III JORNADA CLÍNICA

del Grupo de Estudio Latinoamericano de Linfoproliferativos (GELL)

COMITÉ ORGANIZADOR: JUNTA DIRECTIVA GELL 2022 -2024

CASO CLINICO: LINFOMA MALT Y DESORDEN IgG4

PRESENTACION DE CASO:

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Médico Asistente Oncología Médica HNERM
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DISCUSION DE CASO:

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Instituto de Previsión Social. Hematóloga clínica
en Centro Oncológico REVITA. Asunción, Paraguay.

COI

- No conflicto de intereses para esta presentación

¿Por qué necesitamos hablar de IgG4?

¿Por qué necesitamos hablar de IgG4?

haematologica
Open access journal of the Ferrata-Storti Foundation, a non-profit organization

Vol. 104 No. 3 (2019): March, 2019 > IgG4-related disease: what a hematologist needs to know

REVIEW ARTICLES

IgG4-related disease: what a hematologist needs to know

Luke Y.C. Chen, Andre Mattman, Michael A. Seidman, Mollie N. Carruthers

Vol. 104 No. 3 (2019): March, 2019 https://doi.org/10.3324/haematol.2018.205526

¿Por qué necesitamos hablar de IgG4?

BMC Part of Springer Nature

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The risk of malignancy in patients with IgG4-related disease: a systematic review and meta-analysis

Tingfeng Yu, Yaxian Wu, Jia Liu, Yanyan Zhuang, Xiaoyan Jin & Lingyun Wang

Arthritis Research & Therapy **24**, Article number: 14 (2022) | Cite this article

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A Correspondence to this article was published on 24 May 2022

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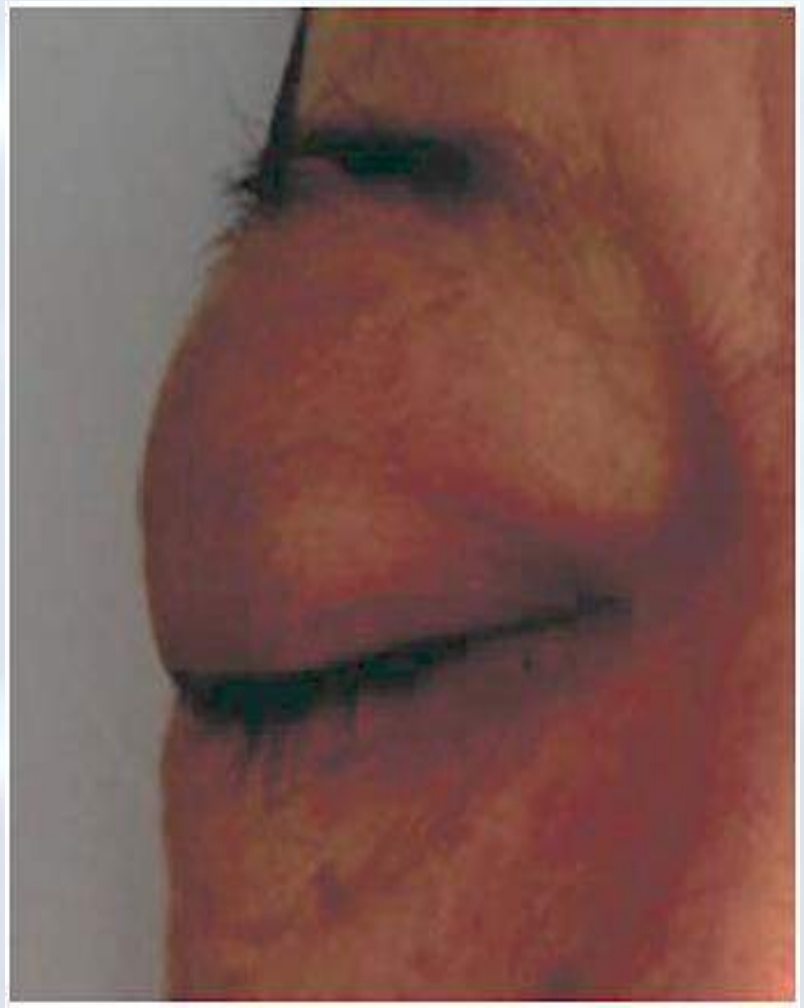
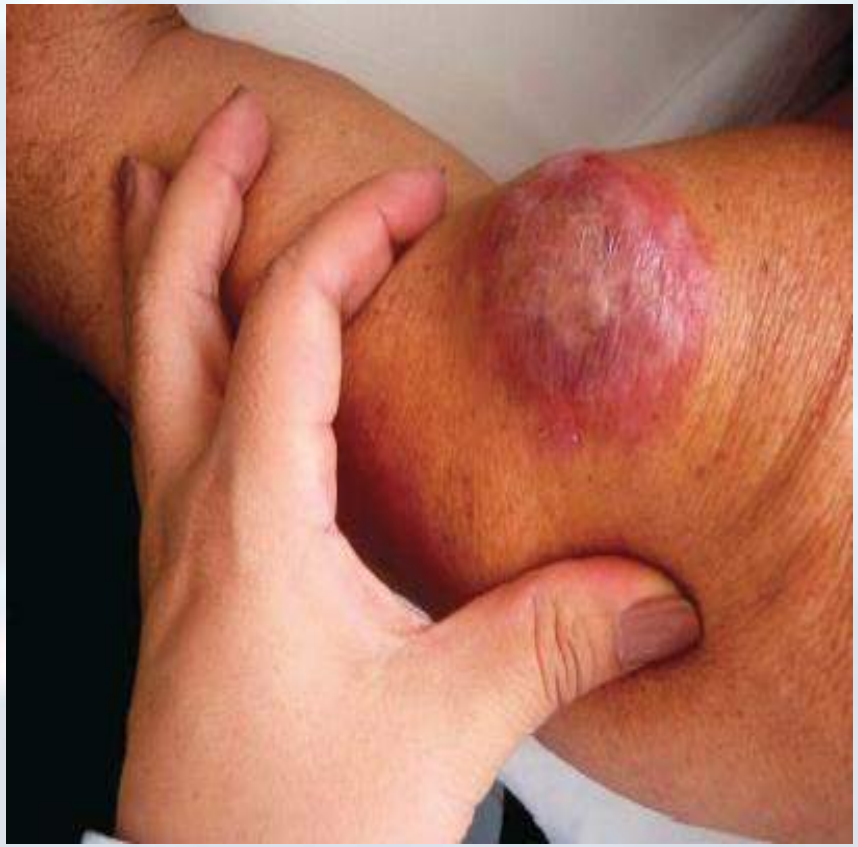
CASO CLINICO

