Inflammation Orbitaire
(Cas cliniques)

Deauville 2016

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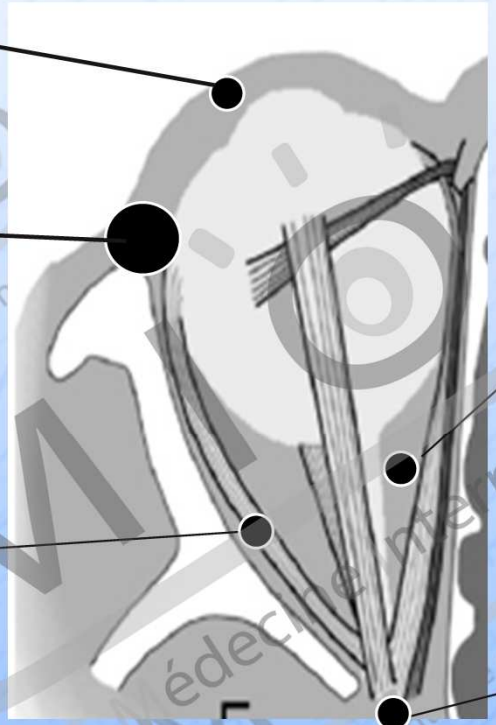
Formes Anatomocliniques

Xanthogranulomes palpébraux

Dacryoadénites

Myosites

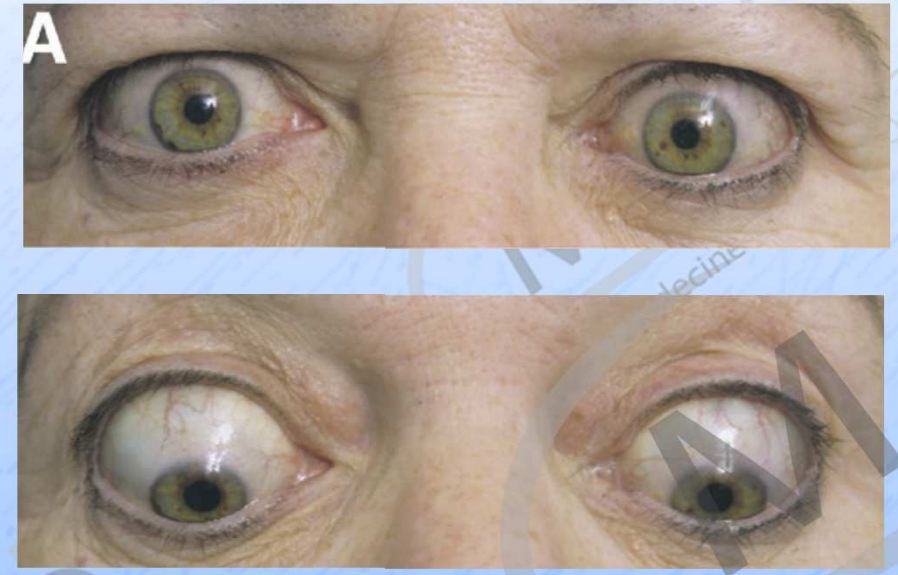


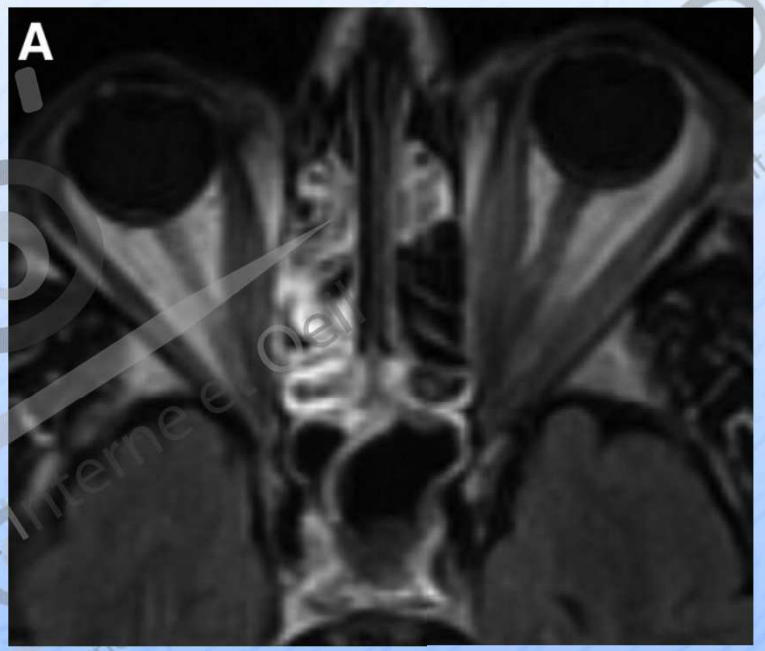
Graisse (atteinte diffuse)

Apex

Cas clinique 1

Femme 50 ans,
Diplopie binoculaire





Cas clinique n° 1

1. Lymphome orbitaire
2. Maladie associée aux IgG4
3. Orbitopathie Basedowienne
4. Sarcoïdose
5. Granulomatose avec polyangéite

