

PARIS  
76<sup>e</sup> CONGRÈS  
SNFMI  
CONGRÈS FRANCO/MAGHRÉBIN

7 Décembre 2017



## CAS CLINIQUES-UYEITES 3

*Sébastien Abad*

Service Médecine Interne. Hôpital Avicenne. Bobigny



# Formes Anatomocliniques

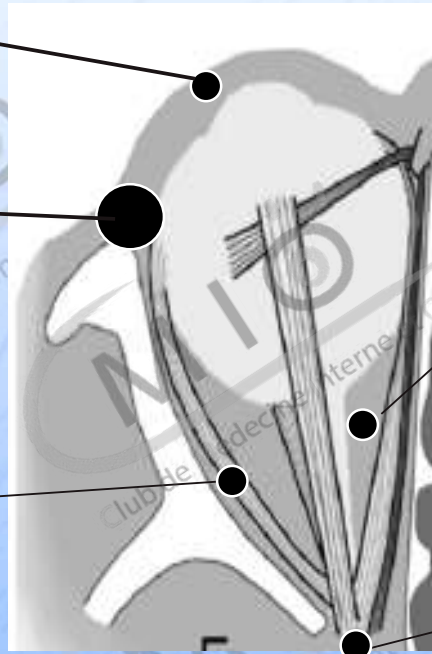
**Graisse palpébrale**

**Dacryoadénites**

**Myosites**

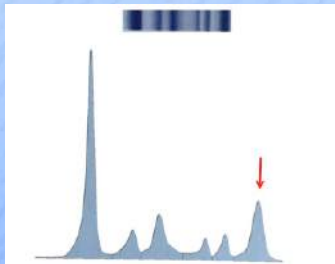
**Graisse  
(atteinte diffuse)**

**Apex**



# Patient 42 ans

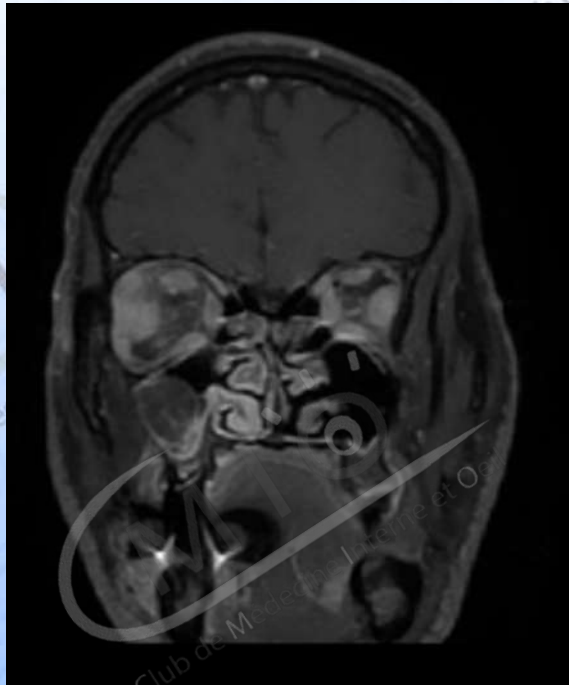
- Septembre 2011:  
« gonflement palpébral »
- Mai 2012:
  - 7 kg
  - Fièvre vespérale: 38,5° C
  - Ggl cerv. / axil. < 1 cm



