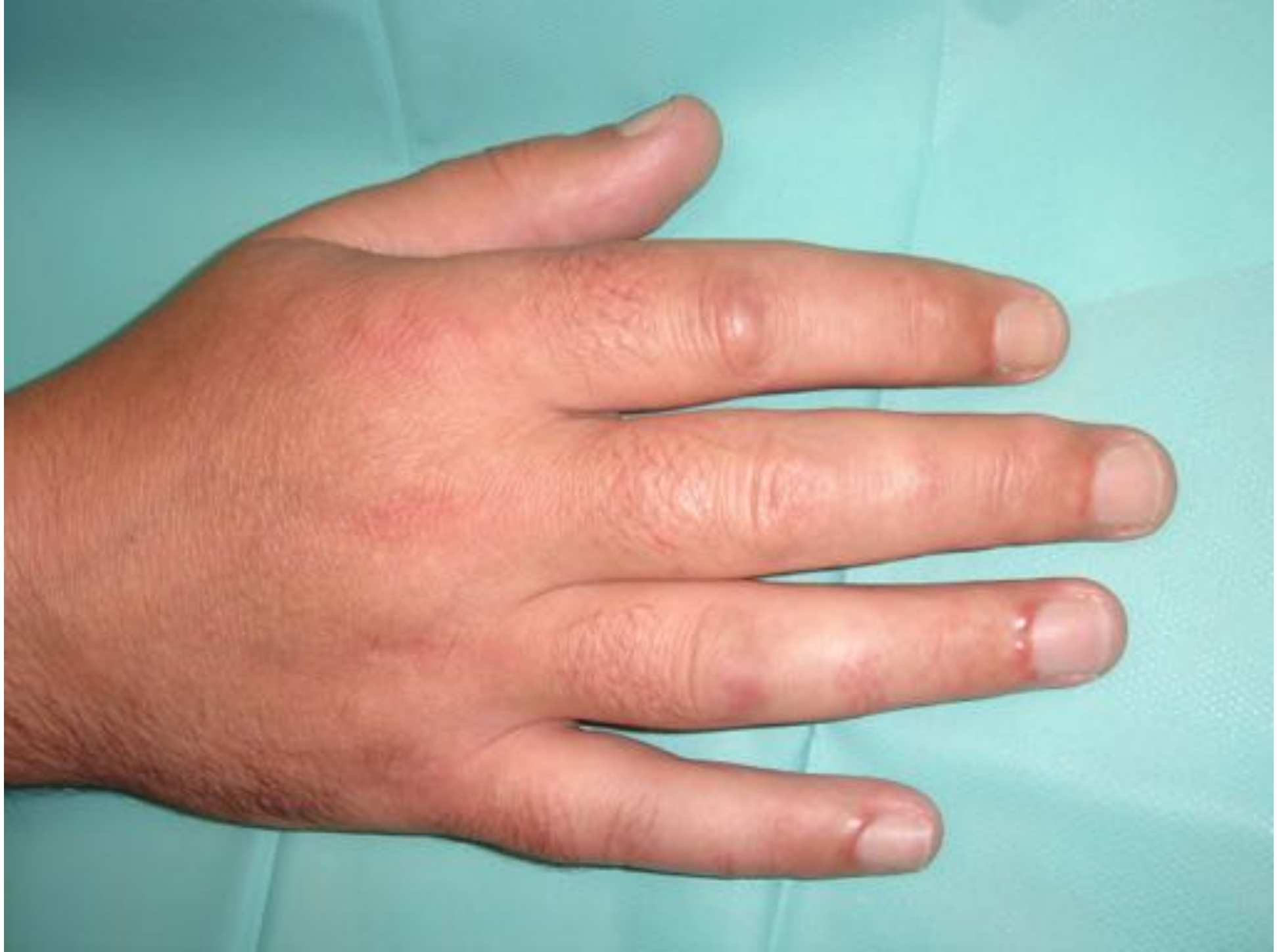
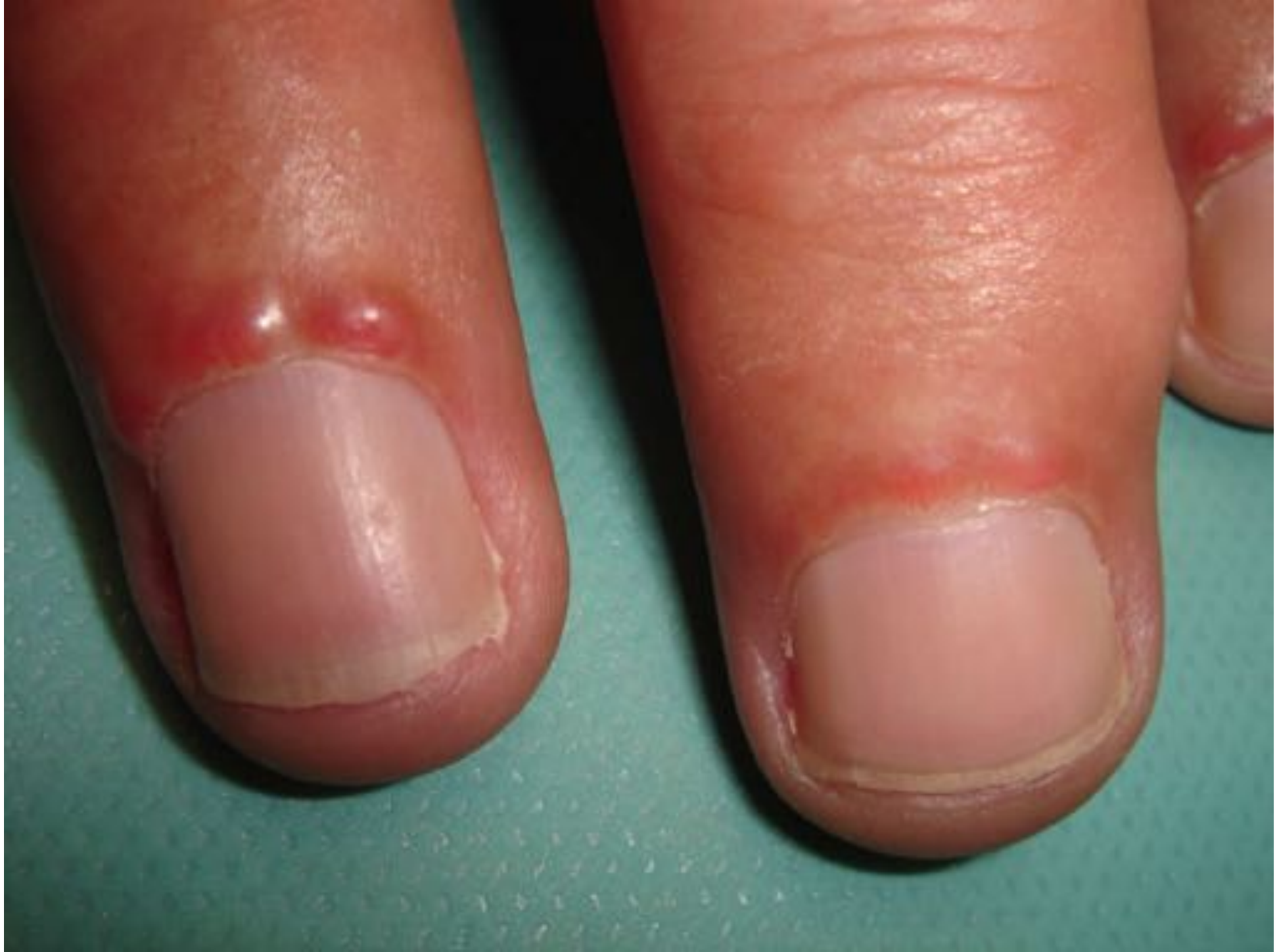
Cas clinique