Cas clinique n° 1

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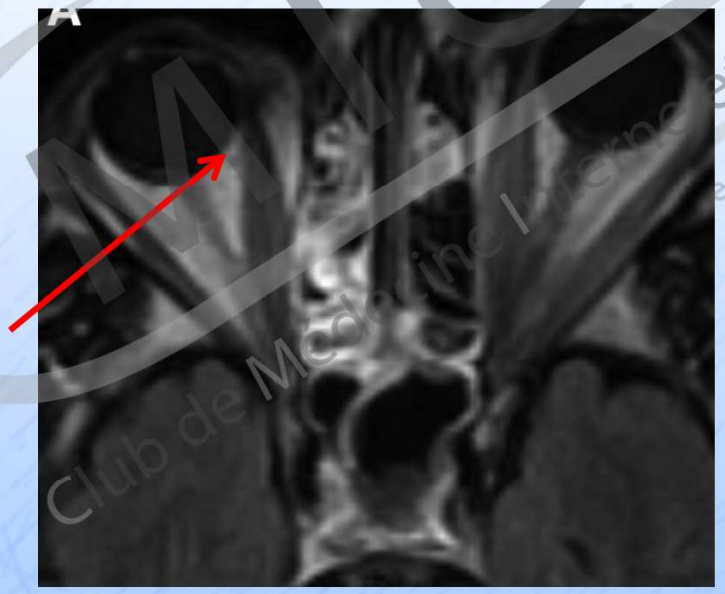
Orbitopathies dysthyroïdiennes

- **Maladie de Basedow (95%)**
- **Rétraction palpébrale!**



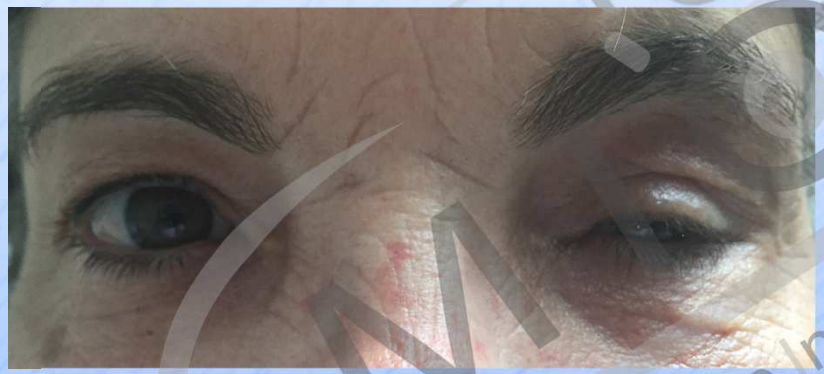
- **TRAK +, TSH (parfois normale!)**

- **IRM orbitaire:**

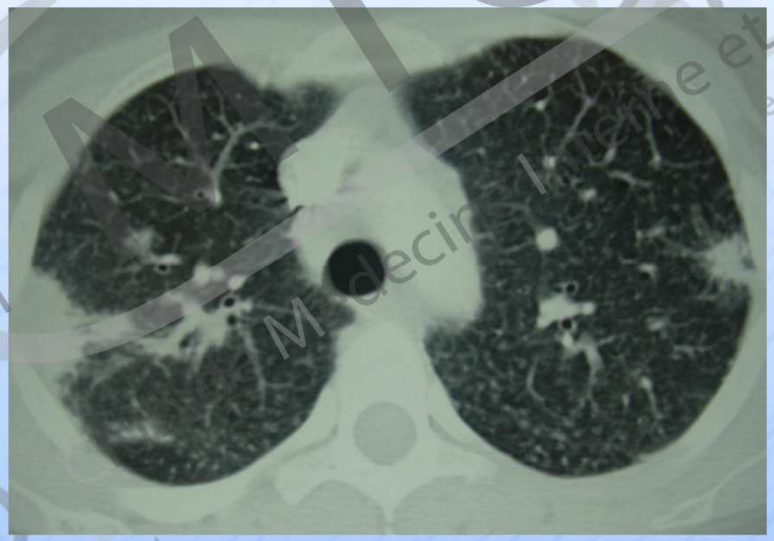
Respect du tendon +++

Cas clinique n° 2

Femme 50 ans,
Oedème palpébral, ptosis
Glande lacrymale hypertrophiée



+ Toux



Quel(s) diagnostic(s) évoqués?

Cas clinique n° 2

1. Lymphome orbitaire
2. Maladie associée aux IgG4
3. Orbitopathie Basedowienne
4. Sarcoïdose
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Cas clinique n° 2

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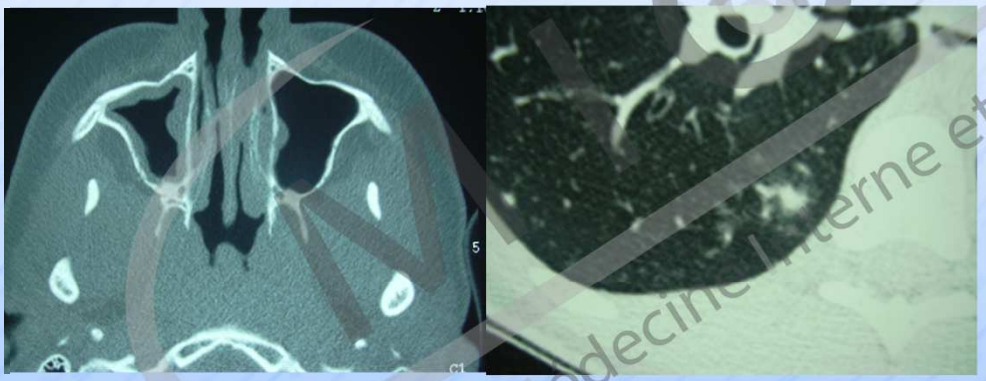
Orbitopathies et MAI