- Pte varón de 58 años con antecedente de Malaria en 3 oportunidades.
- 2009: exoftalmos bilateral asintomático
- 2010: Biopsia de tejido (-) NM



CASO CLINICO

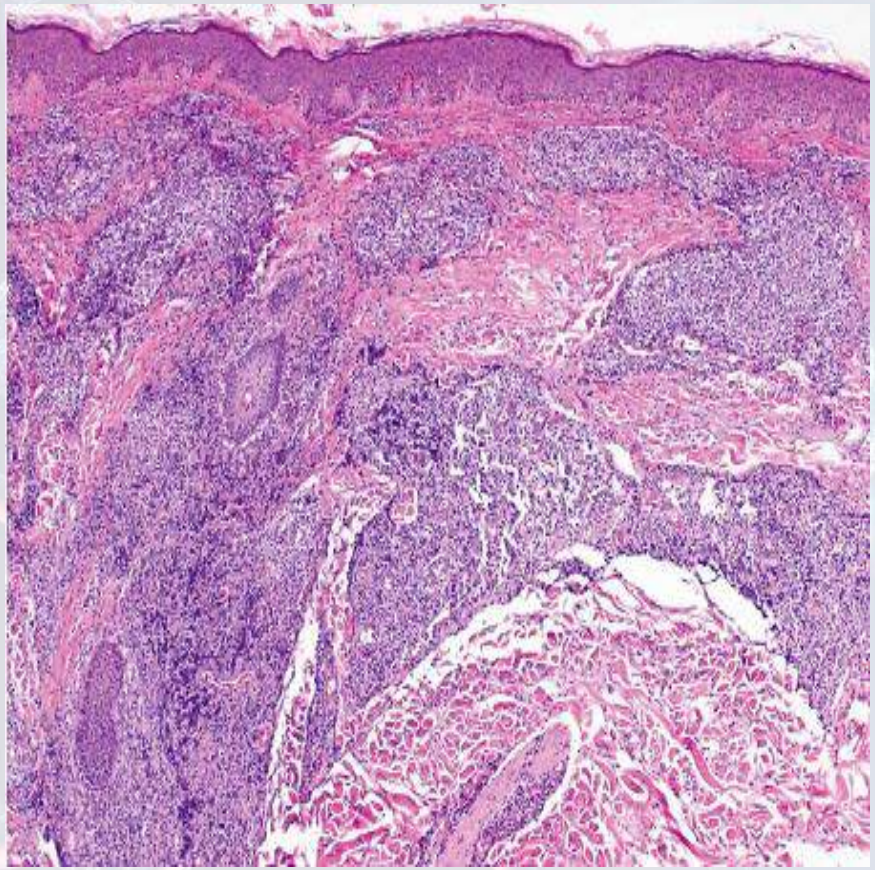
- 2016: Nódulos subcutáneos en partes blandas.
- ECOG 1.
- Múltiples lesiones eritematosas, nodulares e infiltradas en región frontoparietal izquierda y parietal derecha, parte posterior del cuello y superior de la espalda. Tumor exofítico eritematoso de 5 cm en cara interna de brazo izquierdo.