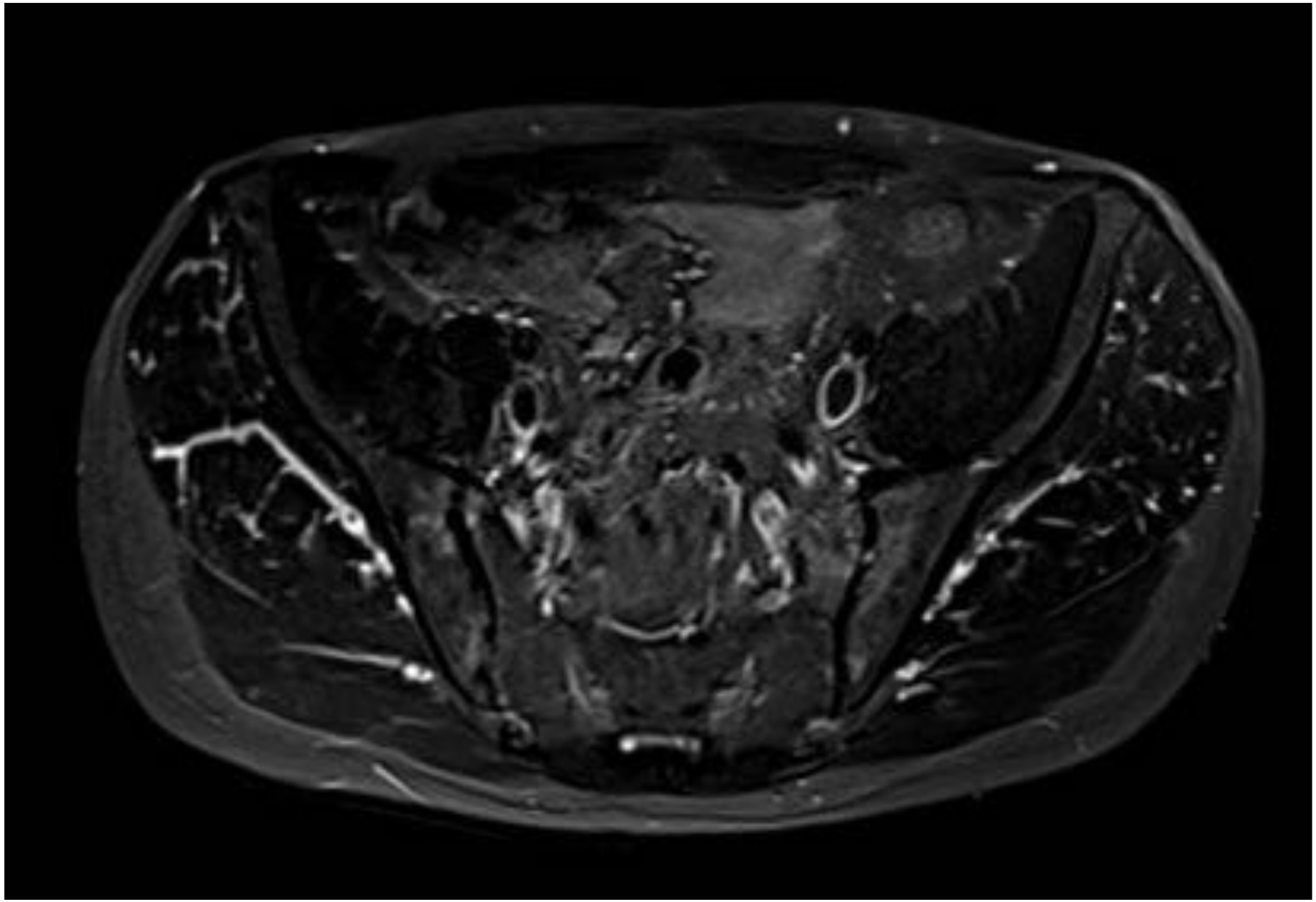
- ♂47 ans.
- Pas d’antécédent.
- Pas de traitement.
- PR chez une tante.
- Polyarthrite, rachialgies, Raynaud.
- Et des lésions cutanés !