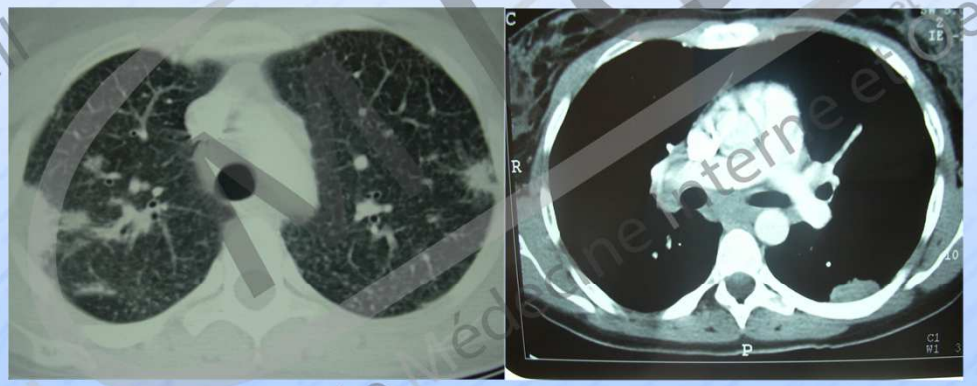
Dacryoadénite (bilatérale) + atteinte pulmonaire

Granulomatose avec polyangéite



→ ANCA, BU (GB, GR)

Sarcoïdose



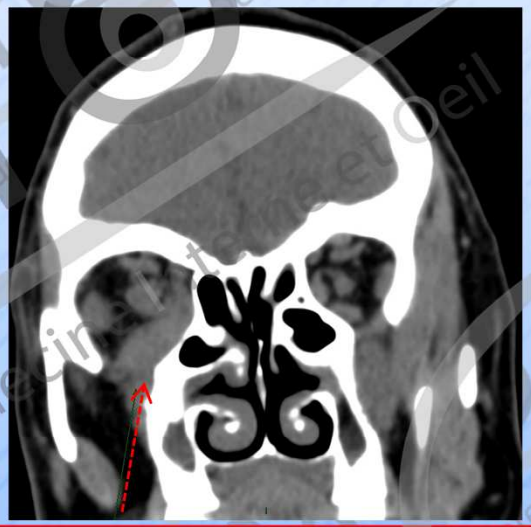
→ BGSA, Fibro bronchique + LBA

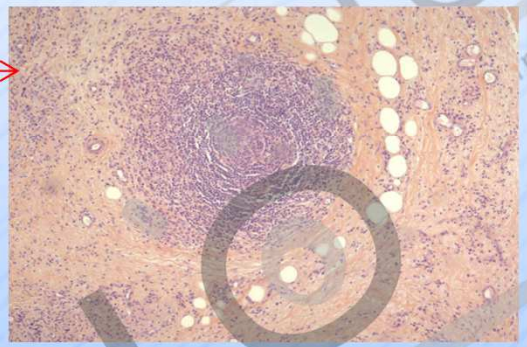
75 % des MAI associées aux orbitopathies !

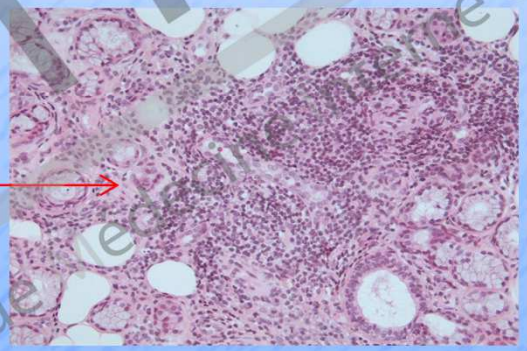
Abad et al. Rev Med Interne 2012

Cas clinique n° 3

50 ans
Asthme de révélation tardive
“Gonflement des glandes lacrymales”







AC anti-SSA/SSB négatifs

Cas clinique n° 3

1. Lymphome orbitaire
2. Granulomatose avec polyangéite
3. Sarcoïdose oculaire
4. Gougerot-Sjögren
5. Maladie associée aux IgG4