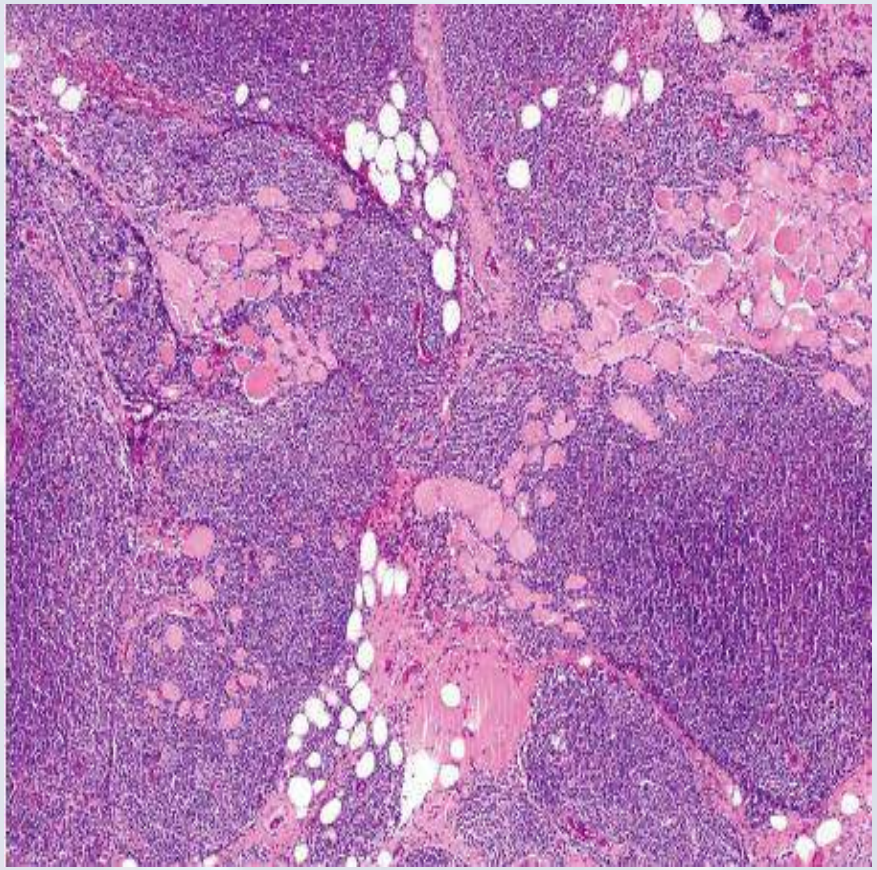


CASO CLINICO

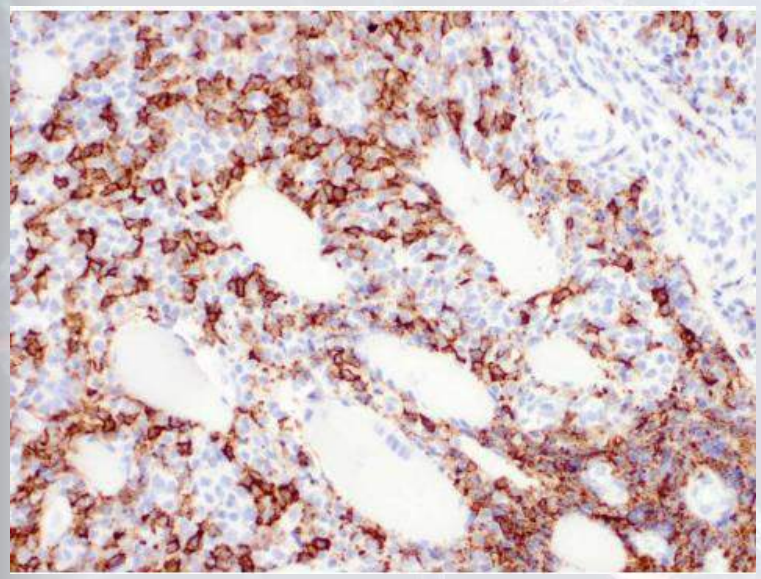
- Lab: HB 15.7 g/dL, Leuc: 5.6 x 10^9/L, RAN 2.9 x 10^9/L, no eosinofilia, plaq rangos normales.
- LDH, B2-MG, electroforesis en rangos normales. No VIH, no VHB, no VHC. Serologia C. psittaci, Chlamydia trachomatis, y Chlamydia pneumoniae negativos.
- IgE: 595 IU/mL (0–99 IU/mL).
- Aspirado medula ósea médula normocelular con hematopoiesis en las 3 líneas.
- TEM CU-TAP CC: compromiso de nódulos subcutaneoa múltiples y adp cervicales bilaterales.