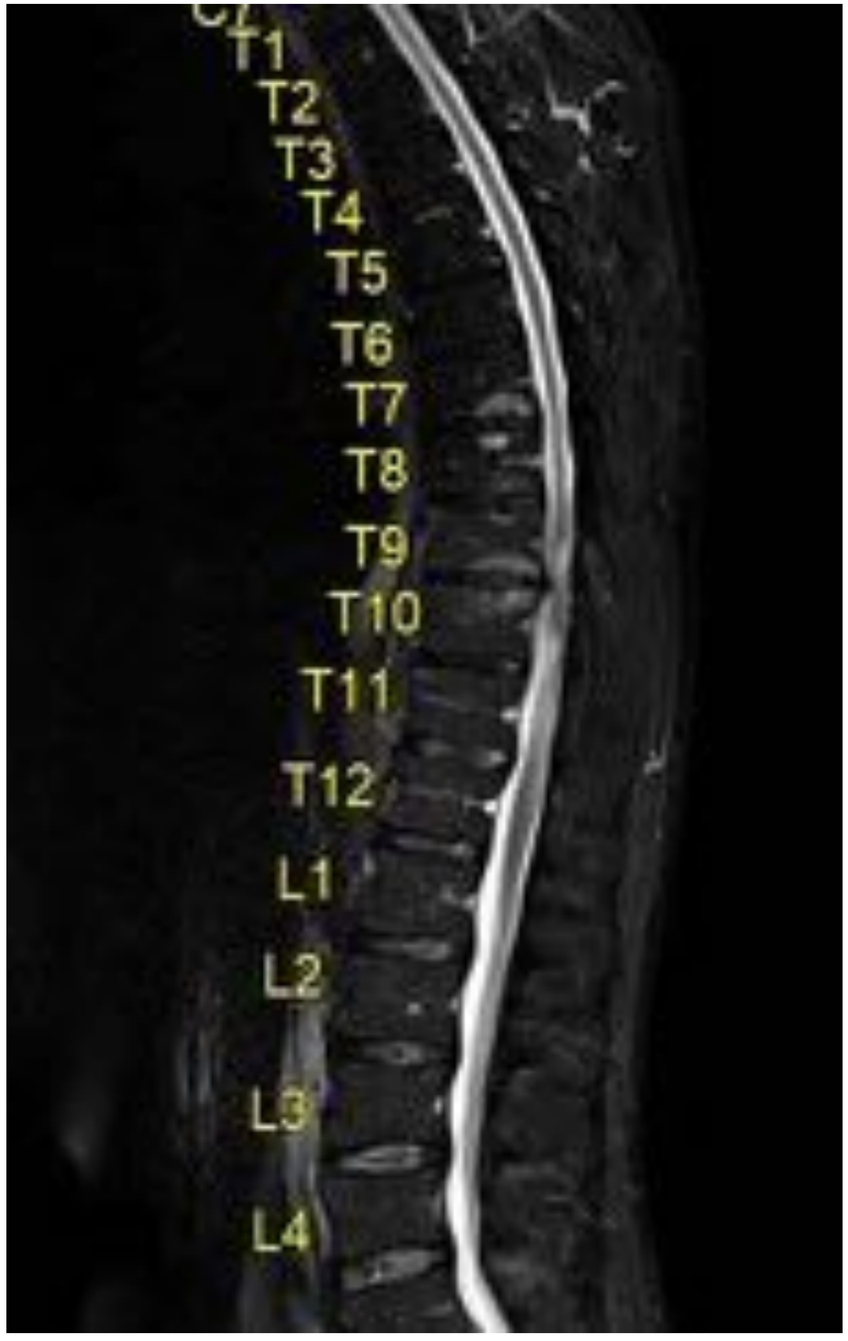


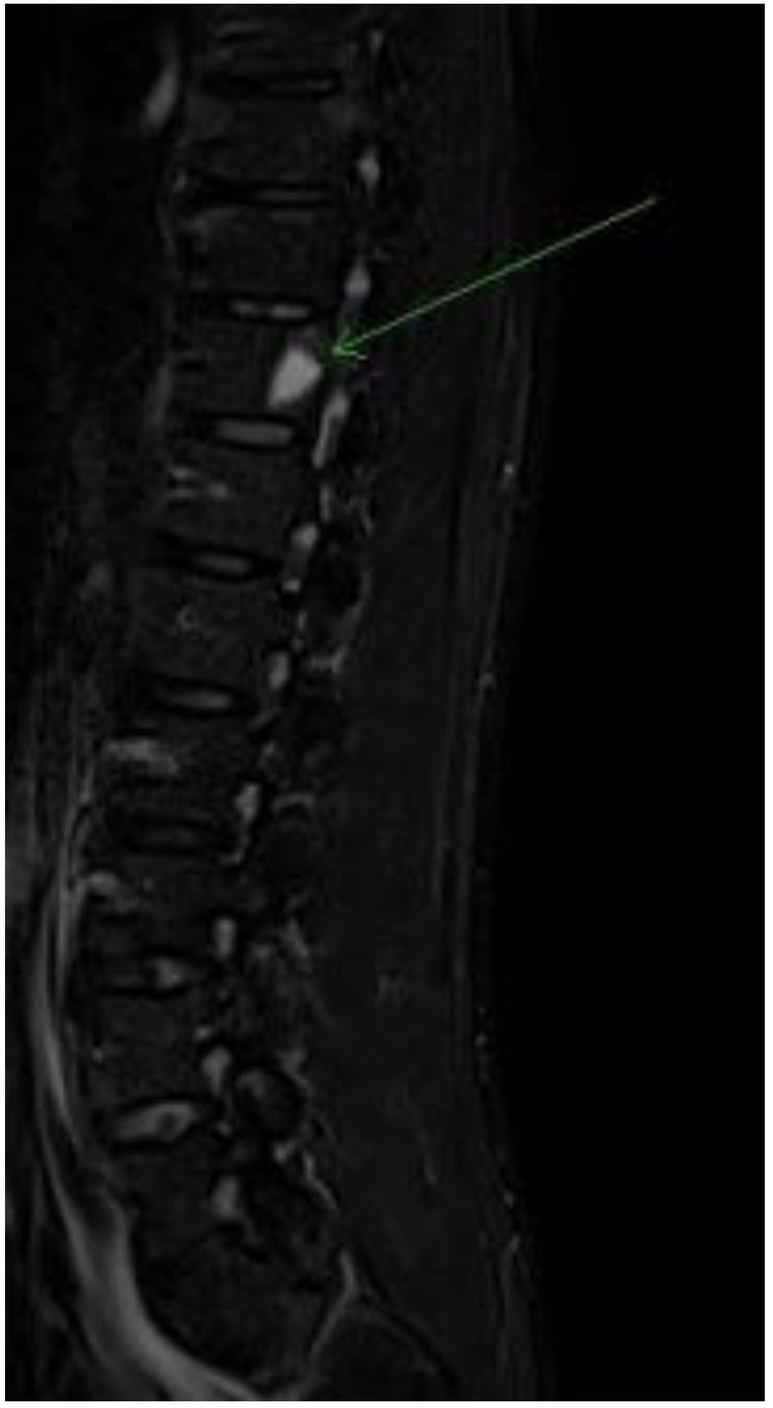


- Bilan standard normal.
- CRP < 3.
- EDP normale.
- Urines normales.
- AAN 1/640, SSA 34.
- BGSA Chisholm 2.
- Capillaroscopie normale.

