Dermatomyositis Complicated by Calcinosis: Keys to the Therapeutic Challenge

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Abstract

Background: There is no curative therapy for dermatomyositis complicated by calcinosis. Many patients with such disorders will continue to have some symptoms. In Iraq many specialist physicians may have limited experience with treatment of such disorders and treatment side effects of long term treatment can be sometimes encountered. It is hoped that advances associated with the accumulating research evidence can help in improving treatment of such conditions.

Materials and Methods: A unique case of pediatric dermatomyositis associated with calcinosis that was referred because treatment was unsatisfactory and was associated with serious side effects was studied. The medical literatures were reviewed for the research evidence suggesting possible therapeutic interventions than improve this condition.

Results: The occurrence of pediatric dermatomyositis associated with calcinosis has not been well documented in Iraq. RH is an eight-year old who had dermatomyositis associated with calcinosis and complicated by bilateral osteonecrosis of the hip joints, pericardial effusion, and chronic hematoma at the right ankle. The girl was treated before referral by some of the traditional therapies that improved her symptoms early in the course of illness, but treatment was associated remissions and relapses and she experienced serious side effects of long-term treatment.

Conclusion: Research evidence suggests several possible therapies for the treatment of dermatomyositis complicated calcinosis despite long-term prednisolone and methotrexate. The choice of the option may have to be individualized as it may depend on the experience of the treating physician, the availability of the medications. As a first line we suggest the use of Adrenocorticotropic hormone (ACTH) combined with probenecid and aluminium hydroxide as a first line.

Keywords: Dermatomyositis; Calcinosis; Osteonecrosis of Hips; Pericardial Effusion

Introduction

Dermatomyositis is a chronic connective tissue disorder characterized by inflammation of the skin and muscle with a highly variable course, and characteristic signs including violaceous "heliotrope" eyelid discoloration with periorbital edema. The diagnosis of this condition is based mostly on clinical features. There is no curative therapy for dermatomyositis complicated by calcinosis. Many patients with such disorders will continue to have some symptoms [1-5].

In Iraq, many specialist physicians may have limited experience with treatment of such disorders and treatment side effects of long term treatment can be sometimes encountered. It is hoped that advances associated with the accumulating research evidence can help in improving treatment of such conditions.

Materials and Methods

A unique case of pediatric dermatomyositis associated with calcinosis that was referred because treatment was unsatisfactory and was associated with serious side effects was studied. The medical literatures were reviewed for the research evidence suggesting possible therapeutic interventions than improve this condition.
Results

The occurrence of pediatric dermatomyositis associated with calcinosis has not been well documented in Iraq. RH is an eight-year old who had dermatomyositis associated with calcinosis and complicated by bilateral vascular osteonecrosis of the hip joints, pericardial effusion, and chronic hematoma at the right ankle. The girl was treated in the province of Maysan in the South of Iraq before referral by some of the traditional therapies that improved her symptoms early in the course of illness, but treatment was associated remissions and relapses and she experienced serious side effects of long-term treatment.

The illness of R.H started at the about the age of four years two weeks after an episode of diarrheal illness. She developed progressive weight loss and weakness with difficulty in walking in association with characteristic violaceous “heliotrope” eyelid discoloration in the upper lid with periorbital edema, and she gradually lost the ability to walk occurring in association of proximal muscle wasting. She was treated with long-term prednisolone and treatment was associated remissions and relapses, and methotrexate was used.

During the eight year, she had radiologic evidence of osteoporosis and tissue calcifications (Figure 1A) developed bilateral osteonecrosis of the hip joints (Figure 1B), pericardial effusion, and chronic hematoma at the right ankle. Pericardial effusion was performed and smears showed a large number of fresh and old degenerated red blood cells and very few scattered inflammatory cells suggesting hemorrhagic pericardial effusion. Pleural fluid sugar was 247 mg/dL. MRI of the right ankle showed a well-defined multi-locular septated soft tissue lesion with no bone involvement suggesting chronic hematoma. She was also experiencing thrombocytosis and platelet counts ranged between 509 - 707 x 10⁹/L.

Figure 1A: Radiographs of the hands showing osteoporotic changes and tissue calcifications.
She also gradually developed multiple masses (calcinosis) in several areas of her body and some of them were infected and showing purulent discharge (Figure 2). She had high Erythrocyte Sedimentation Rate (ESR) at the level of 80 mm/hr, suggesting poor control of the inflammatory process, and mildly reduced blood calcium (8.78 mg/dL, normal 9 - 11 mg/dL). She was also anemic (Hemoglobin 9 g/dL).

Figure 2: On referral, the girl had multiple masses (calcinosis) in several areas of her body and some of them were infected and showing purulent discharge.
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Literature review revealed that several therapeutic interventions have been used with a beneficial effect in this condition including ACTH [6], aluminium hydroxide [7,8], excision calcified masses [9], probenecid [10-12], Colchicine [13], oral diltiazem with or without oral pamidronate [14-16], alendronate [17], intravenous infliximab [18], Pamidronate [19-22], intralesional corticosteroids [23,24], intravenous immunoglobulin [25], Sodium thiosulfate [25].