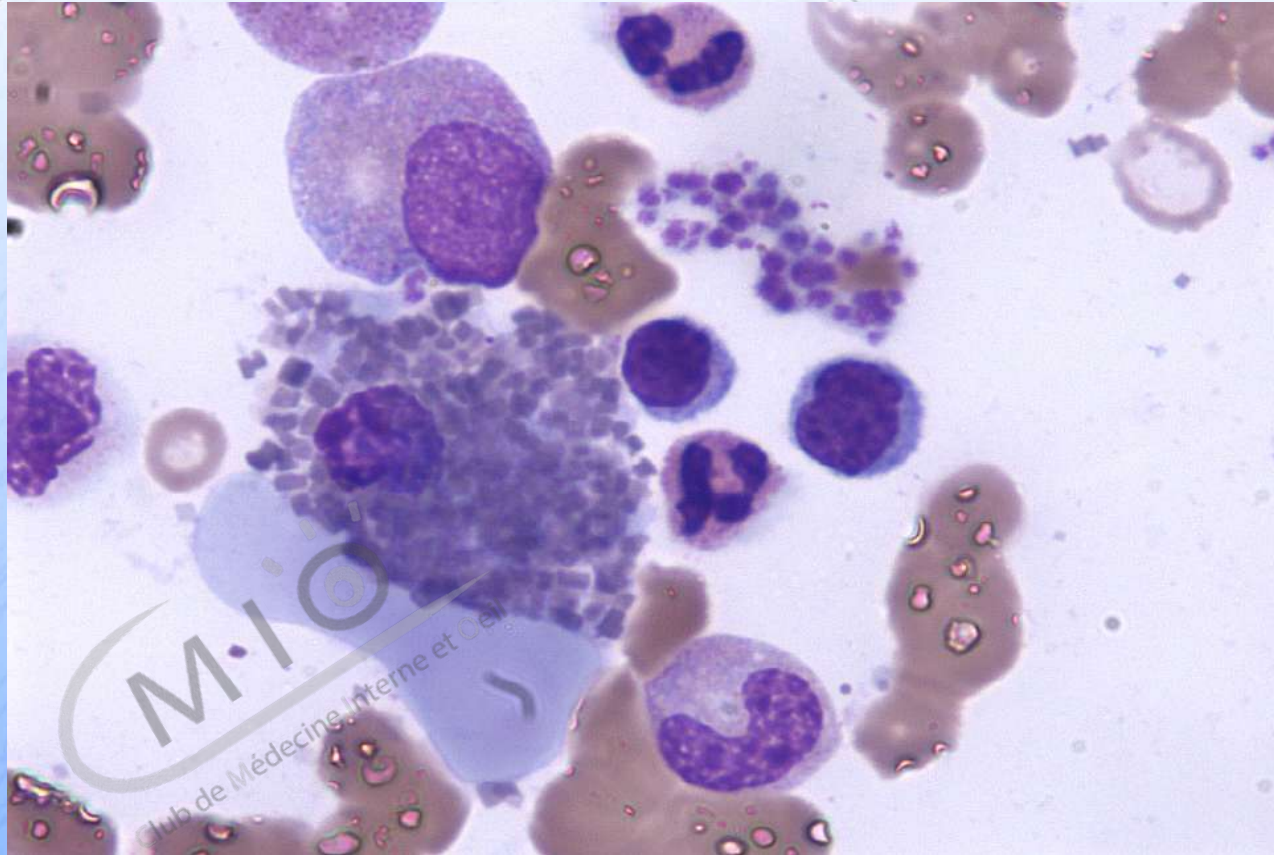
- Pic  $\Leftrightarrow$  IgA Kappa
- IgA: 9 gr/l, IgG: 12gr/l, IgM: 0.4 gr/l
- Ratio K/ $\lambda$ : 1.8 (N<1.8)