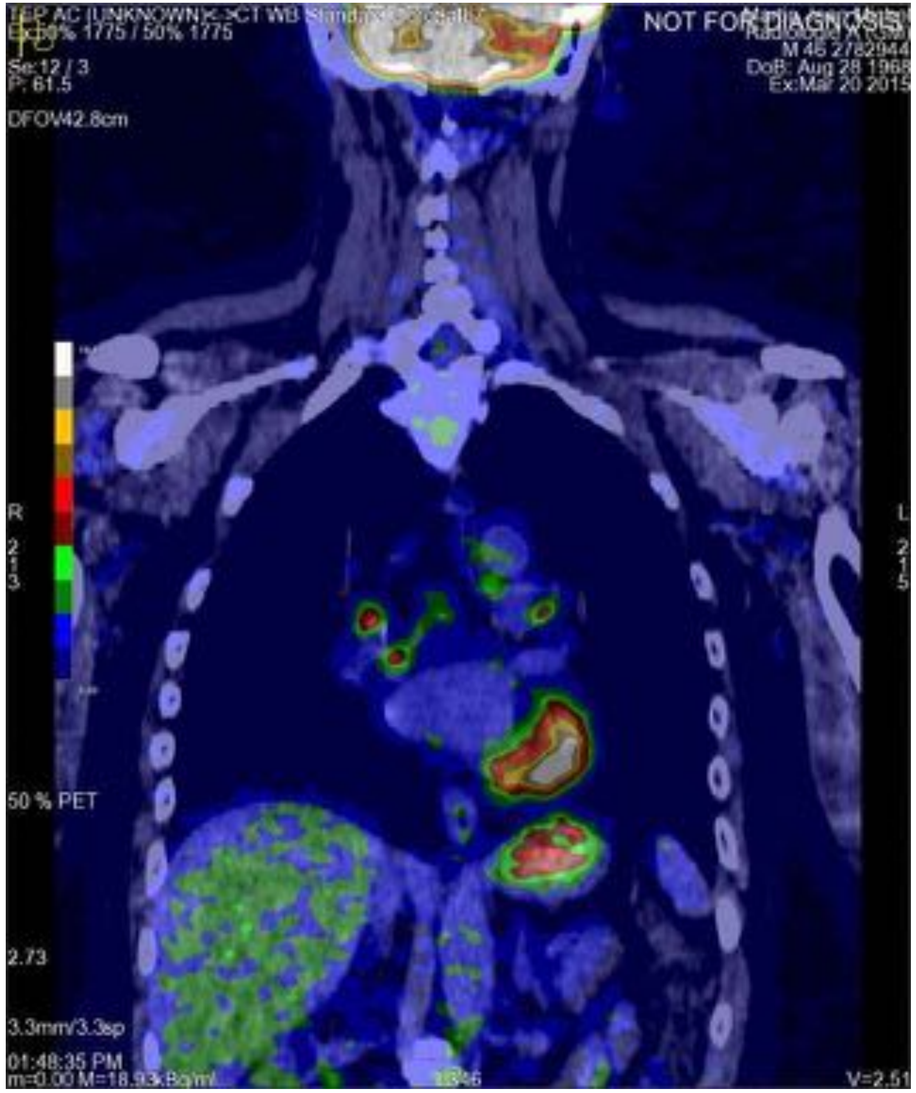
IRM

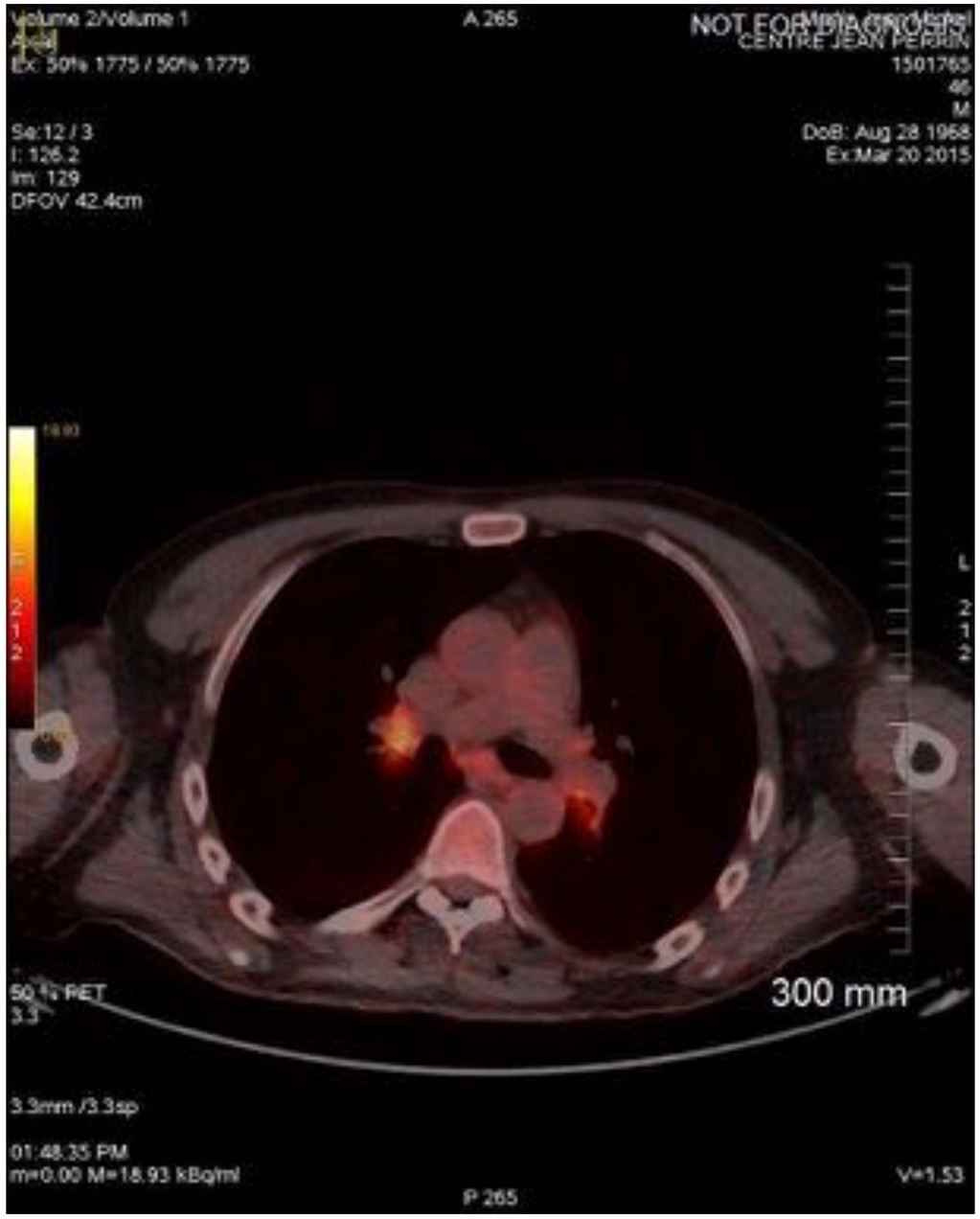



T1
T2
T3
T4
T5
T6
T7
T8
T9
T10
T11
T12
L1
L2
L3
L4



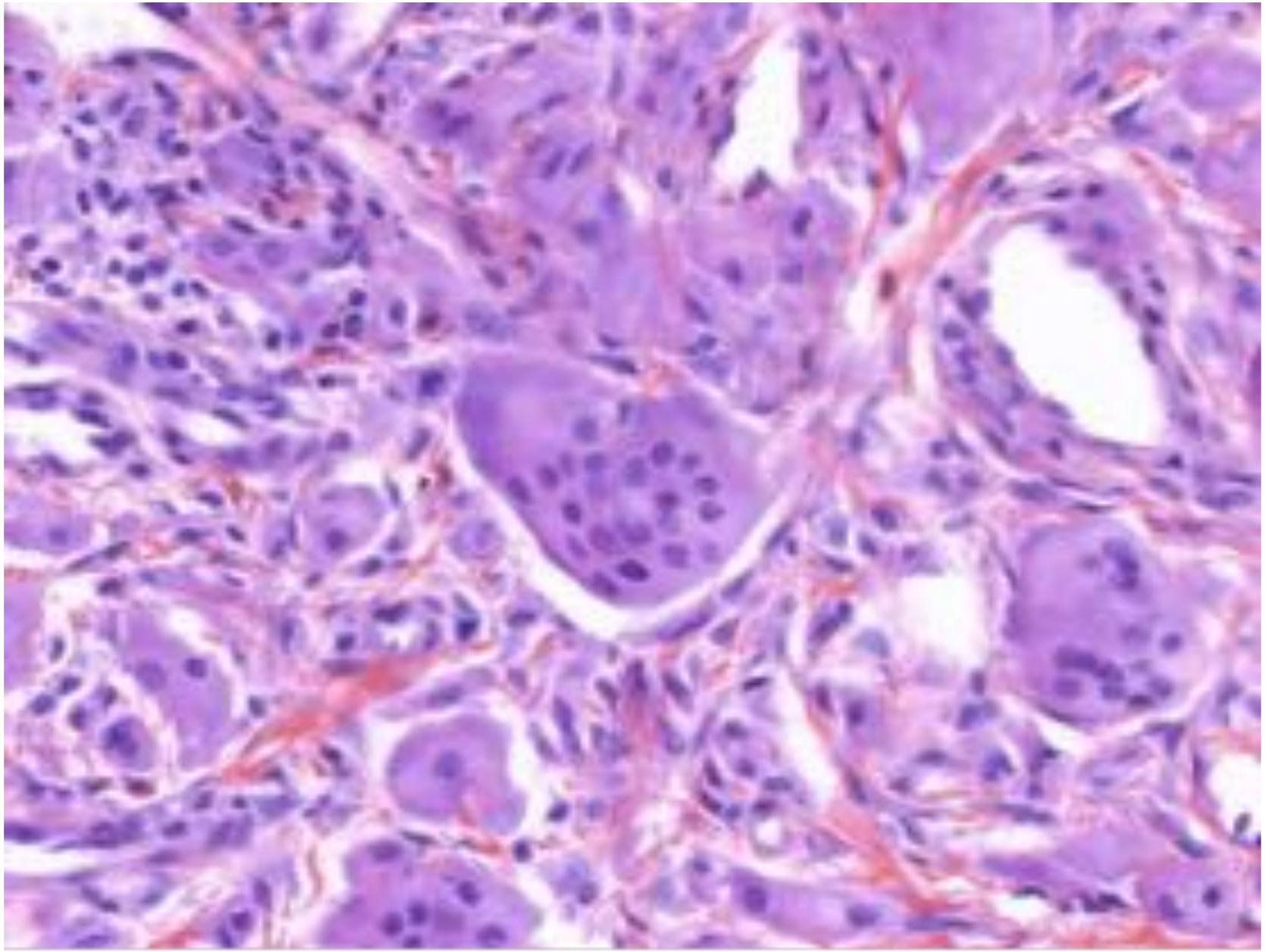
TEP


NOT FOR DIAGNOSIS
Se:12 / 3
DFOV42.8cm
50 % PET
2.73
3.3mm/3.3sp
01:48:35 PM
V=2.51


A 265
NOT FOR DIAGNOSIS
CENTRE JEAN PERRIN
Se:12 / 3
I: 126.2
Im: 129
DFOV 42.4cm
DoB: Aug 28 1968
Ex:Mar 20 2015
300 mm
3.3mm /3.3sp
01:48:35 PM
V=1.53
P 265

Biopsies

- CD68+
- CD1a et S100-



Diagnostic ?

