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Oral PUVA therapy in a four-year-old girl with an invalidating nodular cutaneous mastocytosis

Fototerapia oral con UVA en una niña de cuatro años con mastocitosis cutánea nodular invalidante

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Key words:

Mastocytosis, phototherapy, child, PUVA.

Palabras clave:

Mastocitosis, fototerapia, niño, fototerapia con UVA.

ABSTRACT

Mastocytosis comprises a heterogeneous group of disorders characterized by mast cell proliferation and accumulation within various organs, most commonly the skin. Treatment of cutaneous mastocytosis is aimed at reducing symptoms, avoiding agents that precipitate mediator release, use of antihistamines, glucocorticoids, sodium cromoglycate and ketotifen. Phototherapy with UVA or UVB has proven effective in controlling pruritus and cutaneous wheals, and reducing the number of mast cells in the affected skin, but to date there are few pediatric cases reported in the literature. We present a case of a four-year-old girl with a nodular cutaneous mastocytosis and systemic symptoms with minimal response to previous therapies. The patient tolerated well the treatment with PUVA, achieving an improvement in her systemic symptoms, skin lesions, and quality of life, with no immediate side effects. Although the experience reported in the literature is limited and the long-term effects are not known, we propose that PUVA could be a useful alternative in severe cases of cutaneous mastocytosis in children.

RESUMEN

La mastocitosis comprende un grupo heterogéneo de trastornos caracterizados por la proliferación y acumulación de mastocitos dentro de varios órganos, más comúnmente, la piel. El tratamiento de la mastocitosis cutánea tiene como objetivo reducir los síntomas, evitando agentes que precipitan la liberación del mediador, el uso de antihistamínicos, glucocorticoides, cromoglicato de sodio y cetotifeno. La fototerapia con UVA o UVB ha demostrado ser eficaz en el control del prurito y las ronchas cutáneas, así como en la reducción del número de mastocitos en la piel afectada, pero hasta la fecha hay pocos casos pediátricos reportados en la literatura. Presentamos un caso de una niña de cuatro años con mastocitosis cutánea nodular y síntomas sistémicos, con respuesta mínima a la terapia anterior. La paciente toleró bien el tratamiento con UVA y logró una mejora en los síntomas sistémicos, las lesiones cutáneas y su calidad de vida, sin efectos secundarios inmediatos. Aunque la experiencia reportada en la literatura es limitada y los efectos a largo plazo no son conocidos, proponemos que la fototerapia con UVA podría ser una alternativa útil en casos severos de mastocitosis cutánea en niños.

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INTRODUCTION

Mastocytosis comprises a heterogeneous group of disorders characterized by mast cell proliferation and accumulation within various organs, most commonly the skin. Activating mutations in c-kit have been related to the pathophysiology. Treatment of cutaneous mastocytosis is aimed at reducing symptoms, avoiding agents that precipitate mediator release, use of antihistamines, glucocorticoids, sodium cromoglycate and ketotifen.^{1,2} Phototherapy with UVA or UVB³ has proven effective in controlling pruritus and cutaneous wheals, and reducing the number of mast cells in the affected skin, but

to date there are few pediatric cases reported in the literature.

CASE REPORT

A four-year-old girl was diagnosed with severe nodular cutaneous mastocytosis from birth (Figure 1A). The lesional skin biopsy was positive for c-kit and tryptase, and negative for CD 25. Serum tryptase levels were elevated to 114 ng/mL (N < 11). She developed systemic symptoms such as flushing, hypotension and episodes of diarrhea, bleeding and bronchospasm. She was treated with high doses of H1 and H2 antihistamines, ketotifen, topical and systemic cromoglycate and oral

corticosteroids. The relief of the systemic symptoms was minimal, so twice-weekly oral psoralen plus UVA (PUVA) therapy were begun. 8-Methoxypsoralen (8-MOP) was given orally in a dose of 0.3 mg/kg, and 5 mg of dexchlorpheniramine half an hour before each treatment. Protective sunglasses were used in the cabin and 24 hours after the treatment.