Cas clinique n° 3

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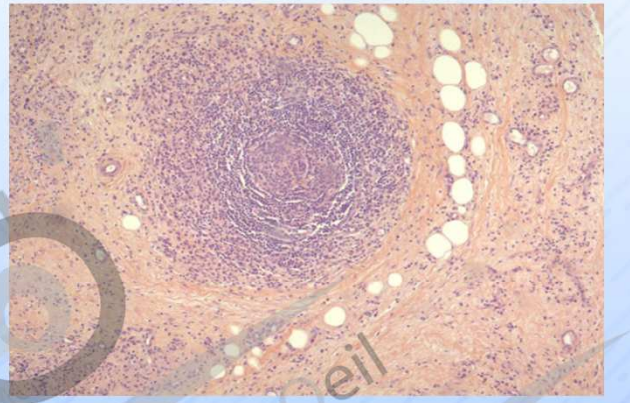
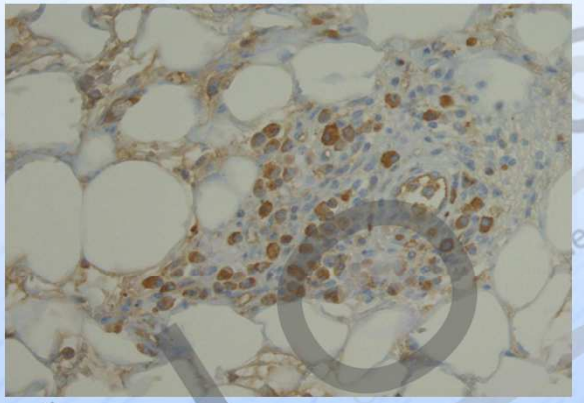
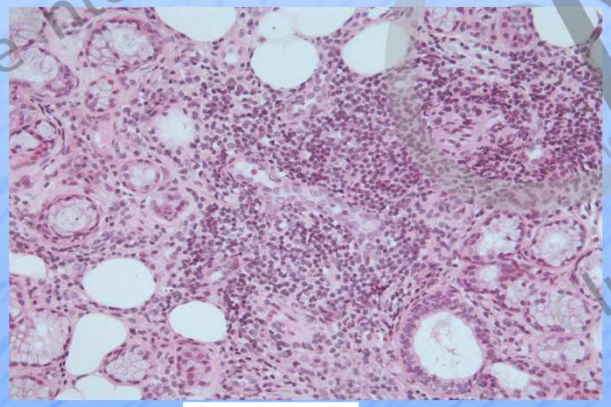
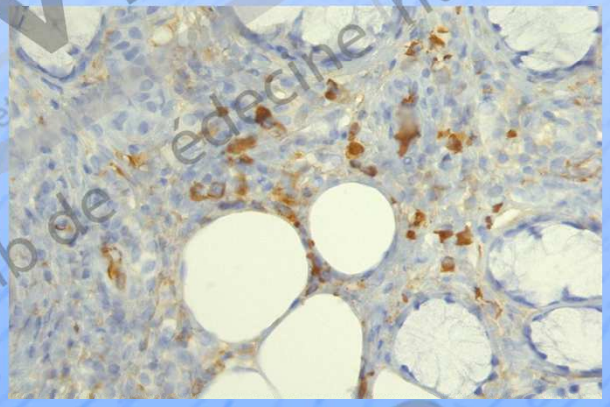
Cas clinique n° 3





+ IgG4 sérique: 7,8 g/l !





High prevalence of IgG4-related lymphoplasmacytic infiltrative disorder in 25 patients with orbital inflammation: a retrospective case series

Romain Deschamps,[1] Lydia Deschamps,[2] Raphael Depaz,[1] Sophie Coffin-Pichonnet,[3] Georges Belange,[4] Pierre Vincent Jacomet,[3] Catherine Vignal,[3] Paul Benillouche,[3] Marie Laure Herdan,[3] Marc Putterman,[5] Anne Couvelard,[2] Olivier Gout,[1] Olivier Galatoire[3]

Critères Umehara

- Lympho-plasmocytes IgG4 (> 10/ HpF et IgG4/IgG total plasma cells > 40%)
- Fibrose (Storiforme)
- IgG4 sérique > 135 mg/l
- Eliminer diagnostics différentiels