Réticulohistiocytose multicentrique

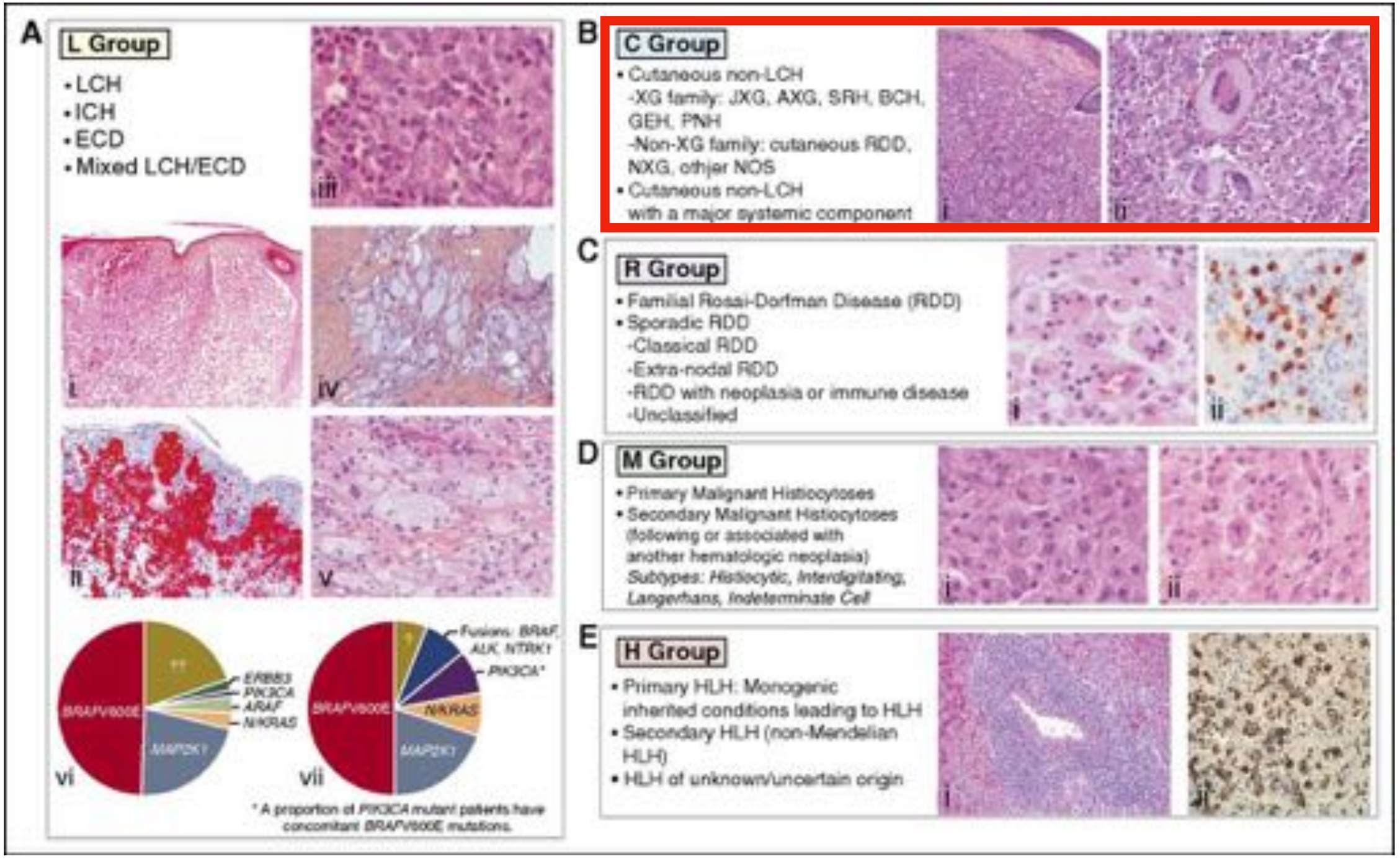
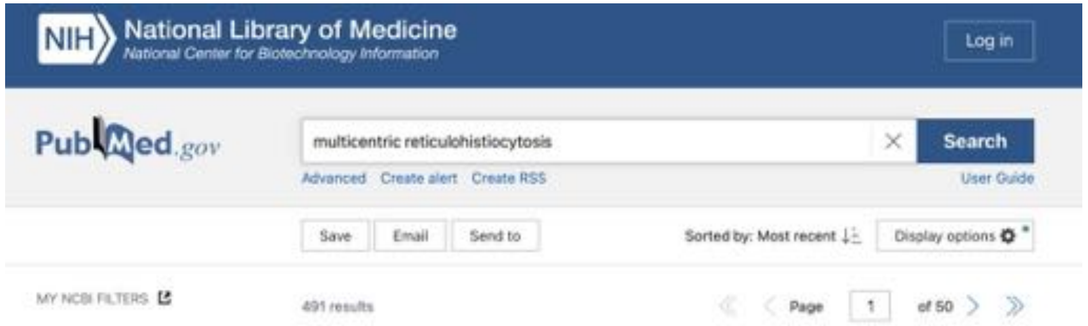

A
L Group
• LCH
• ICH
• ECD
• Mixed LCH/ECD
i
ii
iii
iv
v
vi
vii
BRAFV600E
MAP2K1
ERBB3
PIK3CA
ARAF
NRAS
Fusions: BRAF, ALK, NTRK1
PIK3CA*
* A proportion of PIK3CA mutant patients have concomitant BRAFV600E mutations.
B
C Group
• Cutaneous non-LCH
-XG family: JXG, AXG, SRH, BCH, GEH, PNH
-Non-XG family: cutaneous RDD, NXG, othjer NOS
• Cutaneous non-LCH with a major systemic component
C
R Group
• Familial Rosai-Dorfman Disease (RDD)
• Sporadic RDD
-Classical RDD
-Extra-nodal RDD
-RDD with neoplasia or immune disease
-Unclassified
D
M Group
• Primary Malignant Histiocytoses
• Secondary Malignant Histiocytoses (following or associated with another hematologic neoplasia)
Subtypes: Histiocytic, Interdigitating, Langerhans, Indeterminate Cell
E
H Group
• Primary HLH: Monogenic inherited conditions leading to HLH
• Secondary HLH (non-Mendelian HLH)
• HLH of unknown/uncertain origin

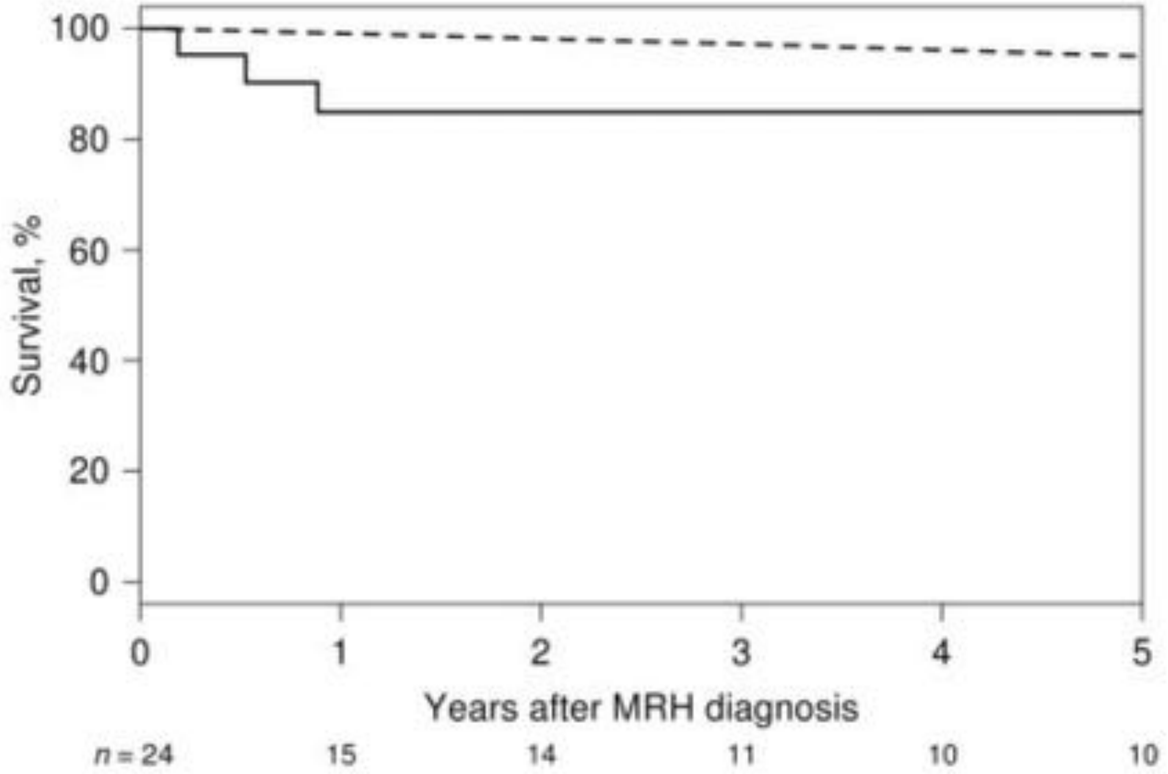
Table 2. Non-LCH of skin and mucosa (C group)