Biopsia del tejido orbitario

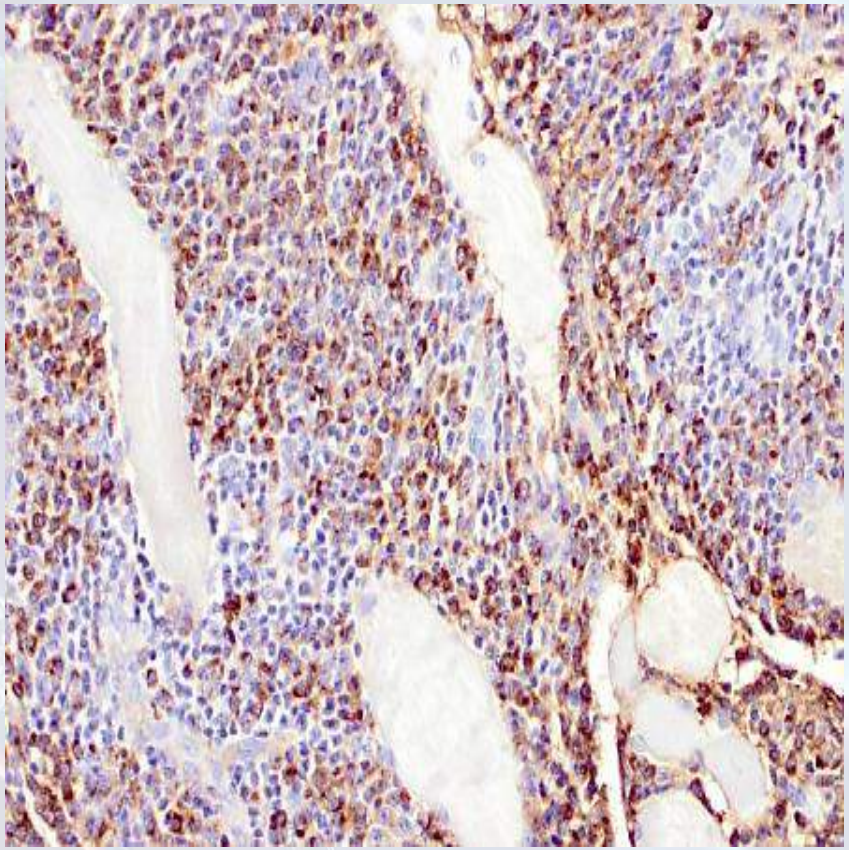




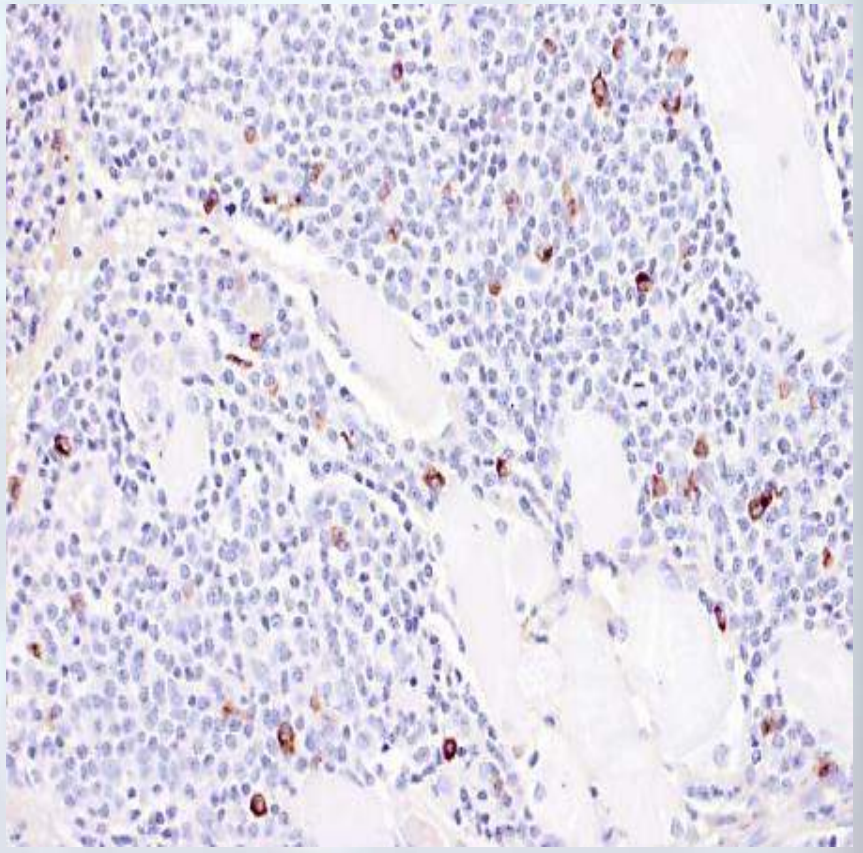
Denso infiltrado superficial plasmocitario y profundo perivascular y perianexial, con patrón nodular