Okazaki K et Umehara H. Int J Rheumatol. 2012

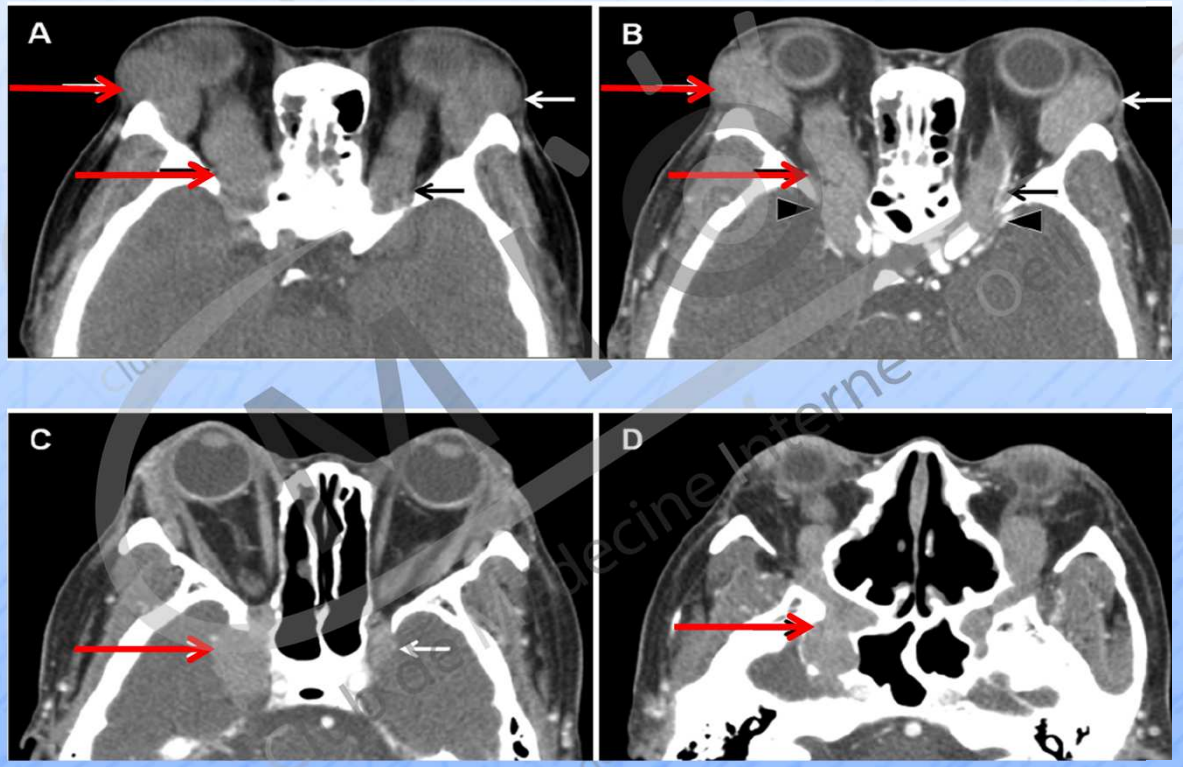
Table 1 Clinical and radiographic data for IgG4-positive and IgG4-negative patients

Patient/ sex/age	Location	Pain	Eyelid or periocular swelling/mass	Other clinical history
IgG4 positive (n=10) (40%)				
1/F/31	LG, EOM, orb	Yes	Yes	Iodine allergy
2/F/81	LG	No	Yes	Breast carcinoma, pacemaker
3/M/40	Orb, ON	No	Yes	Malaria
4/F/57	LG, EOM, orb, ON	Yes	Yes	Ovarian cystectomy, eczema,chronic urticaria
5/F/52	Orb, LG	Yes	Yes	Retinal detachment
6/F/65	Orb, EOM,ON	No	Yes	Nephritis, bladder carcinoma, diabetes mellitus
7/F/28	LG, orb, apex, EOM	No	Yes	Asthma; recurrent sinusitis; left optic neuritis 1 and 4 years earlier
8/M/55	LG	No	Yes	Iodine allergy, bronchomediastinal lymphadenopathy
9/M/26	LG	No	Yes	
10/F/62	LG	No	Yes	Thyroidectomy

Br J Ophthalmol 2013;**97**:999–1004. doi:10.1136/bjophthalmol-2013-303131

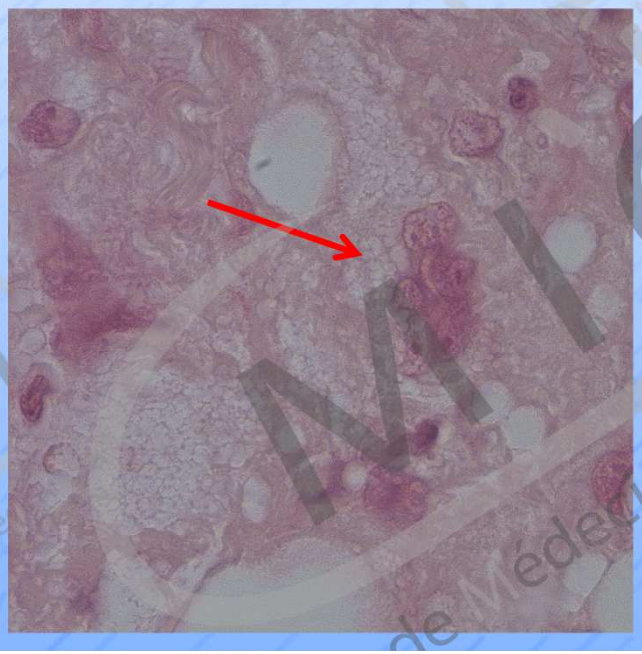
Ocular adnexal IgG4-related disease: CT and MRI findings

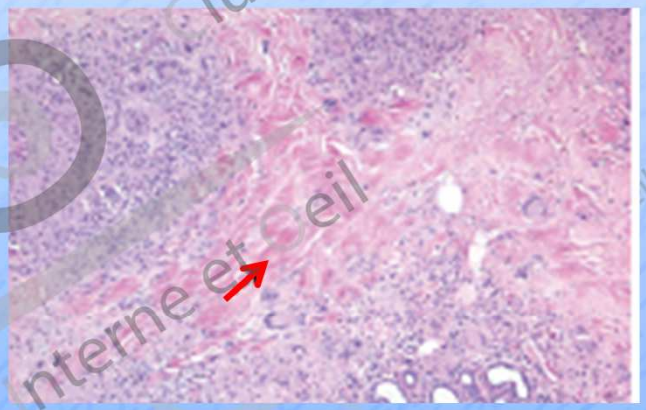
Yong Sub Song,[1] Ho-Kyung Choung,[2,3] Sun-Won Park,[1,4] Ji-Hoon Kim,[1] Sang In Khwarg,[3] Yoon Kyung Jeon[5]