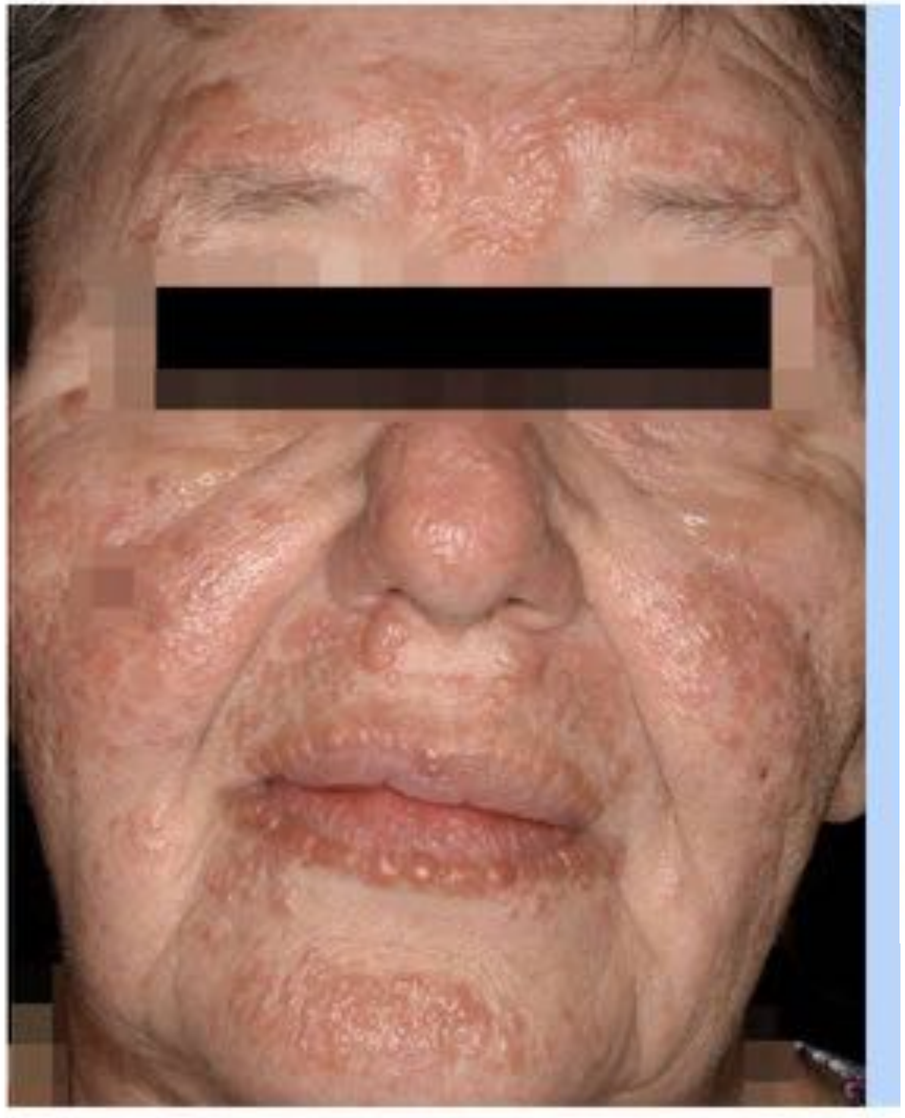
Non-LCH of skin and mucosa	
Cutaneous non-LCH histiocytoses	
XG family	JXG
	AXG
	SRH
	BCH
	GEH
	PNH
Non-XG family	Cutaneous RDD
	NXG
	Cutaneous histiocytoses not otherwise specified
Cutaneous non-LCH histiocytoses with a major systemic component	
XG family	XD
Non-XG family	MRH

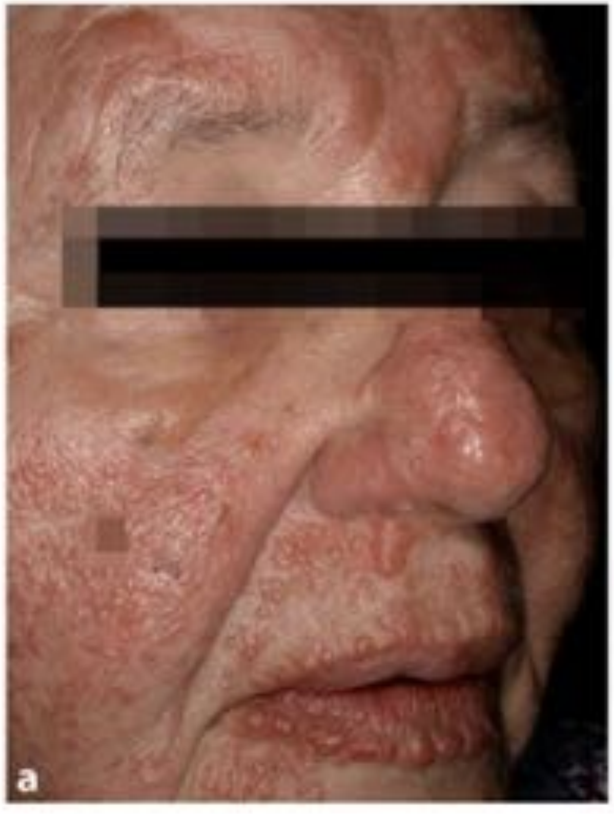
AXG, adult xanthogranuloma; BCH, benign cephalic histiocytosis; GEH, generalized eruptive histiocytosis; JXG, juvenile xanthogranuloma; MRH, multicentric reticulohistiocytosis; NXG, necrobiotic xanthogranuloma; PNH, progressive nodular histiocytosis; RDD, Rosai-Dorfman disease; SRH, solitary reticulohistiocytoma; XD, xanthoma disseminatum; XG, xanthogranuloma.

