CASE REPORT

Mycoplasma pneumoniae - The Culprit for Juvenile Dermatomyositis in A 9-Year-Old Girl: A Case Report

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ABSTRACT

Mycoplasma pneumoniae is frequently known as an "atypical bacterium" that can cause wide-ranging extrapulmonary manifestations. Here, we outline a case of a child, aged 9-year-old, who presented with profound proximal muscle weaknesses and a vague rash, associated with markedly elevated serum creatinine kinase (CK). Muscle biopsy suggested Juvenile Dermatomyositis (JDM) following an upper respiratory illness of *M. pneumoniae* origin. The child responded exceptionally well to a combined therapy of immunoglobulin, intravenous glucocorticoid and methotrexate.

Keywords: Rash, Myopathy, Juvenile dermatomyositis, Mycoplasma

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INTRODUCTION

JDM though "is the most common inflammatory myopathy of childhood" (1 p.595), "however its pathogenesis remains unknown" (1 p.596). While genetic and environmental factors were both known to play significant role, infective cause also has been commonly linked to. Various infectious microorganisms including common bacterial, viruses as well as parasites have been associated with JDM; nonetheless the link with *M. pneumoniae* infection remains uncommon.

It is well known that *M. pneumoniae* commonly caused mild respiratory illnesses in children. But extrapulmonary manifestations of this bacterial infection are relatively scarce, may be severe and may affect any organ system. We came across a case of gradual weaknesses and loss of function in a child after a minor respiratory infection that could result in a long-term impediment.

CASE REPORT

A girl, aged 9, presented to our paediatric clinic with a two months history of generalised symmetrical muscle ache and weaknesses. There was preceded history of upper respiratory tract illness three weeks prior to the onset of symptoms which resolved without medical attention. She gradually had difficulty to climb up stairs at home and have been absent from school due to enormous pain and fatigue while walking and climbing stairs at school. At presentation, she appeared fatigue, but all her vital signs were stable and normal. Physical examination revealed a child with normal Glasgow Coma Scale and behaviour. She had conspicuous generalised swelling of all four limbs with fading facial and lower arms rashes.

Figure 1 showed the vague rash on her face. Her neurological examination revealed reduced power with

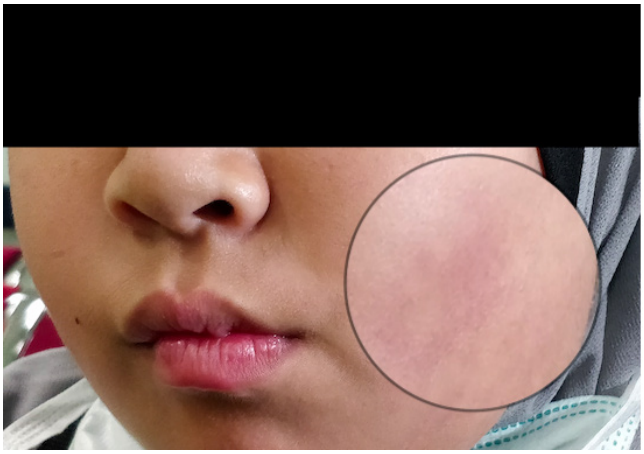

Figure 1: The picture of the patient's left cheek that shows a very vague rash appears intermittently few weeks after an episode of mild respiratory infection. It is an irregular erythematous non itchy patch that can be easily missed. The patient and parents were not aware of the rash until it has been pointed out by clinician.

grade 3/5 in bilateral lower limbs and grade 4 in both upper limbs. She had a marked proximal myopathy as she was not able to stand up from sitting on a chair. Her laboratory investigations revealed serum ESR of 84mm/hr with serial serum CK levels highest at 6470 U/L, and deranged liver transaminases. Her immunoglobulin (Ig) M serology for *M. pneumoniae* turned out positive. Other investigations including TORCH (Toxoplasmosis, Rubella, Cytomegalovirus and Herpes Simplex) screen, hepatitis serology, respiratory viruses screen and antinuclear antibody were all negative.

She was initially treated with oral Azithromycin and fluid hydration with appropriate analgesia. Immunosuppressive agent was not started yet at that point. However, she showed further clinical deterioration on daily routine function; her serum CK remained elevated and her proximal myopathy did not improve at all. A whole-body MRI was performed and showed increase signal enhancement in muscle groups of both upper and lower limbs. Figure 2 showed sagittal view of paravertebral muscular in the lumbosacral regions on T2-weighted images. Muscle biopsy was taken from left vastus lateralis muscle which showed muscle fibres atrophy especially around the perifascicular region leading to a diagnosis of JDM. Figure 3 showed the microscopic appearance her muscle biopsy. Treatment was started with intravenous immunoglobulin, followed by high dose methylprednisolone and methotrexate therapy. She showed good progression and improvement following the treatment, in addition to regular limbs physiotherapy. Currently, she is on small dose oral prednisolone, weekly oral methotrexate, multivitamins and oral calcium supplementation.

