%) [3].

Up to 10-43% of patients with DM develop interstitial lung disease (ILD) [4] of varying severity. Lung complications are categorized into intrinsic (ILD) or extrinsic disease (aspiration pneumonia, thoracic muscle weakness and pulmonary arterial hypertension). There are also several case reports of DM complicated by pneumomediastinum (PnM) [4-15]. This article summarises what is currently known of the incidence, complications and management of PnM occurring in patients with DM.

PnM is the presence of free air within the mediastinum. Many cases are caused through trauma e.g. a knife wound to the chest. Non-traumatic cases are considered 'spontaneous' and may occur in the presence of other underlying factors which cause rupture of para-mediastinal alveoli; such as illicit drug use, profuse vomiting/cough or barotrauma (e.g. diving) [4]. Symptoms of PnM include chest pain, facial swelling and dyspnoea. PnM is considered a rare complication of DM; incidence less than 2.2% (the quoted figure for the incidence of PnM in all inflammatory myositis) [5]. Some authors call for a new understanding of its true prevalence due to the increasing number of case reports suggesting that the rate of occurrence in DM is underestimated [6]. Furthermore, PnM may occur prior to the diagnosis of DM and as not all cases of PnM are symptomatic, clinicians should be alert to its development, especially in view of the significant mortality associated with it (see below).

Risk Factors

PnM occurring in DM has been hypothesized to be due to distortion of lung architecture or vasculitic lesions causing alveolar necrosis [4, 7]. Based on reporting of several cases in the literature, risk factors that are suggested for the development of PnM in DM are:

- Male gender [4]
- Younger age [8]
- Within early stages of DM [9,10]
- Underlying ILD [4]
- Corticosteroids (debated) [4,8,9]
- Presence of cutaneous ulcers [10-12]
- Chinese origin (due to high frequency of anti-MDA5 antibodies) [11]
- Normal/mildly elevated CK [8,10,11]
- Anti-Jo-1 negativity [13]

Complications

PnM is a significant complication of DM, with a relatively high mortality. Complications of this phenomenon include pneumothorax and surgical emphysema. The poorest survival appears to be associated with absence of muscle weakness, decrease in muscle capacity or carbon monoxide diffusion capacity before PnM onset [4]. Whilst most case reports in the literature discuss patients experiencing resolution of PnM, Le Goff et al describe a Kaplan Meier survival rate of 64% at one year and 55% at two years [5].

Management

Whilst pneumomediastinum alone is usually managed conservatively, PnM in the context of DM is associated with a poor outcome and therefore is often managed more aggressively. Oxygen therapy [4,8,10] is an initial step, however there is some debate regarding medical management. Whilst there is an argument that weakening of the alveolar wall, leading to subsequent air leak, can be caused by steroids [14], the majority of reports in the literature describe oral and IV corticosteroids as being a mainstay of management of PnM in DM. There is evidence of higher mortality when high dose corticosteroids are used alone [10]. The use of numerous immunosuppressant therapies has been described, including cyclophosphamide, azathioprine, methotrexate [11] cyclosporine [15], Rituximab [13] and IVIG. There has been little to no significant evidence of the efficacy of immunosuppressants [5] nevertheless their use remains prevalent in these cases.

Discussion

PnM is an uncommon complication of DM, most often occurring in the presence of ILD. The aetiopathophysiology of PnM in DM remains unclear. Case reports have provided us with an indication of possible risk factors. More work is needed to understand the true prevalence of PnM as a complication of DM. Current treatment approaches in the literature have been variable and present ambiguous conclusions, although immunosuppression appears to be the preferred choice of management in most reported cases. In view of the high mortality associated with this complication, more research is required to provide clinicians with evidence-based management recommendations.

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