CD 20 +, infiltrando músculo y tejidos blandos



Cadena Kappa



Cadena Lambda

CASO CLINICO

- **DX: Linfoma de la zona marginal con diferenciación plasmocítica 1° anexo ocular EC IVB x comp. Partes blandas (2016).**

Qué entidades pueden tener un patrón de Ig E elevada, infiltrados plasmocitarios y linfadenopatías?

Qué características son de interés en este caso?

Qué entidades pueden tener un patrón de Ig E elevada, infiltrados plasmocitarios y linfadenopatías?

Mimicker of IgG4-related disease	Areas of overlap	Distinguishing features of the mimicker not typically seen in IgG4-related disease
Multicentric Castleman disease (MCD)	• Lymphadenopathy (particularly the MCD-like variant of IgG4-LAD) • High serum IgG4 • IgG4+ plasma cells in tissue* • Polyclonal hypergammaglobulinemia	• MCD is a "hyper-IL-6" syndrome associated with B symptoms and highly elevated CRP and IL-6 not seen in IgG4-RD
Cutaneous and systemic plasmacytosis (CSP)	• Lymphadenopathy • Polyclonal hypergammaglobulinemia • Polyclonal plasmacytosis, including IgG4+ plasma cells in bone marrow and skin*	• IgG4-RD rarely involves skin whereas cutaneous lesions (round/oval, red/brown poorly circumscribed macules, papules and plaques) are an obligatory feature of CSP; serum IgG4 in CSP is normal or mildly elevated
Rosai-Dorfman-Destombes disease (RDD, a.k.a "Sinus Histiocytosis with Massive Lymphadenopathy; "R" group histiocytosis)	• Lymphadenopathy • Extranodal RDD can involve the nasal cavity, and retro-orbital tissue • Meningeal involvement of RDD may mimic IgG4-pachymeningitis • Salivary gland involvement • CNS involvement including pachymeningitis • Increased IgG4+ plasma cells in tissue*	• Massive cervical lymphadenopathy is atypical for IgG4-RD • RDD may present with B symptoms • Large histiocytic cells with hypochromatic nuclei and pale cytoplasm; emperipolesis; positive for S100, CD68, CD14 and CD163

Haematologica 2019 Volume 104(3):444-455

Qué entidades pueden tener un patrón de Ig E elevada, infiltrados plasmocitarios y linfadenopatías?

Mimicker of IgG4-related disease	Areas of overlap	Distinguishing features of the mimicker not typically seen in IgG4-related disease
Hypereosinophilic syndrome (HES)/chronic eosinophilic leukemia (CEL) [particularly lymphocyte-variant HES]	• T-cell clonality by PCR • Atopy/asthma/elevated IgE • Lymphadenopathy • Eosinophilia • Elevated serum IgG4 • Aberrant T cell phenotype in peripheral blood ($CD4^+/3^-$, $CD4^+/7^-$, $CD3^+/4^-/8^-$)	• Increased blasts or myeloid clone in CEL • PDGFR-alpha/PDGFR-beta/FGFR1/PCM1-JAK2 positivity are not seen in IgG4-RD • More marked and persistent eosinophilia in HES
Plasma cell myeloma/monoclonal gammopathy of undetermined significance	• Plasma cell infiltrate • Hypergammaglobulinemia • Renal failure • Proteinuria	• Plasma cell clonality • Monoclonal paraprotein ± suppression of polyclonal immunoglobulins • Lytic boney disease/hypercalcemia • Light chains in urine rather than albuminuria
Vasculitis, particularly eosinophilic granulomatosis with polyangiitis and granulomatosis with polyangiitis	• Eosinophilia in blood and tissue • Polyclonal hypergammaglobulinemia with elevated serum IgG4 • Peri-aortitis and rarely, Kawasaki-like coronary	• Highly elevated CRP in vasculitis • Extravascular granulomas, small-medium vessel vasculitis • Mononeuritis multiplex not a feature of IgG4-RD

Haematologica 2019 Volume 104(3):444-455

Qué entidades pueden tener un patrón de Ig E elevada, infiltrados plasmocitarios y linfadenopatías?