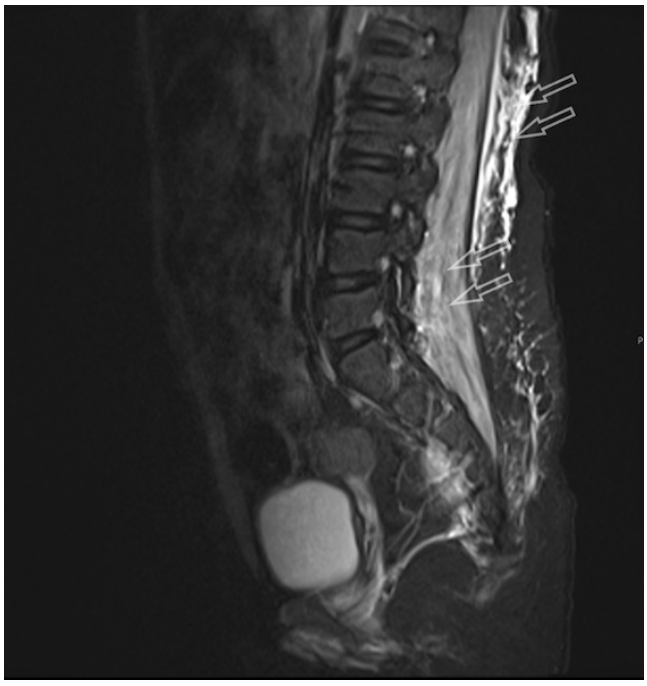

Figure 2: The image of sagittal view of lumbosacral region magnetic resonance imaging showed increased signal intensity on subcutaneous layer (top arrows) and paravertebral muscles (bottom arrows).

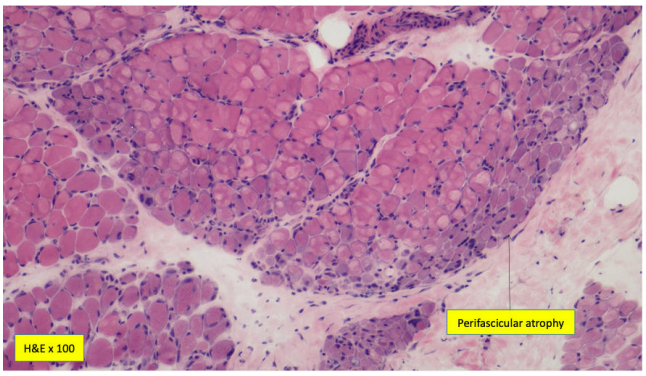

Figure 3: The image of haematoxylin and eosin (H&E) staining (x100) of the patient muscle biopsy showing perimysial and perivascular inflammation and perifascicular atrophy

DISCUSSION

JDM remains the most common inflammatory myopathies of childhood although the exact aetiology of the disease is still not completely well understood. Apart from environmental and genetic factors, several hypotheses using infection-triggered autoimmunity have been suggested which include anti-idiotypic antibodies induction, molecular mimicry and modification of self-antigens through microbial proteins. "Environmental and genetic factors affect the child's susceptibility to these conditions which also alter the inflammatory response, adding heterogeneity to disease pathophysiology" (2).

In our case, the patient sought treatment only after two months after seeking medical attention elsewhere, with symptoms suggesting JDM; symptoms are gradual in onset and progressing over time. This interval is not unusual as shown in a retrospective study in Nationwide Children's Hospital in Ohio over 23 years period which showed "the mean intervals between onset of symptoms to diagnosis in younger and older age groups were 5.6 months and 4.5 months respectively" (3 p. 4). The population under three year of age is defined "younger age" and above three year of age at the onset of symptoms is defined "older age".

Upon admission, blood investigations and imaging performed suggested the diagnosis of JDM, fulfilling some of the criteria according to modified Bohan and Peter which are; classic skin rash with symmetrical muscle weakness and elevation of muscle enzyme. The abnormal finding at muscle biopsy and MRI findings of generalised myositis further confirmed our suspicion of inflammatory myopathy. The positive results of *M. pneumoniae* antibody suggested the likely causative agent in this case, although at the time of presentation, the child had no more symptoms such as cold or cough.

In a study done in Toronto, it is reported that "thirty of 78 patients (38%) were found to have a history of clinical symptoms, not typically associated with JDM, that were suggestive of a probable infection" (4 p. 527). Among

them, respiratory infection was most common accounted for at least 80% of probable infection (4 p. 527). There are other infectious agents that have been associated with inflammatory myositis such as parvovirus, Borrelia burgdorferi, various viruses including influenza and hepatitis B, streptococcal infections, herpes simplex and Toxoplasma gondii (1 p. 596). In this case, the serologic confirmation for other infectious agents were not done to due cost implications and in the absence of typical clinical signs.

M. pneumoniae is a common bacterium known to affect paediatric age group. Cascades of autoimmune reactions are assumed to be the anchor that resulted in these extrapulmonary complications which may present before, during, after, or in the absence of pulmonary signs. It can be explained by three possible mechanisms the first being the direct effect in which *M. pneumoniae* directly blights certain organ and cause local cytokines induction and second; via an indirect route with formation of immune complexes formation and the third is vascular occlusion causing complications such as vasculitis and thrombosis (5 p. 162).

Thus, the above-mentioned pathogenesis can to a certain extent, result in inflammatory myopathy. In our case, it is analytical that apart from polymyositis, *M. pneumoniae* infection can be complicated with JDM. There are still very limited case studies and reports regarding JDM associated with *M. pneumoniae* compare to *M. pneumoniae* associated polymyositis, hence the incidence of JDM associated with *M. pneumoniae* is relatively unknown. More exposure is needed among clinicians to avoid late diagnosis and associated complications and poor outcomes.

CONCLUSION

JDM can be seen as an extrapulmonary complication even in a mild respiratory illness caused by *M. pneumoniae* and a high index of suspicion is required in all children presented with proximal myopathy and rash following this presumed benign infection. We presented this case with a degree of complexity as the child was initially treated as post *M. pneumoniae* complication and the investigations were stepped up in view of clinical deterioration. This case highlighted that proximal myopathy in an event of recent infection should be considered as "Idiopathic Inflammatory Myopathy", hence warranted such investigations.

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