

Table. Patient's characteristics and outcomes

Patient	Age at JDM diagnosis (years) / Sex	Clinical involvement at onset of JDM / At initiation of JAK I (*)	Auto-Ab	Muscle biopsy scoring (total score, (0- 27) / vascular score, (0-3)/ connective tissue score (0-2) (Time elapsed from diagnosis to MB)	Previous treatments	Indication for JAKi/ (Time elapsed from JDM diagnosis to onset of JAKi)	JAKi Dose	Achievement of a CID (time from JAKi initiation to CID, months); Treatments associated with JAKi at time of CID	Outcomes/treatment at last FU in responders; (duration of follow-up from onset of JAKi)
1	13/F	Muscle* (CMAS 2/52, MMT 38/80), skin rash* (DAS skin 4/9), bowel*	Anti-NXP2	24/3/1 (first, 2 months) 5/0/1 (second, 44 months)	CS#*, IVIg (d-32§), MTX, RTX (d-107§), MMF, tacrolimus, IA*	Refractory severe muscle disease (22 months)	Ruxolitinib 0.7mg/kg/d (15 mg bid)	Yes (2.6); CS (0.3 mg/kg/d)	CID Progression of calcinosis Ruxolitinib (35 months)
2	8 /F	Muscle* (CMAS 5/52, MMT 48/80), skin rash* (DAS skin 8/9), arthralgia, Calcinosis*	Anti-NXP2	26/3/1 (2 months)	CS#*, IVIg*, MTX, HCQ*, RTX (d-50§), MMF, sirolimus, PEX*	Refractory severe muscle and skin disease (18 months)	Ruxolitinib 0.6mg/kg/d (10 mg bid)	Yes (5.3); CS (0.15 mg/kg);	CID Improvement of calcinosis Ruxolitinib (27 months)
3	10/F	Muscle* (CMAS 32/52, MMT 69/80), severe skin ulceration* (DAS skin 2/9), polyarthritis,	Anti-MDA5	5/1/0 (3 months)	CS#*, IVIg*, MMF, MTX, RTX (d+106§)*	Refractory severe skin ulcerations (4 months)	Ruxolitinib 0.7mg/kg/d (10 mg bid)	Yes (5) CS (0.2 mg/Kg/d)	CID Ruxolitinib CS (0.1 mg/kg/d) (15 months)
4	12 /F	Muscle* (CMAS 43/52, CMAS 74/80), severe skin ulcerations* (DAS skin 3/9) polyarthritis*,	Anti-MDA5	15/2/1 (2 months)	CS#*, IVIg*, PEX*, RTX (d-35)	New-onset JDM with severe skin ulcerations and polyarthritis (2 weeks)	Baricitinib 0.1 mg/kg/d (2 mg bid)	Yes (1.7) CS (0.4 mg/kg/d)	CID Baricitinib, CS (0.2 mg/kg/d) (8 months)
5	5/F	Muscle* (CMAS 7/52, MMT 26/80), skin rash* (DAS skin 6/9)	Anti-NXP2	NA	CS#*, MTX, IVIg*, HCQ, RTX (d-69§)	Refractory severe muscle disease (4 months)	Baricitinib 0.5 mg/kg/d (4 mg bid)	Yes (5); CS (0.4 mg/kg/d)	CID Baricitinib CS (0.3 mg/kg/d) (8 months)
6	7/F	Muscle* (CMAS 3/52, MMT 44/80), skin rash* (DAS skin 7/9) involvement	Anti-TIF1 $\gamma$	NA	CS#*, MTX, IVIg (d-7), PEX*	Refractory severe muscle disease (1 month)	Ruxolitinib - 0.7 mg/kg/d (7.5 mg bid)	No	-
7	12/M	Muscle* (CMAS 26/52, MMT 49/80), skin* (rash, DAS skin 8/9), bowel*	No MSA Anti-U1-RNP and anti-	23/3/2 (54 months)	CS#*, HCQ*, MTX, MMF, IVIg (d+453), RTX (d-294§), tacrolimus	Refractory muscle, skin and digestive disease (40 months)	Ruxolitinib 0.8 mg/kg/d (20 mg bid)	No	-

			SSA/ Ro- 60kD)						
8	11/F	Muscle* (CMAS NA MMT NA), skin rash* and ulcerations* (DAS skin 8/9)	Anti-NXP2	NA	CS#*, MTX, IA*, IVIg (d-35§)	Refractory severe muscle disease (2 months)	Ruxolitinib - 0,8 mg/kg/d (10 mg bid)	No	-
9	9.5 /M	Muscle* (CMAS 6/52 MMT 50/80), skin rash (DAS skin 6/8)	Anti-TIF1 $\gamma$	10/2/1 (1 month)	CS#*, MTX, IVIg (d-57)	Refractory severe muscle disease (10 months)	Baracitinib (4 mg bid)	No	-
10	3/M	Muscle* (CMAS 14/52; MMT 45/80), skin rash and ulcerations (DAS skin 8/9)*	Anti-TIF1 $\gamma$	24/3/2 (20 months)	CS#*, MTX; MMF, RTX (d-137), IA, IVIg*	Refractory severe muscle and skin disease (27 months)	Ruxolitinib (0.7 mg/Kg/d)	No	-

CID, clinically inactive disease; CS, corticosteroids; F, female, FU, follow-up; HCQ, hydroxychloroquine; IA, immunoadsorption; IVIG, intravenous immunoglobulins; JAKi: JAK inhibitors; JDM, juvenile dermatomyositis; M, male; MMF, mycophenolate mofetil; MTX, methotrexate; NA: not available; PEX, plasma exchange; RTX, rituximab.

\* Indicates involvement and treatments at onset of JAKi

# Steroid dosage  $\geq 0.5$ mg/kg/day

§ Delay between the last rituximab or IVIg infusion and JAKi introduction (in days)