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DERMATOMYOSITIS WITH ANTI-NXP2 POSITIVE ANTIBODIES REVEALED BY RECURRENT CUTANEOUS CALCINOSIS IN AN 8-YEAR-OLD BOY: CASE REPORT AND LITERATURE REVIEW

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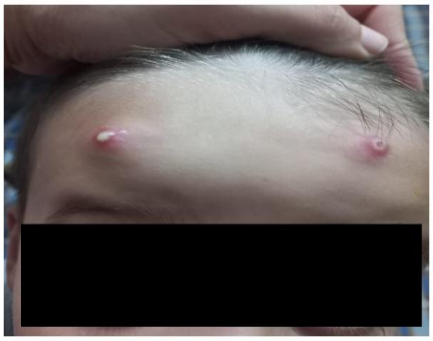
ABSTRACT
We report the case of an 8-year-old boy with no significant family history who presented with subcutaneous nodules and diffuse cutaneous calcinosis. His symptoms included non-inflammatory arthralgia, myalgia, muscle fatigue, and photosensitivity. Clinical examination revealed proximal muscle weakness and characteristic cutaneous lesions of dermatomyositis. The diagnosis was confirmed by the presence of anti-NXP2 positive antibodies and the absence of biological inflammatory syndrome. Oral corticosteroid therapy was initiated. This case highlights the importance of early diagnosis in dermatomyositis, especially in atypical pediatric forms associated with cutaneous calcifications.

KEYWORDS: Oral corticosteroid therapy was initiated.

INTRODUCTION
Dermatomyositis (DM) is a rare autoimmune inflammatory disease that primarily affects skeletal muscles and the skin. In its pediatric form, it can be associated with severe complications, such as cutaneous calcinosis, particularly in cases with anti-NXP2 antibodies. These antibodies are increasingly recognized as diagnostic markers for the disease in children. Calcinosis, although atypical as an early symptom, can be a key sign of dermatomyositis, as demonstrated by this clinical case.[1,2]

CASE REPORT
The patient is an 8-year-old boy with no significant family history. In his personal history, he had subcutaneous nodules that appeared on the lower third of the tibia, for which he underwent excision surgery in 2022. He also reported non-inflammatory arthralgia in the knees and ankles, myalgia, muscle fatigue, and photosensitivity.

On clinical examination, proximal muscle weakness (score 3/5) was noted. Dermatological examination revealed diffuse cutaneous calcinosis (on the forehead, ears, genital area, and tibial regions), as well as erythema in photo-exposed areas. Additionally, painful periungual erythema was observed, but no mucosal involvement was noted.



Biological Workup
Biological tests showed that muscle enzymes were normal, with no biological inflammatory syndrome. However, the presence of positive antinuclear antibodies and anti-NXP2 antibodies confirmed the diagnosis of anti-NXP2 positive dermatomyositis. This result is consistent with the clinical and immunological features associated with the pediatric form of the disease.[3,4]

Radiological Workup

A muscle ultrasound was performed, revealing calcinosis lesions without significant findings, and a skin biopsy was indicated to confirm muscle inflammation and calcinosis. However, it was deemed unnecessary in light of the biological and radiological results.

Treatment and Management

The treatment initiated consisted of systemic oral corticosteroids (prednisone), which resulted in gradual improvement of muscle and skin symptoms. Regular clinical and biological monitoring was established to evaluate the response to treatment and adjust the doses. Due to the recurrence of cutaneous calcinosis, additional therapeutic options, such as bisphosphonates, may be considered in the future.[5]

DISCUSSION

The diagnosis of pediatric dermatomyositis remains challenging, especially in atypical forms. In this case, the clinical signs of cutaneous calcinosis and photosensitivity, associated with strongly positive anti-NXP2 antibodies, allowed for the diagnosis to be established. Cutaneous calcinosis, although rarely present at the onset of the disease, is a concerning sign and can indicate a severe form, with the risk of progression to greater muscle and joint involvement. Studies have shown that early detection of anti-NXP2 antibodies can lead to better disease control and more effective management.[6,7]

Anti-NXP2 antibodies, found in this case, are particularly associated with pediatric forms of dermatomyositis, which are often more severe and resistant to treatment. Their presence, combined with cutaneous and muscular involvement, underscores the need for prompt treatment and close monitoring to prevent complications. Recent studies also suggest that stronger immunosuppressive treatments may be necessary to control severe forms of the disease.[8]

Pediatric dermatomyositis with anti-NXP2 antibodies is often a more severe and treatment-resistant form of the disease compared to classic forms. Recent studies show that anti-NXP2 antibodies are strongly associated with clinical manifestations of cutaneous calcinosis, and early detection of these antibodies allows for rapid treatment adaptation. Therapeutic options, including corticosteroids, immunosuppressants, and bisphosphonates for calcinosis, are continually evolving, and a multidisciplinary approach remains essential.[9,10]

Autoanticorps dans la myosite auto-immune

Auto-anticorps	Signes cliniques
	Dermatomyosite amyopathique, papules palmaires
Anti-TIF1g	Risque accru de cancer
Anti-NXP-2	Calcinose sous-cutanée Risque accru de cancer

[12]

	SAE	Mi2	NXP2	TIF1g	MDA5
Atteinte cutanée	Sévère Atteinte typique Erythème violacé inconstant	Erythème typique	Calcinose Oedème sous cutané	Sévère Papules de Gottron hyperkératosique Lésions psoriasiformes Patch 'red on white'	Mains de mécanicien Ulcère cutané Papule palmaire
Atteinte musculaire	Dysphagie	Atteinte sévère Taux élevé de CPK	Atteinte sévère Myalgie Dysphagie	Atteinte modérée Taux faible de CPK « DM amyopathique » Dysphagie	« DM amyopathique »
Atteintes extra-cutanéo-musculaires	PID: Peu fréquente Arthrite: Peu fréquente Raynaud: Peu fréquent	PID: Peu fréquente Arthrite: Peu fréquente Raynaud: Peu fréquente	PID: Peu fréquente Arthrite: Peu fréquente Raynaud: Peu fréquente	PID: Peu fréquente Arthrite: Peu fréquente Raynaud: Peu fréquente	PID sévère Arthrite: fréquente Raynaud: fréquent Fièvre
Association au cancer	0-20 %	0-20 %	10-35 %	35-80 %	0

[13,14]

CONCLUSION

This case underscores the importance of early recognition of clinical signs of dermatomyositis, particularly in atypical pediatric forms. Cutaneous calcinosis, although rare, can be an indicator of this condition and guide the diagnosis. Early management, especially with corticosteroids, is crucial for improving symptoms and preventing complications. Anti-NXP2 antibodies play a pivotal role in diagnosing severe forms of pediatric dermatomyositis.[11]

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