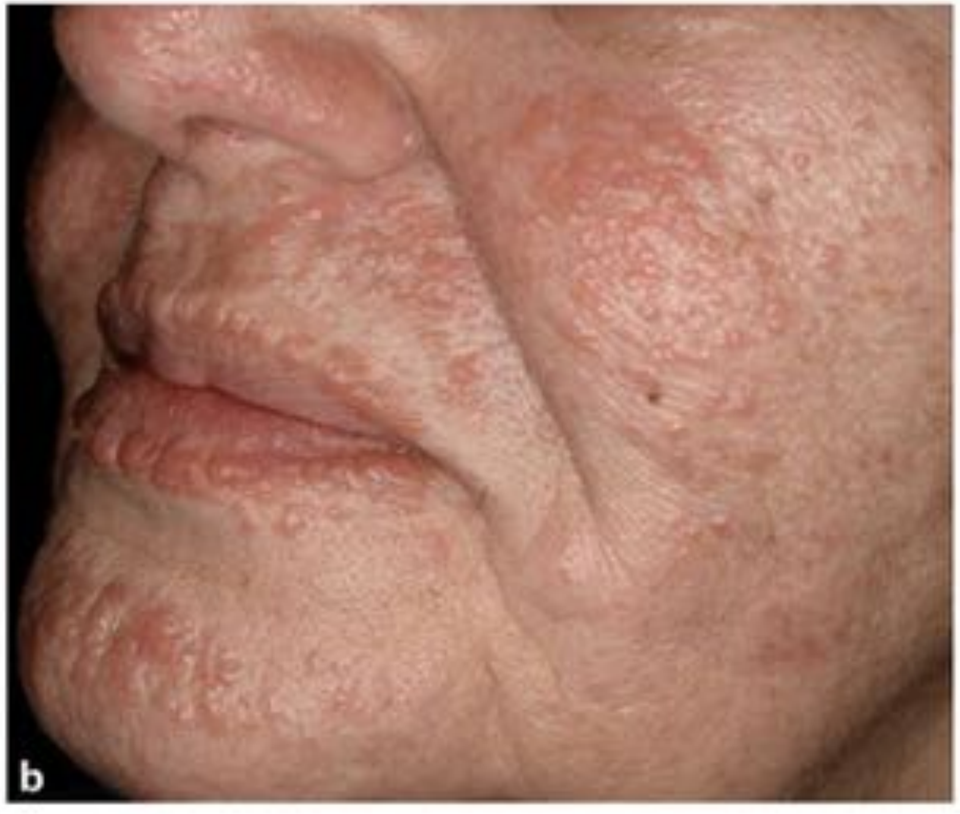


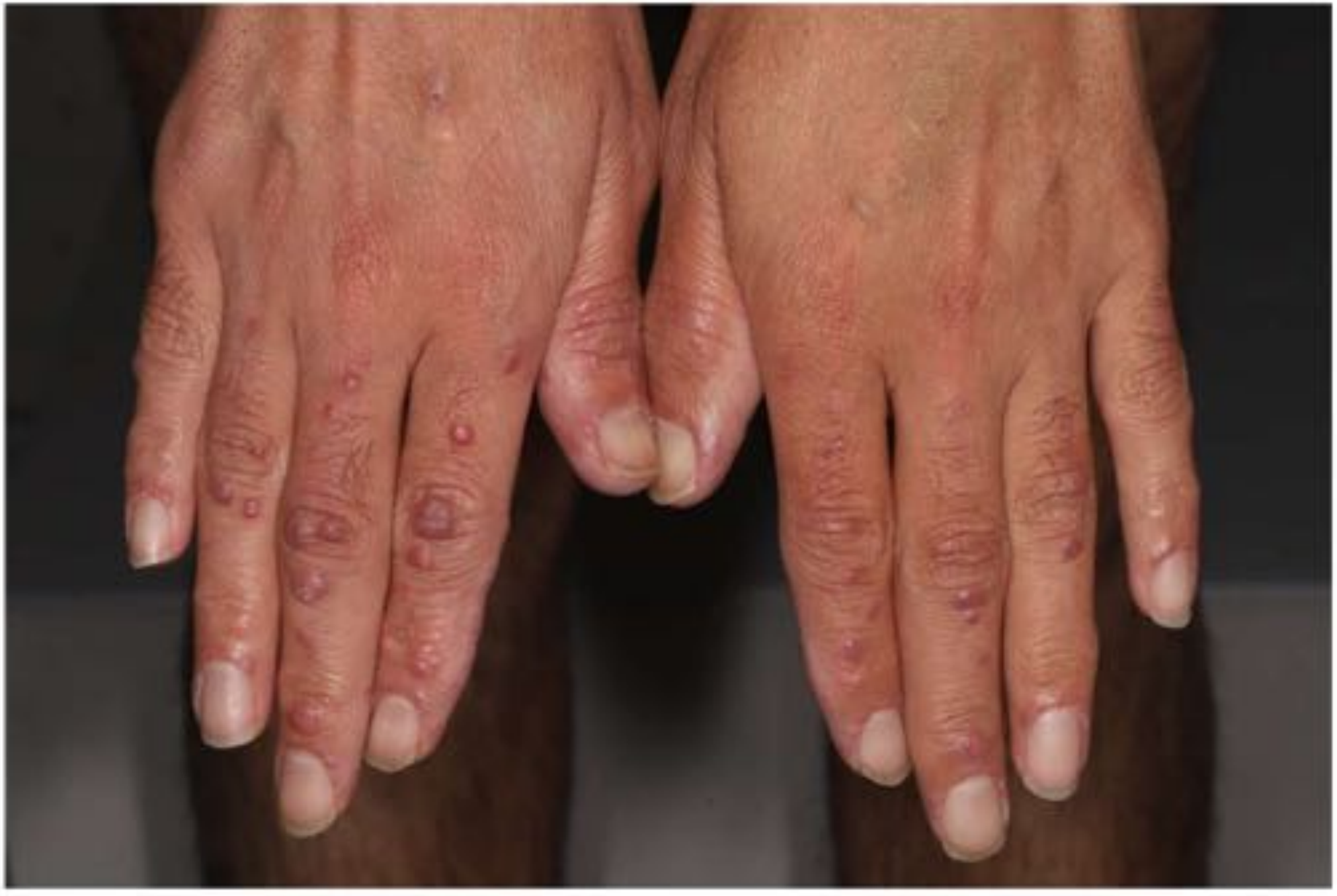
- 1937
- 2-3F/1H
- 50-60 ans
- Tableau cutané/articulaire
- Autres manifestations
- MAI associées
- 15-30% : paranéoplasique

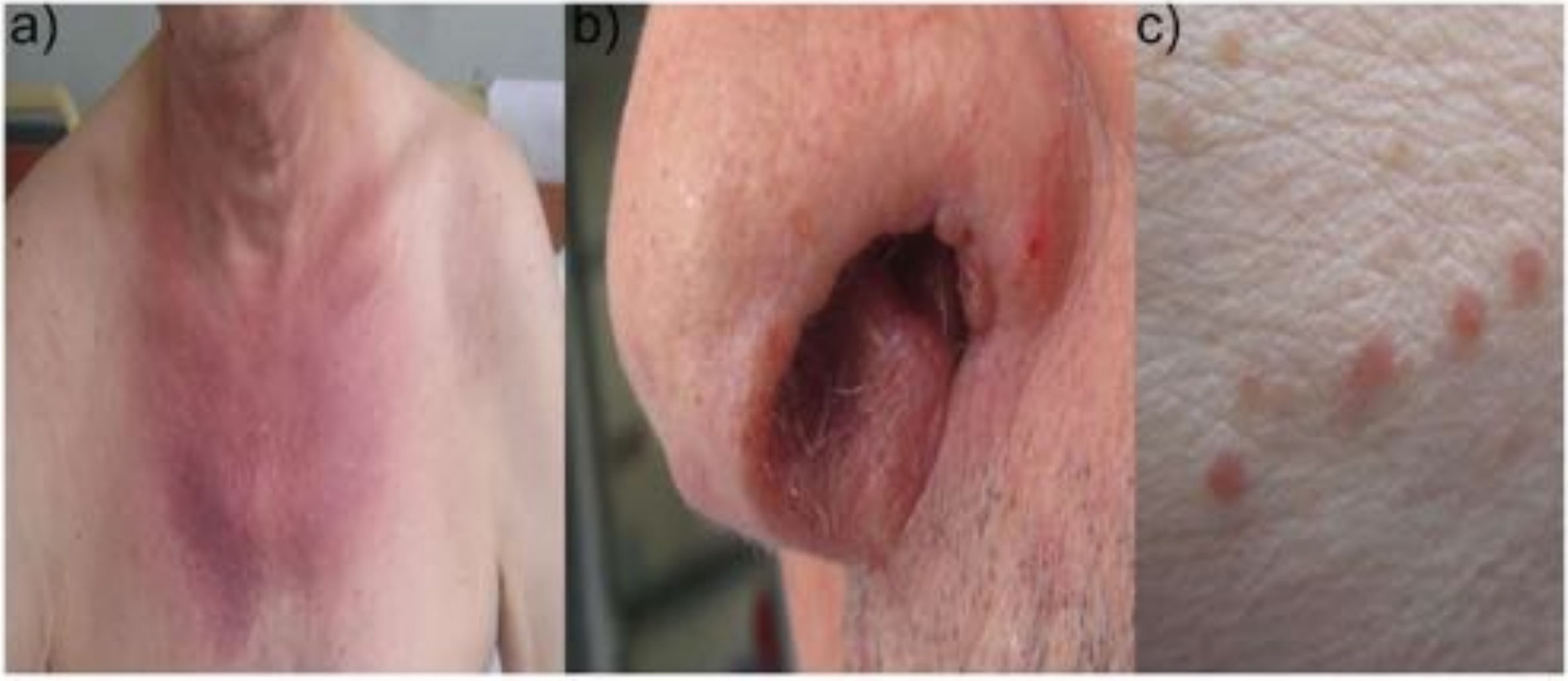





a


b




a)
b)
c)