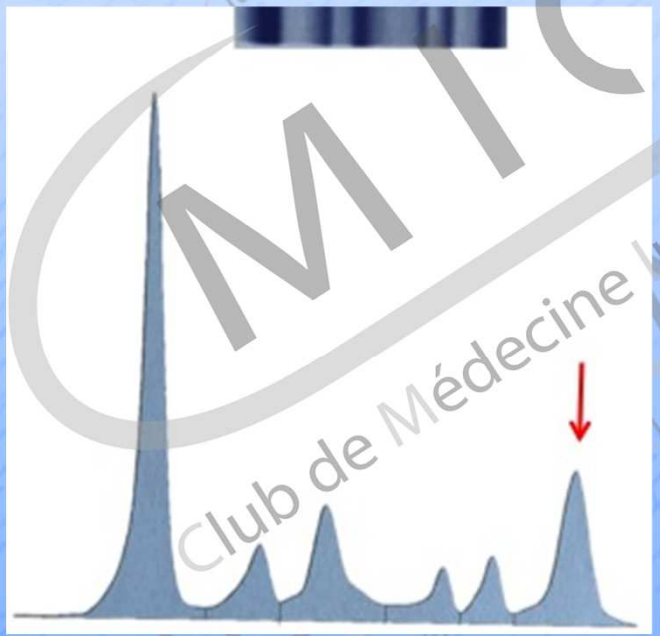
Song YS, *et al. Br J Ophthalmol* 2013;**97**:412–418.

Cas clinique n° 4





+ Nécrose collagène !



Cas clinique n° 4

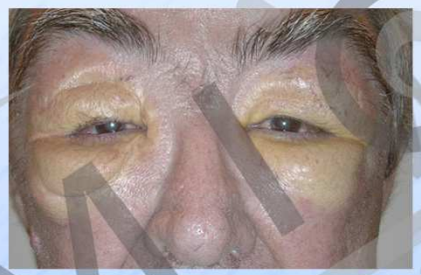
1. Xanthogranulomatose nécrobiotique
2. Maladie de Gaucher
3. Syndrome AAPOX
4. Maladie d'Erdheim Chester
5. Granulomatose avec polyangéite

Cas clinique n° 4

1. **Xanthogranulomatose nécrobiotique**
2. **Maladie de Gaucher**
3. **Syndrome AAPOX**
4. **Maladie d'Erdheim Chester**
5. **Granulomatose avec polyangéite**

Cas clinique n° 5

- Homme, 65 ans
- Martinique
- Exploitant agricole
- Asthme tardif
- HDM
  - 1998 Infiltration palpébrale bilatérale
  - 2005 Trouble de l'occlusion des yeux
- Tuméfaction jugale/ Adénopathies cervicales



- *Gammaglobulines: 16 gr/l (polyclonale)*
- *PNE: 900/mm³*

Cas clinique n° 5

- Homme, 65 ans
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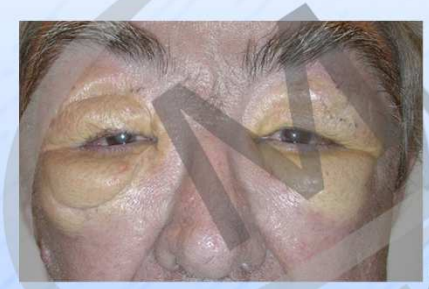
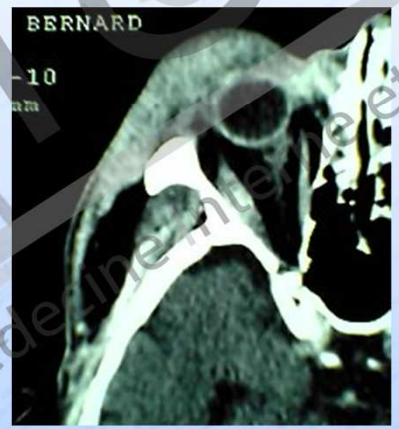
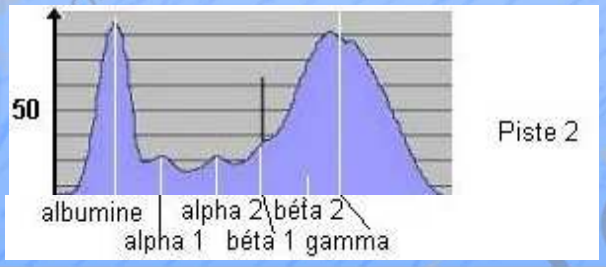




- *Gammaglobulines: 16 gr/l*
- *PNE: 900/mm³*



Cas clinique n° 5

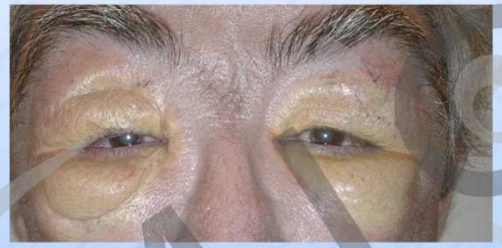
1. Xanthogranulomatose nécrobiotique
2. Maladie d'Erdheim Chester
3. Syndrome AAPOX
4. Granulomatose avec polyangéite
5. Maladie de Kimura

