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# JUVENILE DERMATOMYOSITIS AND HEPATITIS B VIRAL INFECTION

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## ABSTRACT

Juvenile dermatomyositis (JDM) is an uncommon autoimmune illness with unexplained origin. It affects the skin, skeletal muscles, and blood vessels.

This case report emphasizes an unusual case of an 8-year-old girl from Ninevah in Iraq, who presented with limb weakness and rash. She had a family history of hepatocellular carcinoma. The patient had hall mark features of JDM: heliotrope rash, Gottron's papules, and progressive proximal muscle weakness as well as elevated muscle enzymes. A positive hepatitis B surface antigen with high viral load proved concomitant infection. Treatment started with entecavir and intravenous immunoglobulins, followed by methotrexate and corticosteroids leading to steady clinical improvement. Effective antiviral treatment in combination with immunosuppressants is crucial for treating autoimmune diseases without infection reactivation.

The correlation between hepatitis B infection and JDM is still obscure. This case suggests that that hepatitis B may lead to autoimmune diseases when there is a genetic susceptibility. This example adds to the scarce literature on JDM connection with viral hepatitis, highlighting the importance of being alert and using specialized treatment approaches in such cases.

## KEYWORDS

Juvenile dermatomyositis, hepatitis B, Gottron's papules, heliotrope rash

## LEARNING POINTS

- *Rare case insight:* highlights the difficulties in diagnosing autoimmune and viral disorders by presenting a rare case of concurrent juvenile dermatomyositis and chronic hepatitis B infection.
- *Lessons in clinical management:* emphasizes the significance of controlling the risk of viral reactivation and screening for hepatitis B prior to immunosuppressive treatment.
- *Multidisciplinary and educational value:* highlights the importance of a multidisciplinary approach and provides internists with useful knowledge on how to provide individualized care in complicated rheumatic situations.

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## INTRODUCTION

Juvenile dermatomyositis (JDM) is an uncommon autoimmune illness, with incidence ranges between 20 and 40 per 10 million children[1]. The exact cause of JDM is unknown. The disease affects the skin, skeletal muscles, and blood vessels. Patients develop inflammation induced myopathy. Striated muscles are infiltrated by mononuclear cells with microangiopathy. A trigger may be required for a flare of JDM. These triggers may be infectious agents, environmental factors or certain medications[2]. JDM can be triggered by viruses, notably parvovirus B19, Epstein-Barr virus, coxsackie A and B virus and human immunodeficiency virus, bacteria, including *Mycoplasma pneumoniae* and group A *Streptococcus* or protozoa such as *Toxoplasma gondii*[3]. JDM or myositis are rarely reported as complications of viral hepatitis infection. Between 1960 and 2003 only 6 cases with different presentations of myositis related to viral hepatitis infections were reported in the literature as well as 2 cases related to hepatitis vaccinations[4]. To the best of our knowledge, this is the first reported case of JDM in a child with chronic hepatitis B infection; shedding light on a possible association between JDM and hepatitis B infection.

## CASE DESCRIPTION

The patient was an 8-year-old Iraqi girl, living in a rural area, Nineveh governorate, who presented to a private rheumatology clinic in Mosul city with limb weakness and rash for 1 month duration. It started in July 2024 as erythematous rash on the face, cheeks and periorbital areas especially on exposure to sunlight. Two weeks later the rash involved the dorsum of the hands, extensor surface of the elbows and knees. There was also swelling in her face mainly around the eyes. At the same time, progressive symmetrical limb weakness developed involving both upper and lower limbs, which manifested as difficult climbing stairs, getting up from a seat and hair combing. The patient denied having difficulty in swallowing, nasal speech or nasal regurgitation. Daily variation in muscle weakness, ocular involvement, Raynaud's phenomenon or mouth dryness were not reported. The patient complained of pain involving small and large joints of all her limbs. She had mild intermittent fever (not documented) with no rigor or weight loss. Review of other systems, and her past medical and surgical history was unremarkable. There was no family history of a similar condition; however, her grandfather and uncle had been diagnosed with hepatocellular carcinoma. She had many consultations and been misdiagnosed and treated for various conditions with no improvement before coming to our clinic. On physical examination, the patient was conscious, alert, afebrile, looking ill but not distressed, with marked periorbital puffiness with erythematous discoloration of the eyelids (heliotrope sign) *(Fig. 1A)*, and bilateral nonpitting oedema of lower limbs. Her weight, height and BMI were 21 kg, 120 cm and 14.6, respectively. On local examination of the skin, she had erythematous hyperpigmented patches located on both knees and elbows (Gottron's sign) *(Fig. 2A)*.