# IRM orbitaire

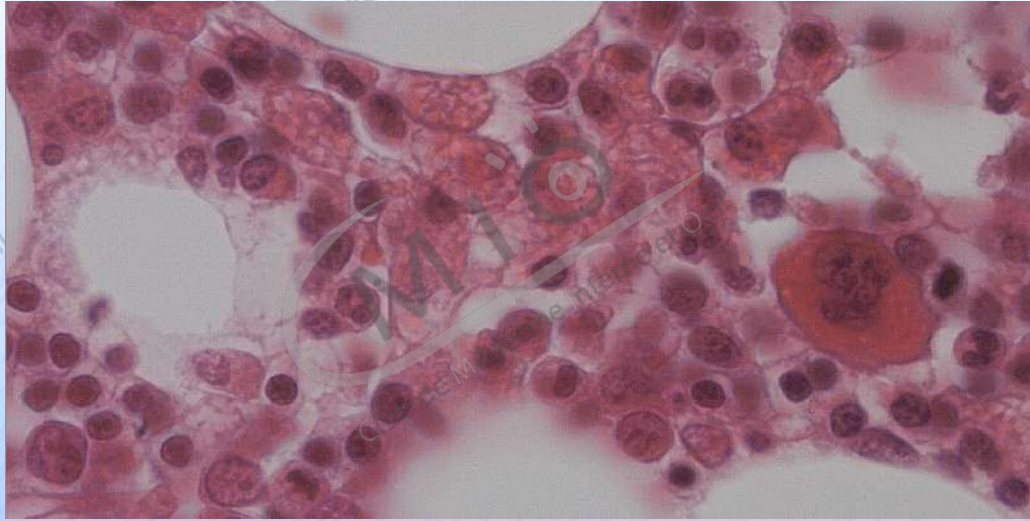


# Myèlogramme

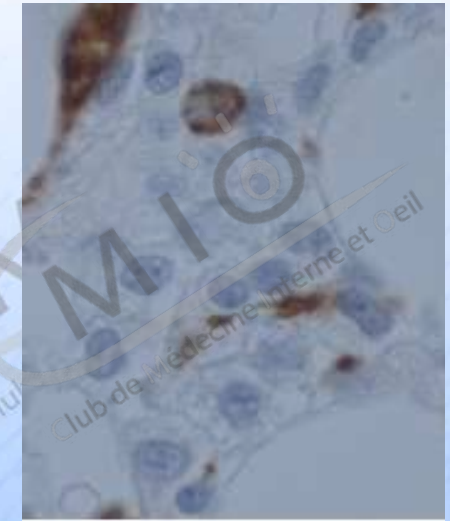
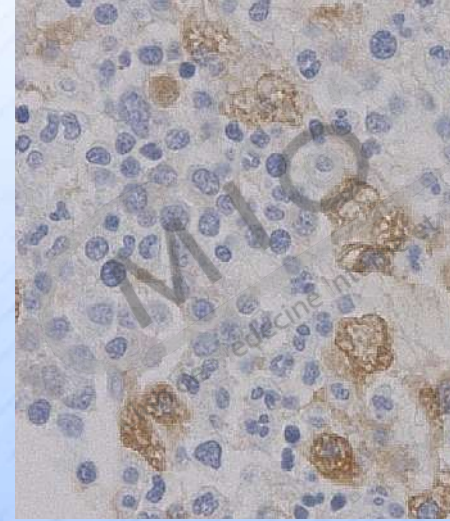