Mimicker of IgG4-related disease	Areas of overlap	Distinguishing features of the mimicker not typically seen in IgG4-related disease
Erdheim Chester disease (ECD; "L" group histiocytosis)	• Retroperitoneal fibrosis • Central nervous system/hypophysitis • Pulmonary involvement	• >95% of ECD patients have bone involvement, which is rare in IgG4-RD (apart from rare IgG4$^+$ angiocentric eosinophilic fibrosis of the head and neck) • Yellow peri-orbital xanthelasmas common in ECD (skin involvement in IgG4-RD is rare, and when present tends to be erythematous or flesh-colored papules) • Foamy multi-nucleated histiocytes, few Touton cells, fibrosis; CD68$^+$, CD163$^+$, CD1a$^-$ • 50% of ECD patients are BRAF V600E-positive
Malignant lymphoma	• Lymphadenopathy • Extranodal mass lesions • Blood and tissue eosinophilia	• B symptoms, bone involvement, brain parenchymal involvement and hypercalcemia are rare in IgG4-RD
Sarcoidosis	• Lymphadenopathy • Pulmonary nodules • Pachymeningitis and/or hypophysitis • Polyclonal hypergammaglobulinemia • Multi-organ involvement	• Non-caseating granulomas • Hypercalcemia and elevated ACE levels

Haematologica 2019 Volume 104(3):444-455

Qué sospecha clínica tener en compromiso ocular?

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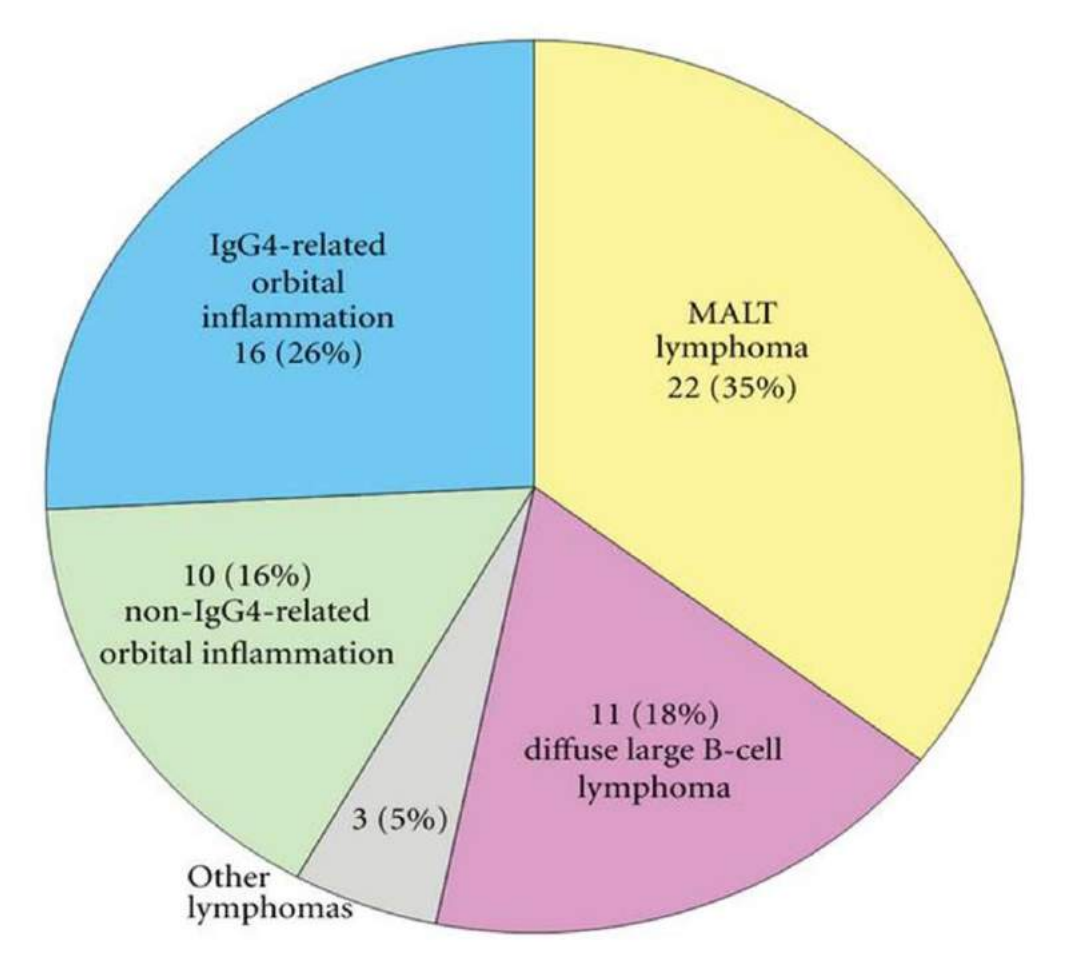