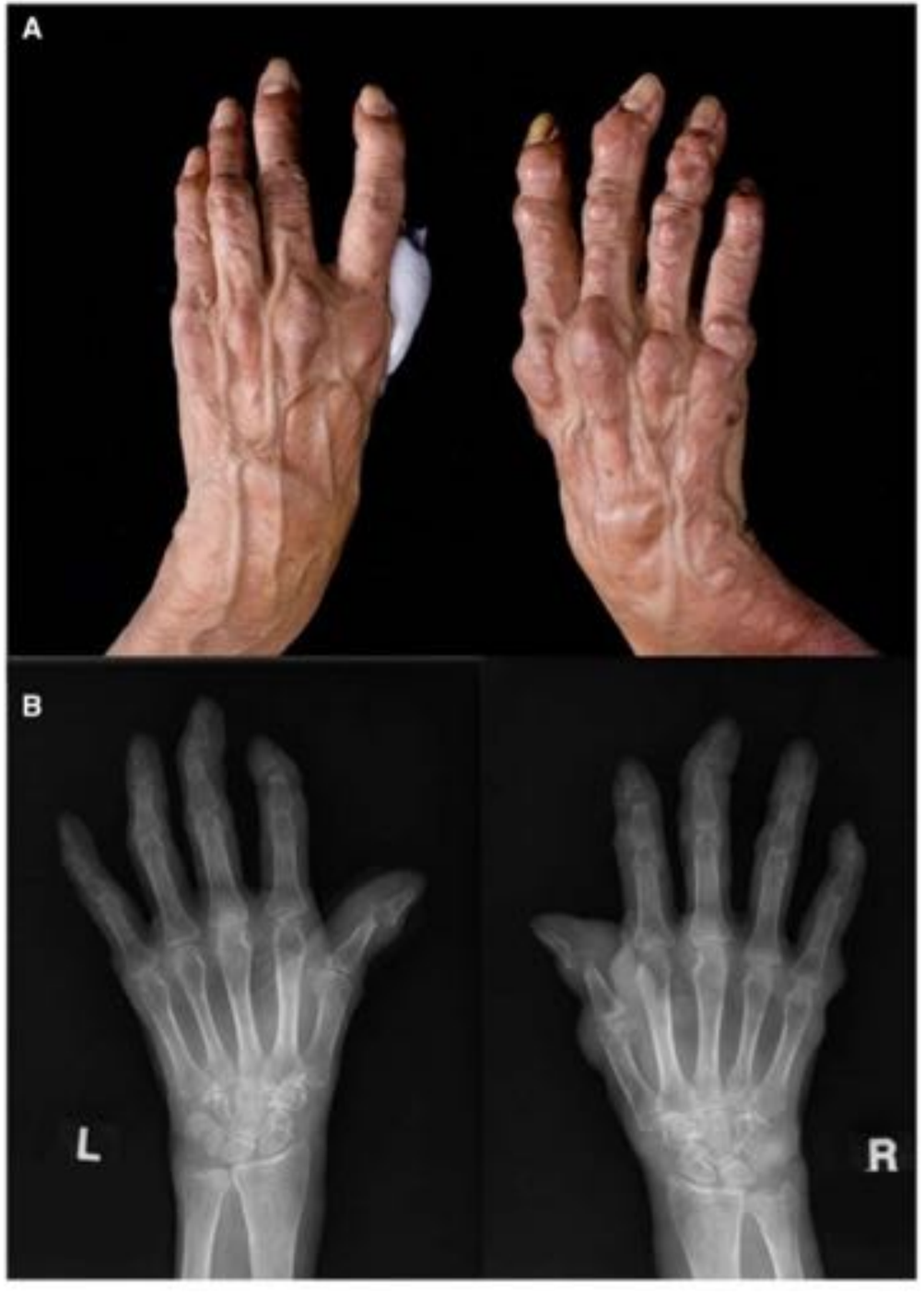

A
B
L
R

Table 1
Clinical Features of multicentric reticulohistiocytosis.

1. Constitutional	Fever, fatigue, and weight loss
2. Musculoskeletal	Symmetrical polyarthritis, involvement of the distal interphalangeal joint, erosive arthritis, and arthritis mutilans
3. Cutaneous	Papulonodular lesions, photodistributed macular eruptions, and periungual telangiectasia
4. Lung	Pleural effusion, pulmonary infiltrates, interstitial fibrosis, and hilar adenopathy
5. Heart	Pericardial effusion and myocarditis
6. Gastrointestinal	Liver and spleen
7. Urogenital	Genital tract and kidney
8. Others	Muscle, thyroid glands, salivary glands, lymph nodes, and eyes

Table 2
Differential diagnosis of MRH from rheumatology perspective.

1. Coexistent disease	Rheumatoid arthritis, systemic lupus erythematosus, Sjögren's syndrome, dermatomyositis, polymyositis, and systemic sclerosis
2. Arthritis	Nodular osteoarthritis, rheumatoid arthritis, psoriatic arthritis, reactive arthritis, and gout
3. Cutaneous	Dermatomyositis, sarcoidosis, lepromatous leprosy, granuloma annulare, and xanthoma
4. Others	Fibroblastic rheumatism, Farber's disease, histiocytosis X, Langerhans, and non-Langerhans cell histiocytosis

Table 3
MRH and malignancy.

• Respiratory	Nasopharyngeal, bronchial, and lung
• Gynecologic	Ovarian, endometrial, and cervix
• Breast	Scirrhous and invasive ductal carcinoma
• Gastrointestinal	Stomach, colon, and liver
• Hematological	Lymphoma, leukemia, and myelodysplastic syndrome
• Skin	Melanoma
• Urogenital	Renal, bladder, and penile
• Others	Sarcoma, unknown origin, mesothelioma, and thyroid cancer

Table 4
MRH and associated conditions.

Connective tissue disease	Others
• Rheumatoid arthritis	• IgG paraproteinemia
• Systemic lupus erythematosus	• Hyperlipidemia
• Sjögren's syndrome	• Pregnancy
• Systemic sclerosis	• Primary biliary cirrhosis
• Dermatomyositis	• Mycobacterial infections
• Polymyositis	• Thyroid disorders
	• Celiac disease
	• Paget disease

Traitements

Table 5
Treatment experience with MRH.

Nonbiologicals	Biologicals
• NSAID	**TNF inhibitors**
• Corticosteroid	• Adalimumab
• Methotrexate	• Etanercept
• Antimalarial	• Infliximab
• Leflunomide	• Golimumab
• Sulfasalazine	**IL-6 inhibitor**
• Bisphosphonates	• Tocilizumab
• Azathioprine	**IL-1 inhibitor**
• Vincristine	• Anakinra
• Cyclophosphamide	
• Mycophenolate mofetil	
• Tacrolimus	
• Nitrogen mustard	
• Isoniazid	
• Minocycline	
• Denosumab	

Observation

May 5, 2021

Treatment of Severe Multicentric Reticulohistiocytosis With Upadacitinib

Omid Zahedi Niaki, MD[1]; Erin Penn, MD, MS[2]; Deborah A. Scott, MD[3]; et al

» Author Affiliations

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Targetable driver mutations in multicentric reticulohistiocytosis

Norihiro Murakami, Tomohisa Sakai, Eisuke Arai, Hideki Muramatsu, Daisuke Ichikawa, Shuji Asai, Yoshie Shimoyama, Naoki Ishiguro, Yoshiyuki Takahashi, Yusuke Okuno, Yoshihiro Nishida

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