# Biopsie Ostéoméduillaire

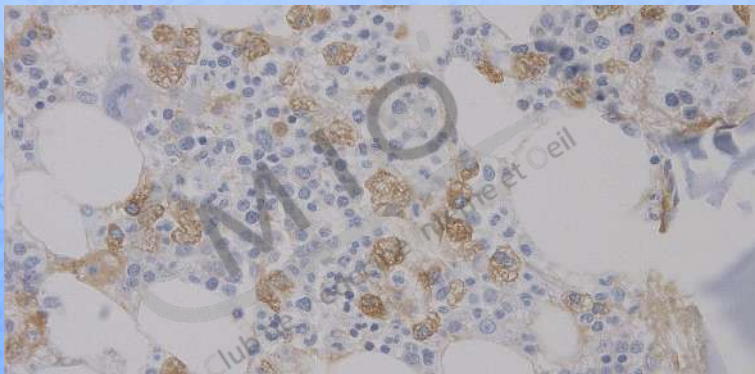
CD138



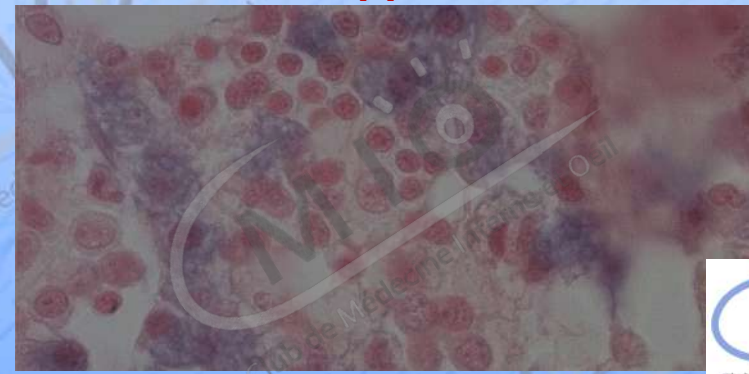
CD68



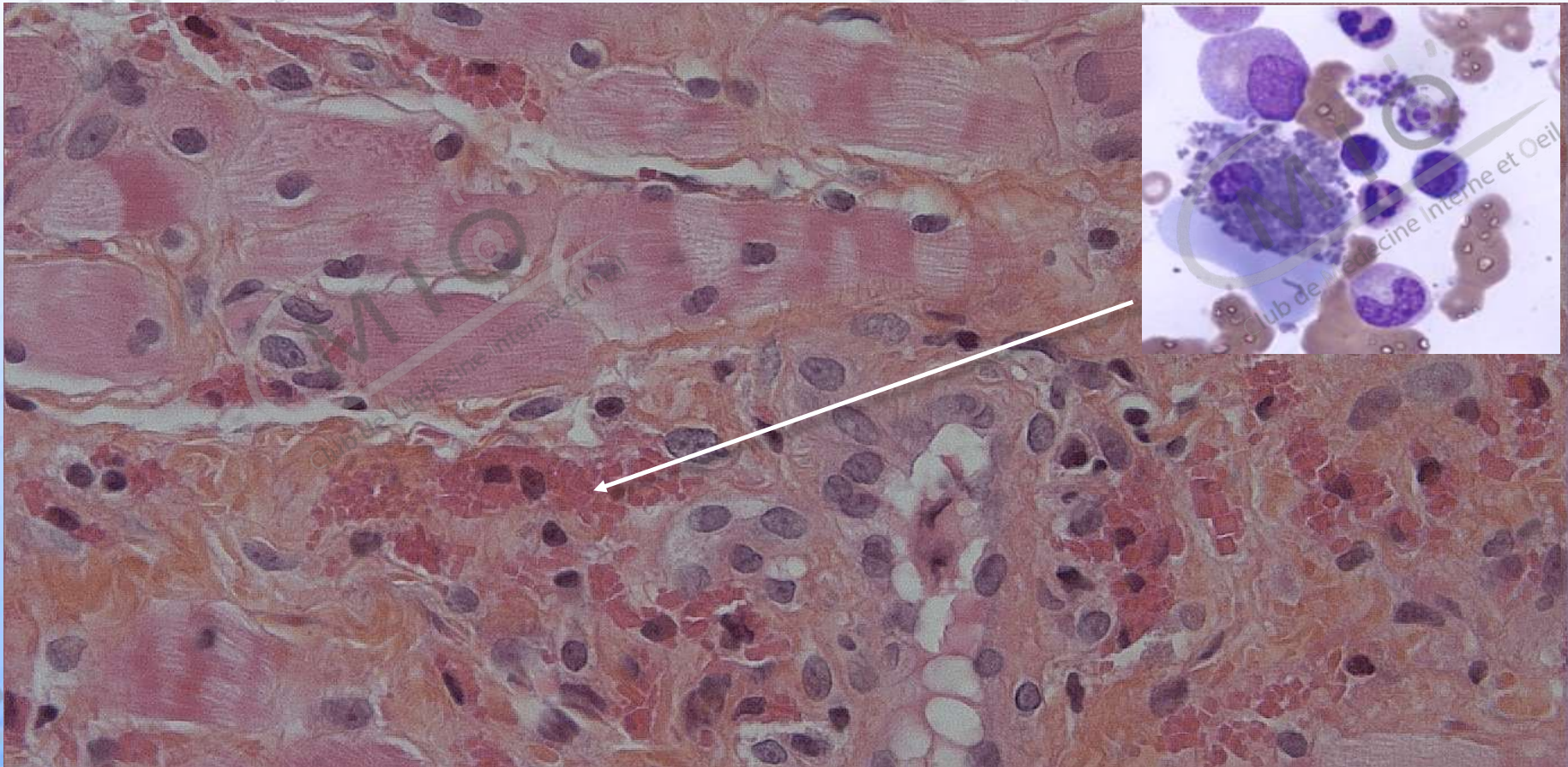
IgA



Kappa

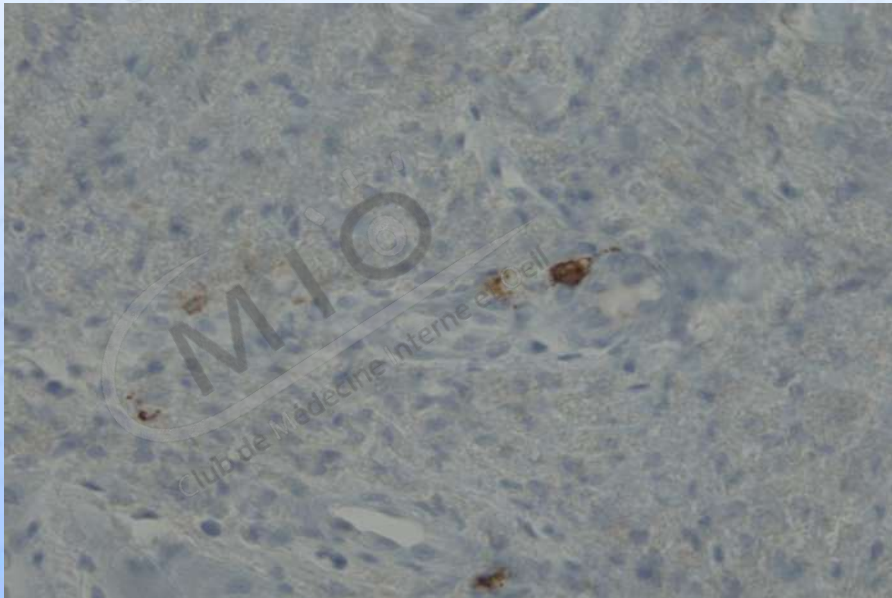


# Biopsie Orbite



# Biopsie Orbite

## CD138





## **Cas clinique n° 1**

- 1. Lymphome orbitaire**