Discussion

Bitnum., et al. (1964) reviewed 1000 cases of dermatomyositis reported during the twentieth century in the literature and found 240 pediatric cases below the age of sixteen years [2].

The occurrence of calcinosis in pediatric dermatomyositis has been recognized as early as 1938 by Bailey [1], while the occurrence of dermatomyositis complicated by calcinosis during early infancy has been recognized as early as 1970 by Dupuis and Sironi [4].

Hill and Wood (1970) emphasized the occurrence of calcinosis as a complication of dermatomyositis in children, and also emphasized the value of steroid therapy and the possibility of the occurrence of some serious complications. Hill and Wood reported that methotrexate was also used in this condition and it was found promising by some authors, but its use was also reported to be without benefit in two cases [5].

In 1981, Skuterud, Sydnes and, Haavik reported that the use of probenecid in the treatment of a 9-year-old girl with dermatomyositis and calcinosis was associated with a dramatic decrease in subcutaneous and intermuscular calcinosis [10]. Harel., et al. (2001) reported of probenecid in the treatment of a 9-year-old girl with dermatomyositis and calcinosis. The calcifications resolved over 18 months of treatment. Harel., et al. attributed the effectiveness of probenecid to increasing renal phosphate clearance [11]. Nakamura., et al. (2006) reported an 11-year-old boy with dermatomyositis who developed calcinosis of both legs despite treatment with corticosteroid and cyclosporine. Probenecid treatment resulted in remarkable improvement of calcinosis, normalization of serum phosphorus level and after 17 months [12].

Fuchs., et al. (1986) reported that the use of oral colchicine in a dosage of 1 mg daily in a girl with dermatomyositis complicated by calcinosis was followed within two months by significant regression of local inflammation and healing of the skin ulcers [13].

Nakagawa and Takaiwa (1993) emphasized that the treatment of calcinosis complicating childhood dermatomyositis was commonly unsatisfactory and reported a patient who was successfully treated with oral aluminum hydroxide. Almost complete clearing of calcinosis was noticed after 8 months of therapy. Treatment was not associated with any adverse effect [8].

Oliveri., et al. (1996) reported an 8-year-old girl with dermatomyositis calcinosis who developed severe steroid induced osteoporosis. Successful treatment of myopathy with methylprednisolone and immunosuppressive drugs didn’t prevent the development of calcinosis. The girl was treated with oral diltiazem (5 mg/kg/day) to control calcinosis and oral pamidronate (4 mg/kg/day) in addition to calcium and vitamin D supplementation, which she had been taking for 3 years. Twenty-one months of treatment was associated with dramatic regression of the calcinosis [14]. Ichiki., et al. (2001) reported a 3-year-old girl with dermatomyositis at age developed progressive calcinosis despite treatment of dermatomyositis with prednisolone and the addition of aluminium hydroxide for calcinosis. Treatment with diltiazem completely suppressed the development of calcinosis in this girl [15].

Ambler., et al. (2005) reported a 6-year-old boy with improving dermatomyositis who developed severe and debilitating calcinosis, unresponsive to diltiazem and probenecid. However, the use of alendronate was associated with dramatic improvement within 1 month and by one calcinosis had virtually resolved [17].
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Riley, et al. (2008) treated five patients who had refractory dermatomyositis with calcinosis with intravenous infliximab. The initial dose was 3 mg/kg. Further doses were given at weeks 2, 6 and every 8 weeks thereafter. The dose and frequency were tailored according to the clinical response. Improvements were seen in all five patients [18].

Marco Puche, et al. (2010) reported three children with dermatomyositis and calcinosis treated with intravenous pamidronate 1 mg/kg/day on three consecutive days every three months. Calcinosis markedly reduced in the three patients, and completely cleared in patient one patient. Treatment was not associated with any adverse effect [19]. Martillotti, Moote and Zemel (2014) reported the use of intravenous pamidronate in the treatment of a 7-year-old girl with dermatomyositis and severe calcinosis refractory to several medication regimens. Three administrations resulted in marked improvement in calcinosis, pain and function and led to remission within less than 1 year of treatment [20].

Giri, et al. (2020) reported three pediatric patients who had dermatomyositis and calcinosis treated with pamidronate. Treatment was complete clearance of calcinosis in one child [22].

Pagnini, et al. (2014) reported the successful use of sodium thiosulfate in the treatment of a patient with pediatric dermatomyositis complicated by ulcerative skin disease and progressive calcinosis [25].

For this case, because of the evidence of persistent inflammatory process and serious steroid side effects, we recommended the use of ACTH combined with probenecid and aluminium hydroxide as a first line. As a second line we recommended pamidronate, and intravenous immunoglobulin.

Conclusion

Research evidence suggests several possible therapies for the treatment of dermatomyositis complicated calcinosis despite long-term prednisolone and methotrexate. The choice of the option may have to be individualized as it may depend on the experience of the treating physician, the availability of the medications. As a first line we suggest the use of ACTH combined with probenecid and aluminium hydroxide as a first line. The girl continued to have high ESR (80 mm/hr) suggesting poor control of the inflammatory process, and the need for steroids; the development of serious steroid side effects makes ACTH therapy the preferable option.

Bibliography


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