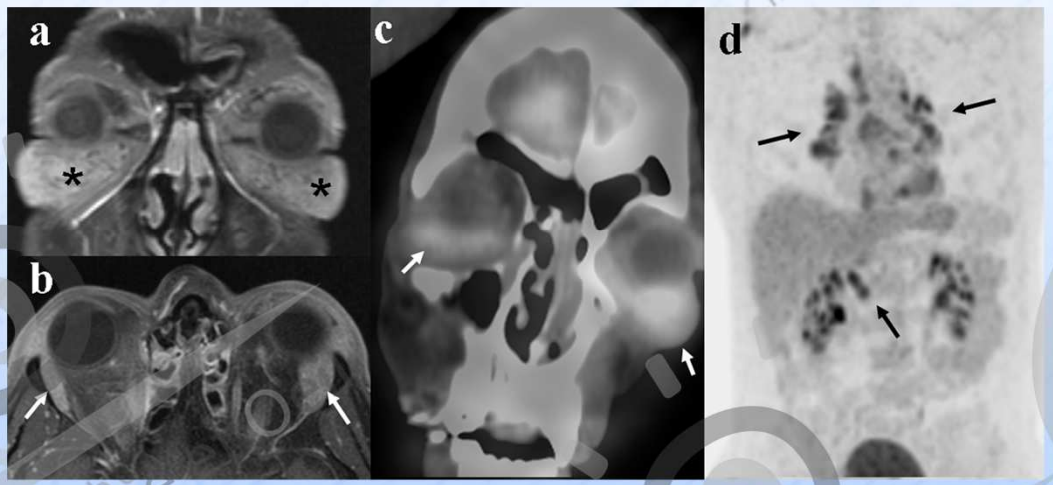
Cas clinique n° 5

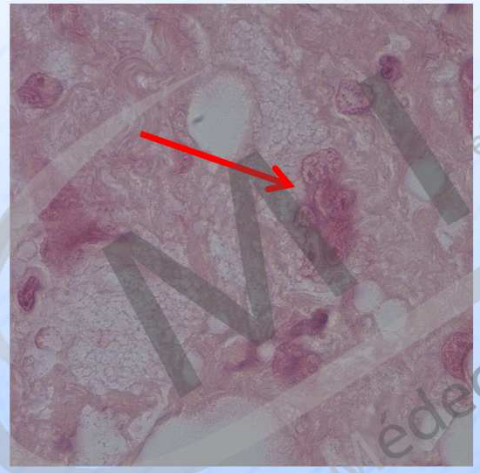
1. Xanthogranulomatose nécrobiotique
2. Maladie d'Erdheim Chester
3. Syndrome AAPOX
4. Granulomatose avec polyangéite
5. Maladie de Kimura

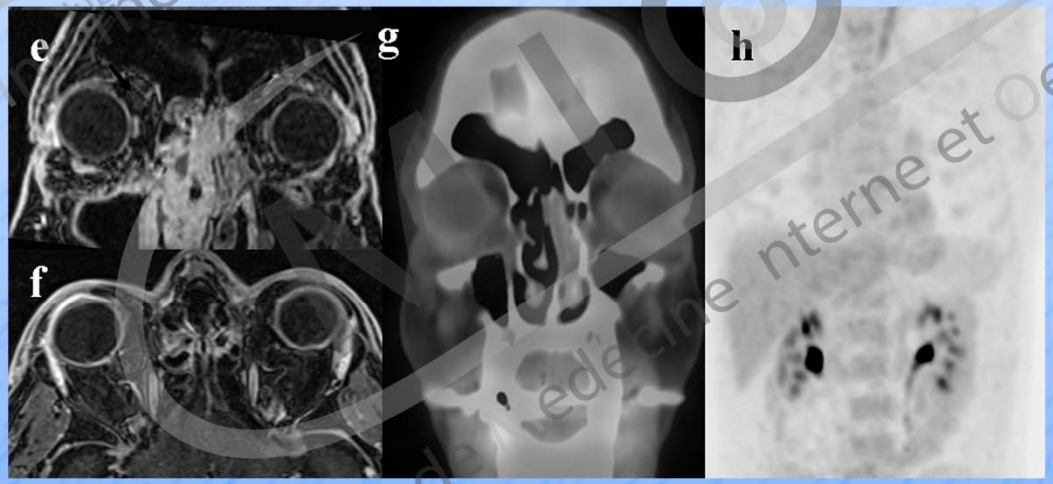
Adult Asthma and PeriOrbital Xanthogranuloma

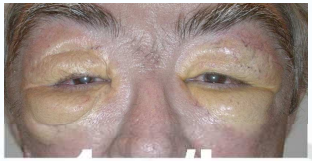
London et al. Ocul Plast Reconstr Surg 2013





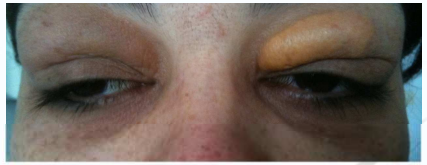






1 g/l

6,5 g/l



9,2 g/l

Témoin positif (IgG4+)