- 2. Maladie associée aux IgG4**
- 3. Xanthogranulomatose nécrobiotique**
- 4. Sarcoïdose**
- 5. Histiocytose cristalline de surcharge**

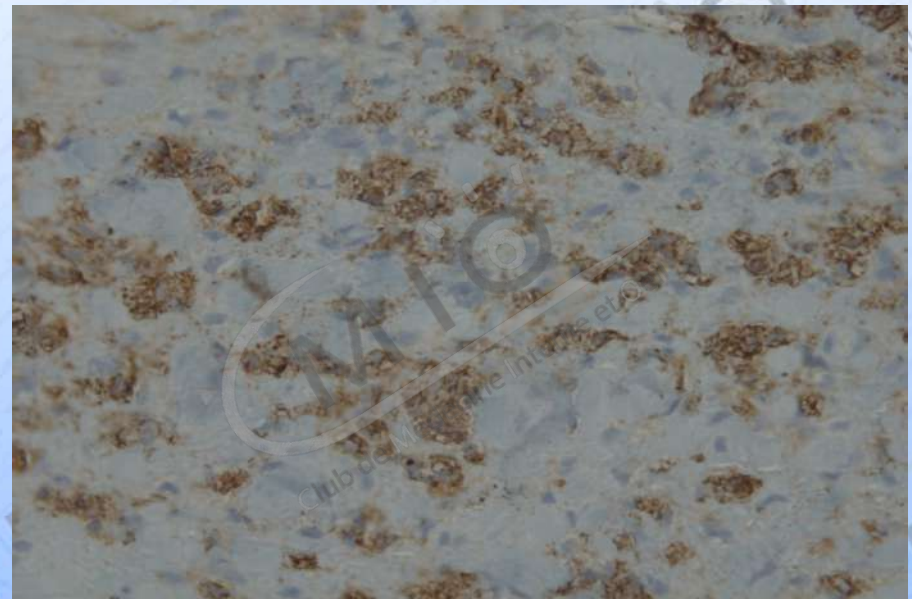
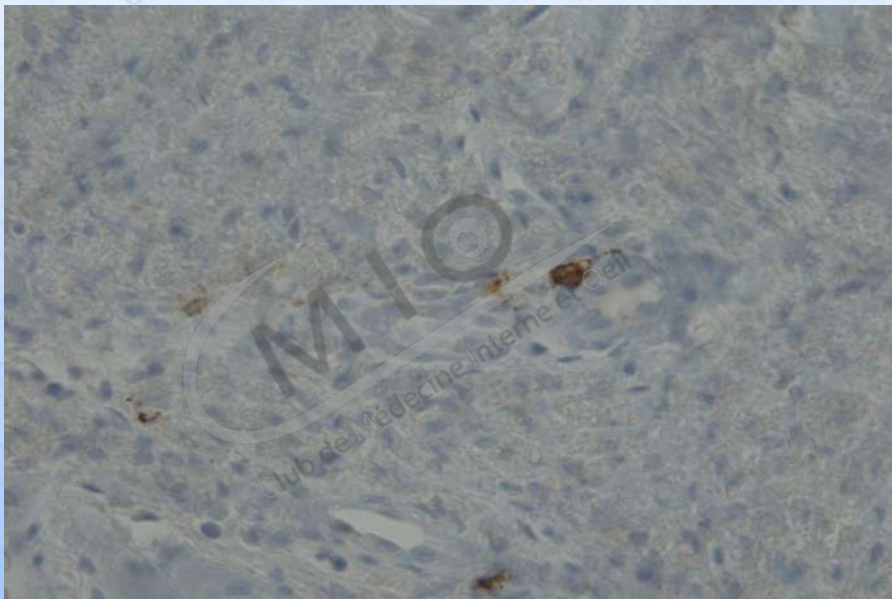
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# Biopsie Orbitale