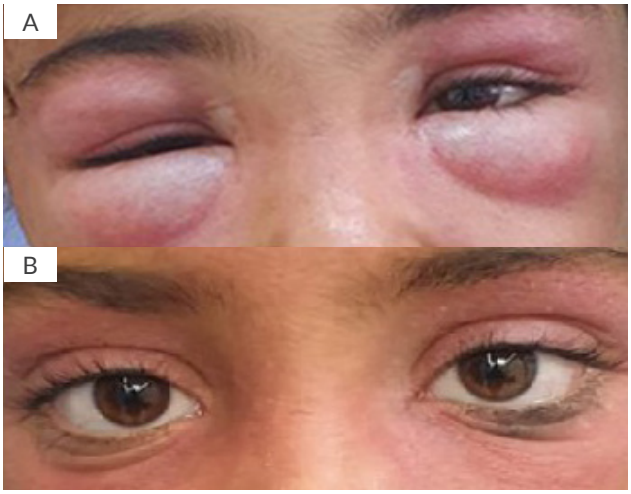



*Figure 1. A) Periorbital puffiness and heliotrope rash at presentation; B) Marked improvement of periorbital puffiness and heliotrope rash 4 months after treatment.*

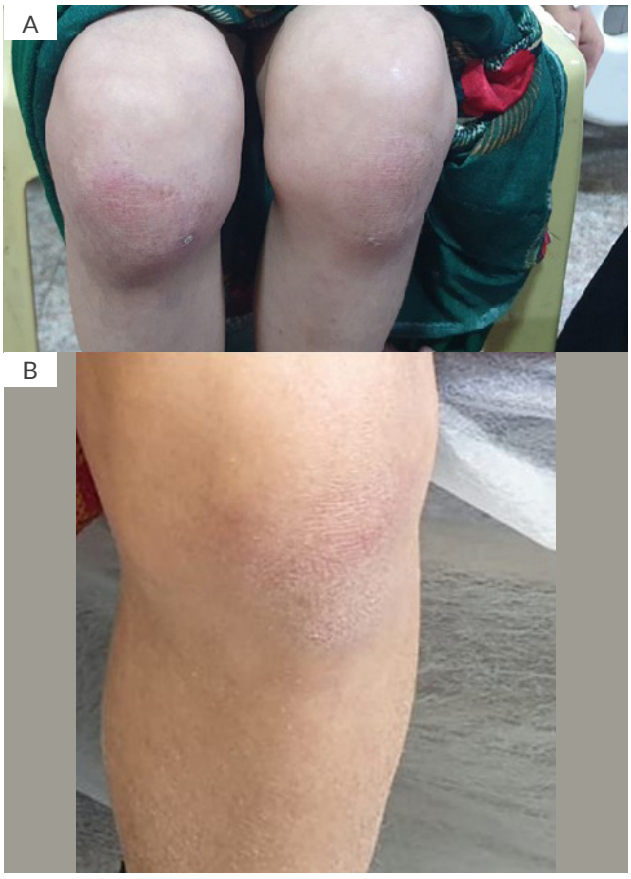



*Figure 2. A) Gottron's sign on the knees at presentation; B) Marked improvement of Gottron's sign on the right knee 4 months after treatment.*

There were also hyperpigmented erythematous firm papules on her metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints (Gottron papules) *(Fig. 3A)*. She had a waddling gait with limited extension and flexion of the knees. However, there is no joint swelling, warmth, or spine deformities were noted. Her muscle bulk was normal, but she had minimal tenderness at the proximal muscles of upper and lower limbs. Muscle power in the proximal muscles of both upper and lower limbs was 2 upper and 2 lower limbs, with a




*Figure 3. A) Gottron's papules on the knuckles of the hands at presentation; B) Marked improvement of Gottron's papules on the hands' knuckles 4 months after treatment.*

positive Gowers' sign. She had no calcinosis cutis. At the time of presentation, the patient was assessed for blood counts, biochemical parameters, serological markers in addition to hepatitis B viral study *(Tables 1 and 2)*.

The patient was initially treated with entacavir syrup 0.35 mg orally once daily and intravenous immunoglobulin (IVIg) 2 g/kg (given over 2 days). One month later, there was mild improvement in her skin rash and, to lesser extent, of her muscle weakness (2+ upper limbs, 2 lower limbs). Creatine phosphokinase (CPK) was 220 µg/l, and lactate dehydrogenase (LDH) was 341 U/l. She was started on prednisolone tab 30 mg daily (in 2 divided doses) and methotrexate tab 7.5 mg weekly with folic acid tab 5 mg weekly. In addition, she continued to receive IVIg (2 gm/kg) monthly. One month later, there was more improvement in her skin rash and muscle power (4 in upper limbs, 3+ in lower limbs). CPK was 85 U/l, LDH 286 U/l. Prednisolone was tapered gradually and consolidated into one daily dose and methotrexate dose was increased to 10 mg weekly and one month later into 12.5 mg weekly. Four months later muscle power was normal with marked improvement in the clinical picture *(Fig.1B, 2B and 3B)*. CPK 80 U/l, LDH 125 U/l.

## DISCUSSION

JDM is a rare autoimmune disease for which there seems to be a genetic predisposition. Infection is one of the most important triggering factors for JDM. Hepatitis B may act as triggering factor as in our patient. A number of studies indicate that there is a link between viral infections and inflammatory myopathies[1].