The UVA were delivered starting at 0.50 J/cm², and increased by 0.25 J/cm² in subsequent visits, up to a maximum dose of 3 J/cm². A total dose of 157.1 J/cm² was

delivered over 24 weeks of treatment during the winter and spring seasons.

The patient's parents or medical staff remained with her inside the cabin for the first weeks. A baseline and a monthly measurement of c-DLQI were taken.

RESULTS

A progressive relief of symptoms was noted after twelve treatments with a dose of 3 J/cm². Three months later, she



Figure 1. A) A four-year-old girl with nodular cutaneous mastocytosis before PUVA therapy. B) After 24 weeks of treatment, a progressive relief of her systemic symptoms was noted, with a slight decrease in the erythema and infiltration of the plaques.

Table 1. Case reports of cutaneous mastocytosis in children treated with PUVA.

Patient	Gender	Clinical	Previous treatment					Age of beginning of photo-therapy	Total dose of UVA J/cm ²	Improvement				Systemic symptoms	Weeks of treatment	Side-effects	Reference
			Anti-H	TC	OC	KT	CR			Skin lesions	Pruritus	Dermoglyphism	Darier's sign				
1	F	UP						7 y	14		x	x	x		4		Christophers et al ⁴
2	M	DCM	x				x	7 m	51	x		x			20		
3	M	DCM						6 m	42.5	x		x			16		
4	F	DCM	x					3 m	70	x		x			16		Smith et al ⁵
5	F	DCM	x	x			x	3 y	41			x			12		Smith et al ⁵
6	F	DCM	x	x	x		x	4 m	99	x					16		Mackey et al ⁶
7	F	UP						1 y	22	x	x		x		NR		Godt et al ⁷
8	M	UP						7 y	4	x	x		x		NR		Godt et al ⁷
9	F	UP						12 y	44	x			x		NR		Godt et al ⁷
10	M	UP						15 y	13	x			x		NR		Godt et al ⁷
11	M	DCM	x		x		x	6 m	40	x				x	NR		Escrignano et al ⁸
12	M	DCM	x	x				16 m	24.2	x		x		x	23	Skin burn	Kinsler et al ⁹
13	F	DCM	x	x	x	x	x	4 y	129.8	x	x			x	20	No	Current case

NR = non registered, UP = urticaria pigmentosa, DCM = diffuse cutaneous mastocytosis, Anti-H = antihistamines, TC = topical corticosteroids, OC = oral corticosteroids, KT = ketotifen, CR = cromoglycate, y = years, m = months.

was left alone in the cabin. A reduction in the infiltration of the plaques was noted (Figure 1B), but the most important observation was a decrease in the frequency of systemic symptoms such as pruritus, diarrhea, episodes of hypotension and flushing. An improvement in her daily living activities was reported, and it persisted even in the summer season. No side effects were observed during the six-month follow-up period at the end of the treatment.

DISCUSSION

Pediatric mastocytosis often differs in presentation and prognosis from adult variants. Currently, there is no standard treatment and the main objective is to reduce the symptoms caused by the release of mast cell mediators.

Phototherapy is used in rare cases of massive cutaneous involvement and severe symptoms that do not respond to other treatments. Oral PUVA therapy has been reported to be effective in cases of diffuse cutaneous mastocytosis in children, although there is little published evidence in the literature and most of it corresponds to isolated case reports (Table 1). We proposed the PUVA therapy as a

treatment alternative in our patient to improve her systemic symptoms and quality of life.

CONCLUSIONS

We report a case of a four-year-old girl with a nodular cutaneous mastocytosis and systemic symptoms, with minimal response to previous therapies. The patient tolerated well the treatment with PUVA, achieving an improvement in her skin lesions, systemic symptoms and quality of life, with no immediate side effects.

Although the experience reported in the literature is limited and the long term effects are not known, we propose that PUVA could be a useful alternative in severe cases of cutaneous mastocytosis in children.

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