6 children had had positive mixed nut challenges which included walnut. We were unable to identify the causative allergen to these positive challenges so these children were not included in the analysis.
82.5% (33) had had both skin prick testing and specific walnut IgE. No patients had had component testing.
Of the 2 children who had had positive challenges, one had had both walnut IgE and skin prick testing (SPT 1 mm and IgE 2.15kua/l) and the other only skin prick testing (SPT 1 mm).
Of the 6 children who had had a positive combination challenge including walnut, walnut SPT ranged from zero to 4 mm, with 50% being zero to 2 mm, 66% (4) children had no specific walnut IgE obtained prior to challenge.
Conclusion: We had the majority of children undergoing walnut challenges having had both skin prick testing and specific IgE to walnut prior to challenge. Improvements could still be made, especially in relation to adding component walnut testing (rJugr1 and r Jugr3), in those children who have had previous reactions and whom had had inconclusive skin prick test, and walnut specific IgE results.

1525 | Clinical, genetic and therapeutical findings of two Brazilian patients with cutaneous mastocytosis associated with systemic autoinflammation

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Background: Autoinflammatory diseases (AID) comprise a broad spectrum of disorders characterized by unchecked activation of the innate immune system. The unremarkable progress in genomics led to an increasing number of disorders classified as autoinflammatory or containing an AID component. Cutaneous mastocytosis is a rare myeloproliferative disease, characterized by excessive proliferation and accumulation of mast cells limited to the skin without systemic inflammation. Cryopyrin/NLRP3-associated periodic syndromes (CAPS) is a group of rare spectral AID characterized by the constitutive activation of NLRP3 inflammasome and the consequent elevate production of inflammatory cytokine IL-1ß. Besides macrophages, monocytes, and other innate immune cells, mast cells (MCs) were shown to express functional inflammasomes too.
Method: Clinical review of patients data. Peripheral genomic DNA sequencing. Parallel peripheral monocytes isolation. IL-1B measurement in supernatants after stimulation with ATP, LPS and ATP-LPS.
Results: Clinical Findings: All patients presented clinical features compatible with both: 1- cutaneous mastocytosis; and 2 - CAPS (Table 1). Genetic Findings: Both patients were sequenced with the next generation sequencing (NGS) based autoinflammatory gene panel described in Rusminiet al., ARD 2015. P1 was negative for the panel whereas P2 was found to have pathogenic heterozygous mutation in NLRP12 (exon 3 - c.c910T; pH304y). Functional Findings: NLRP3 –IA in P1 peripheral monocytes cells revealed high level of IL-1ß when challenged with LPS, and ATP did not amplify cytokine production, suggesting a defect in NLRP3 inflammasome when compared to healthy controls (LPS 2196.5 pg/µL; LPS+ATP 2442.5 pg/µL; ratio LPS+ATP/LPS 1.11 pg/µL; media health control: LPS 80 pg/µL; LPS +ATP 234.05 pg/µL; ratio LPS+ATP/LPS 3.373 pg/µL). Therapeutical: Isolated or combined, anti-histaminic with cromoglycate was not effective in symptoms control. Corticosteroids achieved complete clinical control although side effects were observed. Steroids sparing agents, such as anti-IL1 was effective but divergent clinical response was observed between anakinra and canakinumab, as described in Table 1.
Conclusion: This is the first report of the clinical association of cutaneous mastocytosis and systemic autoinflammation. We propose that these findings are prototype of a novel autoinflammatory syndrome (AIMAS syndrome) as well as a key role for MC inflammasome inflammation.

	P1	P2
Age onset (<1 year)		
. AID symptoms	No (14 months)	Yes (1 month)
. Mastocytosis symptoms	Yes (3 months)	Yes (15 days)
Autoinflammation Symptoms		
Recurrent fever	Yes	Yes
Cold/stress induced triggered episodes	yes	yes
Sensorineural hearing loss	No	No
Urticarial rash	Yes	Yes
Musculoskeletal symptoms	Yes	Yes
Abdominal Pain	Yes	No
Chronic Aseptic meningitis	No	No
Skeletal abnormalities	No	No
Raised inflammatory markers	Yes	Yes
Mastocytosis Symptoms		
Cutaneous	Positive Darier's Test	Positive Darier's Test

	P1	P2
Gastrointestinal	Abdominal pain, oral ulceratin and diarrhea	Abdominal pain, oral ulceratin and diarrhea
Anaphylaxis	Insect and Ibuprofen	Insect
Serum Tryptase levels (<11)	21.9	<11
Skin biopsy	>30 mast cells	Not counted but compatible with mastocytosis
NGS Target Panel (IL1RN, LPIN2, MEFV, MVK, NLRP12, NOD2, PSTPIP1, PSMB8, NLRC4, NLRP3, NLRP12 TNFRSF1A)		
	No mutation	Pathogenic mutation on NLRP12; Exon 3; c.C910T;PH304Y
Treatment		
Anti-histaminics isolated	Partially controlled	Partially controlled
Cromoglycate	Partially controlled	Partially controlled
Corticosteroids	Completely controlled	Completely controlled
Response to anti-IL1		
. anakinra	Partially controlled	Partially controlled
. canakinumab	Completely controlled	Completely controlled