Autoimmune humoral immune response was initiated and attacks endothelium of the endomysial capillary leading to perimysial inflammation. In addition, cytotoxic T cells attack muscle fibres causing polymyositis[5]. Vaccination with the hepatitis B vaccine has been found in 2007 to be a triggering factor for JDM in a 6-year-old age boy correlated to the immune response against vaccine Autoimmune mechanisms related to hepatitis B virus (HBV) infections have been linked to some of the extrahepatic clinical features in acute or chronic disease such as the prodromal pre-icteric flu-like illness, vasculitis, primarily polyarteritis nodosa , and HBV-related glomerulonephritis[6].

Clinically JDM has dermatological and muscular features. There is weakness of the proximal muscles of shoulders and the flexor groups of both neck and hip. The affected muscles are inflamed, painful and tender. Progression of muscle weakness may involve oral, nasopharyngeal and oesophageal muscles causing difficulty in eating, swallowing as well as speech abnormalities. Joints may be involved and develop arthritis. Skin manifestations are diagnostic for JDM as the symmetrical violet colour heliotrope rash in addition the gleaming Gorton's papules, facial erythema,

| **Blood counts** | |
|---|---|
| Haemoglobin (g/dl) | 11.6 |
| White blood cell ($\times 10^9$/l) | 9.1 |
| Platelets ($\times 10^3$/µl) | 344 |
| Erythrocyte sedimentation rate (mm/hr) | 45 |
| **Biochemical parameters** | |
| Creatine phosphokinase (U/l) | 460 |
| Serum glutamic-oxaloacetic transaminase (U/l) | 30 |
| Creatinine (µmol/l) | 45 |
| Lactate dehydrogenase (U/l) | 425 |
| Serum glutamic pyruvic transaminase (U/l) | 20 |
| **Serology** | |
| Antinuclear antibody | Negative |
| C-reactive protein | Negative |

*Table 1. Laboratory investigations.*

| **Viral study** | **Results** |
|---|---|
| Hepatitis B surface antigen | Positive |
| Viral load ($\times 10^6$ copies/ml) | 345 |
| Hepatitis B core antigen, IgM | Negative |
| Hepatitis B core antigen, IgG | Positive |

*Table 2. Viral study.*

nails beds erythema and photosensitivity. Vasculitis affects tiny blood vessels of the fingers, toes and arms with ulcerations and calcium deposited in soft tissues due to calcinosis, which occurs in about one third of the patients. Death may occur due to respiratory problems as interstitial lung disease or respiratory muscle weakness. Another rare fatal complication is vasculitis of the intestinal tract[7,8]

Enzymes revealing muscle diseases as CPK, LDH, serum glutamic-oxaloacetic transaminase (SGOT), and serum glutamic pyruvic transaminase (SGPT) are elevated in JDM. A study by Shenavandeh and colleagues found that CPK was high in around 63% of patients with different categories of inflammatory myositis and ranges between, while LDH was elevated in around 92% of cases. SGOT was high in about 65% of cases. On the other hand, SGPT was high in only 51% of cases. Erythrocyte sedimentation rate (ESR) was found to be more than 25mm in the first hour in all patients at presentation[9].

In this case the patient had two problems, an autoimmune disease, JDM, and infection with hepatitis B. The challenge was treating the autoimmune disease and at the same time preventing exacerbation of the viral infection due to use of immunosuppressants. Entecavir was our drug of choice to control viral hepatitis, which is a first line treatment for HBV, acting against viral deoxyribonucleic acid with a low rate of resistance[10]. Timing of starting antiviral therapy is important before giving patient immunosuppressant drugs. In this case the antiviral entecavir treatment started 4 weeks before starting immunosuppressive treatment[10]. This regimen will be beneficial to limit activity of viral infection when using steroids in medium to high doses for mor than one month to avoid activation of the virus. Moreover, methotrexate is regarded one of the cytotoxic drugs with low risk of reactivation of HBV[11]. Immunoglobulins were given initially with the antiviral therapy for short course treatment of JDM in a dose of 2 g/kg for 2 days, before giving immunosuppressants and continue with 2 gm/kg monthly. Although short courses of immunoglobulins were found to provide control of symptoms for only a short time, they were found to be effective treatment in JDM even if there are complications as dysphagia, with lower fatality rate than treatment with corticosteroids or immunosuppressants[8].

In conclusion, this case report sheds light on the potential relation between viral infections, especially HBV, and the development of JDM in genetically susceptible subjects. Although the specific relationship is still unknown, the case highlights how significant it is to pinpoint viral infections as a potential cause of autoimmune diseases. It also underscores the need for a careful balance between antiviral therapy and immunosuppressive therapy in order to achieve correct control of autoimmune diseases without reactivation of infections. By underlining the importance of a watchful and individualized treatment approach in similar patients, this work adds to the scant evidence on the relationship between JDM and viral hepatitis.

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