Takahira M et al 2012 Inter J Rheumatol

Qué sospecha clínica tener en compromiso ocular?

Ocular Adnexal Lymphoma Associated With IgG4+ Chronic Sclerosing Dacryoadenitis: A Previously Undescribed Complication of IgG4-related Sclerosing Disease

Wah Cheuk, MBBS, Hunter K. L. Yuen, MBChB,† Alexander C. L. Chan, MBBS,* Lee-Yung Shih, MD,‡ Tseng-Tong Kuo, MD,§ Ming-Wai Ma, MBBS,‖ Yan-Fai Lo, MBChB,¶ Wai-Kong Chan, MBChir,# and John K. C. Chan, MBBS**

F/69	3 yr ago: right IgG4-related dacryoadenitis	Follicular lymphoma
M/69	Bilateral proptosis, 1 yr ago – NP↑IgG4+ cells left orbital tumor	MALT lymphoma
M/60	1 yr ago: right IgG4-related dacryoadenitis	MALT lymphoma

Qué sospecha clínica tener en compromiso ocular?

Pathology International 2008; **58**: 465–470 doi:10.1111/j.1440-1827.2008.02257.x

Original Article

Ocular adnexal IgG4-related disease has uniform clinicopathology

Yasuharu Sato,[1] Koh-ichi Ohshima,[2] Kouichi Ichimura,[1] Masakazu Sato,[3] Ichiro Yamadori,[3] Takehiro Tanaka,[1] Katsuyoshi Takata,[1] Toshiaki Morito,[1] Eisaku Kondo[1] and Tadashi Yoshino[1]

Pathology International 2010; **60**: 247–258 doi:10.1111/j.1440-1827.2010.02524.x

Review Article

IgG4-related disease: Historical overview and pathology of hematological disorders

Yasuharu Sato,[1,*] Kenji Notohara,[2,*] Masaru Kojima,[3,*] Katsuyoshi Takata,[1] Yasufumi Masaki[4] and Tadashi Yoshino[1]

7 cases of MALT lymphoma arising in ocular-adnexal IgG4-RD,concomitant or metachronous?

Cuáles son las diferentes presentaciónes clínicas relacionadas en IgG4??