**CD138**

**CD68**



## Crystal-Storing Histiocytosis: Report of a Case, Review of the Literature (80 Cases) and a Proposed Classification

Snjezana Dogan · Leon Barnes ·  
Wilhelmina P. Cruz-Vetrano



### Abstract

We report a case of crystal storing histiocytosis (CSH) of the upper lip and cheek in a 51-year-old woman and review the clinicopathologic features of 80 cases in the literature. These occurred in 41 men and 39 women with a respective mean age of 59 and 61 years (range 17-81 years). **Forty-six patients (58%) had localized CSH, and, of these, 16 (35%) occurred in the head and neck, with the most common site being the eye/orbit.** The remaining 34 patients (42%) had generalized CSH primarily involving bone marrow, liver, lymph nodes, spleen and/or kidney. Regardless of whether the CSH was localized or generalized, the vast majority of patients (90%) had an underlying lymphoproliferative or plasma cell disorder, especially multiple myeloma, lymphoplasmacytic lymphoma, or monoclonal gammopathy of undetermined significance. In 7 cases (8.8%), the CSH was associated with a variety of benign disorders, often with an inflammatory background, and no evidence of a clonal lymphoproliferative or plasma cell disorder. Treatment and prognosis varied according to the underlying disease. A classification of CSH based on etiology and/or associated disease and chemical composition of the crystal is proposed, rare non-immunoglobulin variants of CSH are discussed, and a differential diagnosis of other potentially confusing lesions is provided.

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# Evolution

2012

**Cristal storing histiocytosis**



2014  
(Février → Avril)

**VTD (4 cures)**  
*Velcade*  
*Thalidomide*  
*Dexaméthasone*

2014  
Mai

**Autogreffe de moëlle osseuse**

2014  
Aout → Novembre

**VTD (3 cures)**

- Pic IgA Kappa: 9 gr/l
- Plasmocytose médullaire 10%

- Pic IgA kappa: 9 gr/l
- TAP : HSM +++
- Gglions rétropéritonéaux
- Infiltration graisse péritonéale

- TAP : HSM (20, 13 cm)

- Pic IgA Kappa ⊙
- TAP : HSM (17, 11 cm)

**Paupières normales!**

Cet intervenant ...

- n'a pas déclaré de lien d'intérêt

Tous les orateurs ont reçu une déclaration de liens d'intérêt