734: Frequency and clinical relevance of myositis-specific autoantibodies in children

S. Mejdoub[1], S. Mallouli[2], H. Hachicha[1], F. Kamoun[2], S. Feki[1], C. Triki[2], H. Masmoudi[1]

1 : Immunology Department, Habib Bourguiba University Hospital, Sfax, Tunisia

2 : Child Neurology Department, Hedi Chaker University Hospital, Sfax, Tunisia

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INTRODUCTION

- Screening for auto-antibodies (Abs) is useful for the management of **idiopathic inflammatory myopathies (IIMs)**.
- Among **myositis-specific Abs (MSA)**, some Abs are reported to be associated with juvenile variants.

OBJECTIVE

to determine the **frequency** and **clinical relevance** of **MSA in children**

METHODS

- Study period: 2013-2023

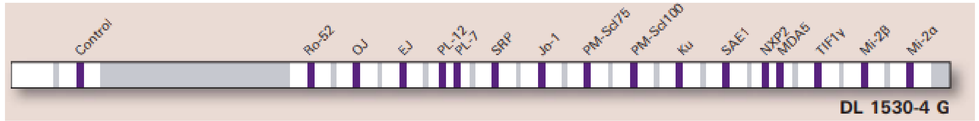
  → 392 serum samples tested for a suspicion of IIM

- MSA screening: immunodot technique (Dot-myositis, Euroimmun®)

  → auto-Abs directed against 16 antigens:

  Mi-2α, Mi-2β, TIF1γ, MDA5, NXP2, SAE1, Ku, PM-Scl100, PM-Scl75, Jo-1, SRP, PL-7, PL-12, EJ, OJ, Ro 52

  → semi-quantitative results for each Ab: negative / + / ++ / +++



- Inclusion of **pediatric cases with positive MSA**

  → collection of clinical data from medical records

RESULTS

129 positive tests for at least one MSA

→ **3 pediatric cases:** three **boys** who had in common **walking difficulties**

Disease-onset: insidious in the 1st case, subacute with a preceding infectious episode in the 2nd and 3rd cases

1st case: anti-NXP2 +++

- 4 years and 9 months
- clinical myopathic syndrome
- electromyographical abnormalities
- suggestive histological features

→**Established diagnosis of IIM**

2nd case: anti-TIF1γ++

- 14 years
- clinical and electromyographical features
- inflammatory biological syndrome
- biopsy not done

→**Probable diagnosis of IIM**

3rd case: anti-MDA5 ++

- 6 years
- ENMG: acute motor axonal neuropathy pattern
- Cerebrospinal fluid analysis: albumino-cytological dissociation

→**acute polyradiculoneuropathy**

→association with a muscular disorder remained to be confirmed

DISCUSSION AND CONCLUSION

❖**Juvenile IIM : extremely rare** (incidence: 2-4 per million)
Dermatomyositis: the most frequent IIM both in adults and children (4-14 years ++, female predominance) *[Marasandra Ramesh H et al, 2022]*

❖Each **MSA** → one type of IIM, particular phenotypes and prognosis → useful to support the existence of a myopathy, its acquired nature, to classify IIM, to definite its prognosis *[Allenbach Y et al, 2014]*

❖ **MSA+ : 50-70% of juvenile IIM**; **the most frequent: anti-TIF1γ, anti-NXP2, anti-MDA5** ≠ in adults: the most frequent MSA are anti-synthetase Abs *[Kim H et al, 2021]* ; no increased cancer risk for children *[Turnier J et al, 2022]*

➢ Our results are in accordance with literature data regarding the low seroprevalence of MSA in children and the antigenic specificities reported in this population.
≠discordance: the 3 reported cases are boys; absence of cutaneous signs

➢ The positivity of these Abs enables to evoke juvenile variants of IIM, which diagnosis relays on clinical, biological, electromyographical and histological arguments.