7,8 g/l

Témoin (IgG4-)

0,2 g/l

MIO Club de Médecine Interne et Oeil

A1 A2 A3 A4 A5

ORBITE

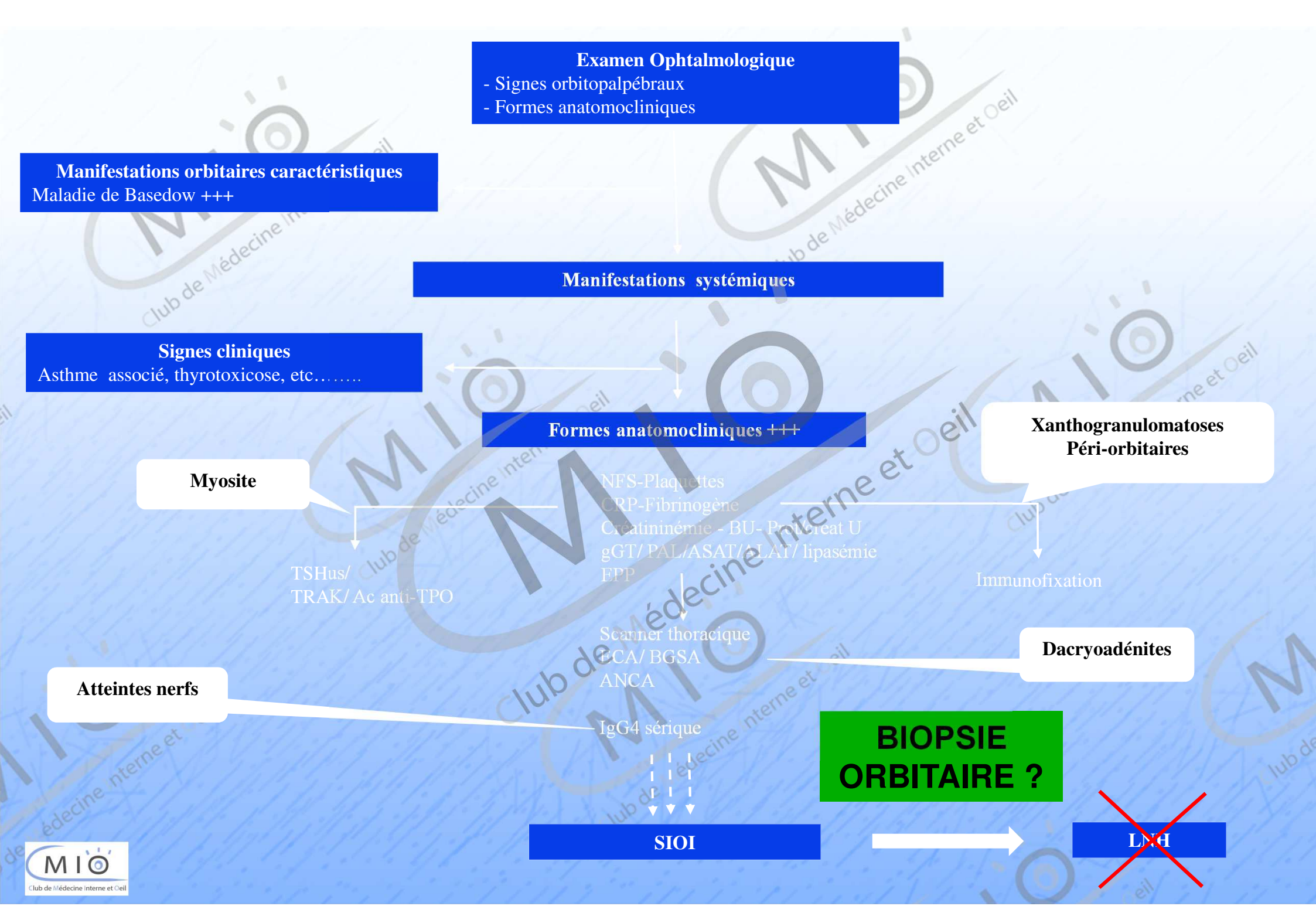
A1* A2* A3* A4* A5*

B1 B2 B3 B4 B6

GLANDE SALIVAIRE

B1* B2* B3* B4* B6*

Merci pour votre attention


Examen Ophtalmologique
- Signes orbitopalpébraux
- Formes anatomocliniques
Manifestations orbitaires caractéristiques
Maladie de Basedow +++
Manifestations systémiques
Signes cliniques
Asthme associé, thyrotoxicose, etc…….
Formes anatomocliniques +++
Myosite
Xanthogranulomatoses
Péri-orbitaires
NFS-Plaquettes
CRP-Fibrinogène
Créatininémie - BU- Protéicréat U
gGT/ PAL/ASAT/ALAT/ lipasémie
EPP
TSHus/
TRAK/ Ac anti-TPO
Immunofixation
Scanner thoracique
ECA/ BGSA
ANCA
Dacryoadénites
Atteintes nerfs
IgG4 sérique
BIOPSIE
ORBITAIRE ?
SIOI
LNH