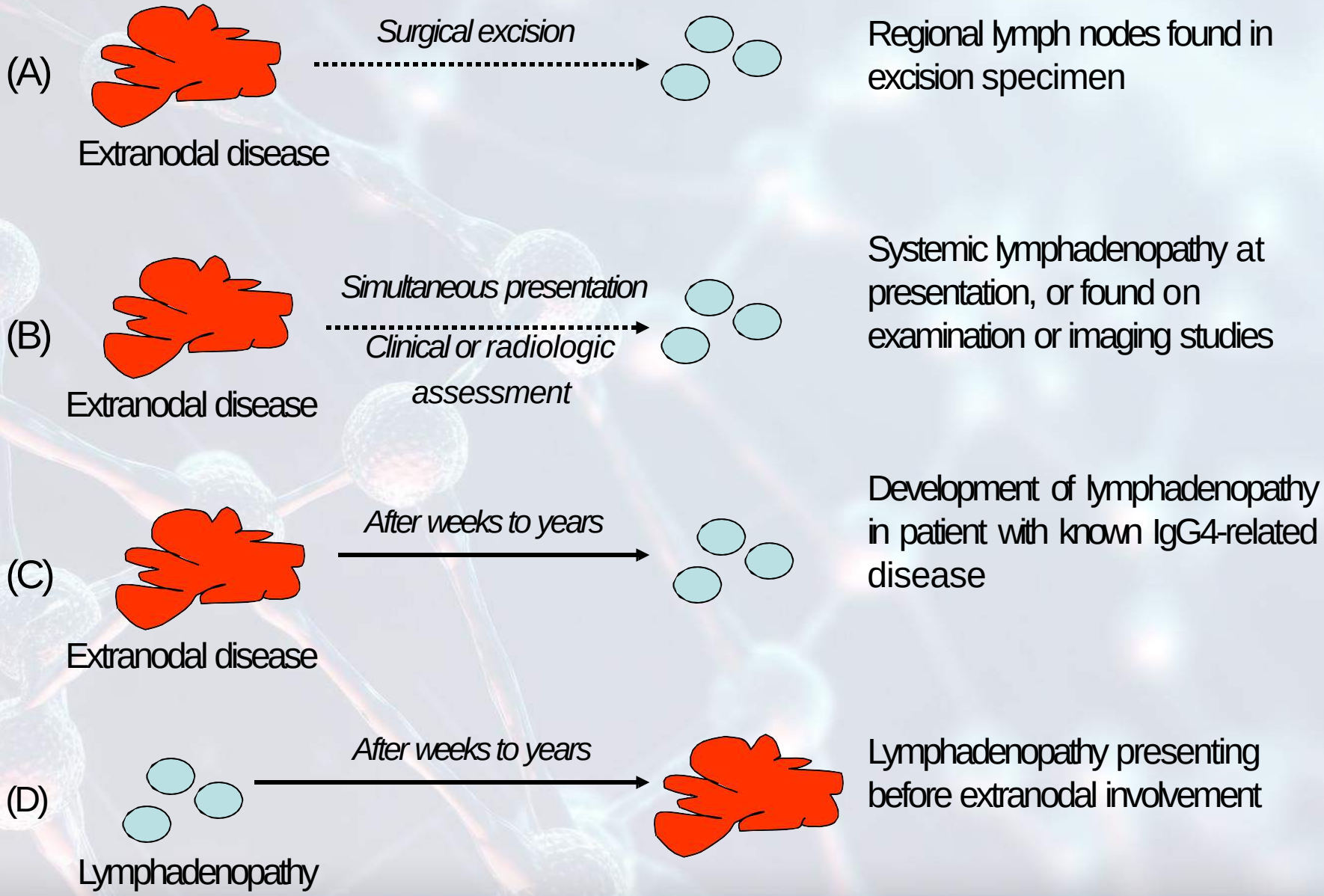
TABLE 1. Cases of MALT Extranodal MZL Involving the Ocular Adnexae and Subcutaneous Tissue Reported in the Literature

Case	Author	Sex	Age	Primary	Initial Treatment	Method Detection
1	Jonak et al[2]	F	58	Orbit	Radiotherapy	CT
2	Jonak et al[2]	F	61	Orbit	Radiotherapy	CT
3	Jonak et al[2]	F	64	Conjunctiva	Radiotherapy	CT
4	Jonak et al[2]	F	49	Orbit	Radiotherapy	PET-CT
5	Jonak et al[2]	F	77	Lacrimal gland	Radiotherapy	PET-CT
6	Jonak et al[2]	F	71	Orbit	CHOP	Self-assessment
7	Jonak et al[2]	F	84	Orbit, cervical node	Radiotherapy, MCP	CT
8	Jonak et al[2]	M	80	Lacrimal gland	Radiotherapy	Self-assessment
9	Jonak et al[2]	M	85	Conjunctiva	Radiotherapy	CT
10	Vincenti et al[3]	M	51	Orbit, mediastinal, abdominal LAD, bone marrow	Pegylated Interferon, ribavirin	Self-assessment, PET-CT
11	Beltran et al[13]	M	58	Orbit, cervical node	R-CHOP	CT, self-assessment

Case	Subcutaneous Involvement	Subcutaneous Localization	Treatment	Follow-up	Survival (mo)
1	Metachronous	Gluteal	Bortezomib	Alive	204
2	Metachronous	Suprapubic	CHOP	Alive	244
3	Metachronous	Lower back	R-CHOP	Alive	195
4	Synchronous	Gluteal	Radiotherapy	Alive	8
5	Metachronous	Periumbilical, paravertebral, lower back	R-CHOP, watchful waiting	Dead (heart disease)	137
6	Metachronous	Forearm	R-CHOP	Alive	93
7	Metachronous	Occipital	Radiotherapy	Dead (heart disease)	26
8	Metachronous	Face	CHOP	Dead (heart disease)	11
9	Metachronous	Lower back	Watchful waiting	Dead (senility)	49
10	Synchronous	Neck, forearm, back, pectoral region, cheekbones	Pegylated Interferon, ribavirin	Alive	19
11	Synchronous	Arm, cervical, frontoparietal, parietal, upper back	R-CHOP	Alive	9

LAD, lymphadenopathy, PET-CT, positron emission tomography–computed tomography; R-CHOP, rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone.

Cuáles son las diferentes presentaciones clínicas relacionadas en IgG4??



Cuáles son las diferentes presentaciones clínicas relacionadas en IgG4??

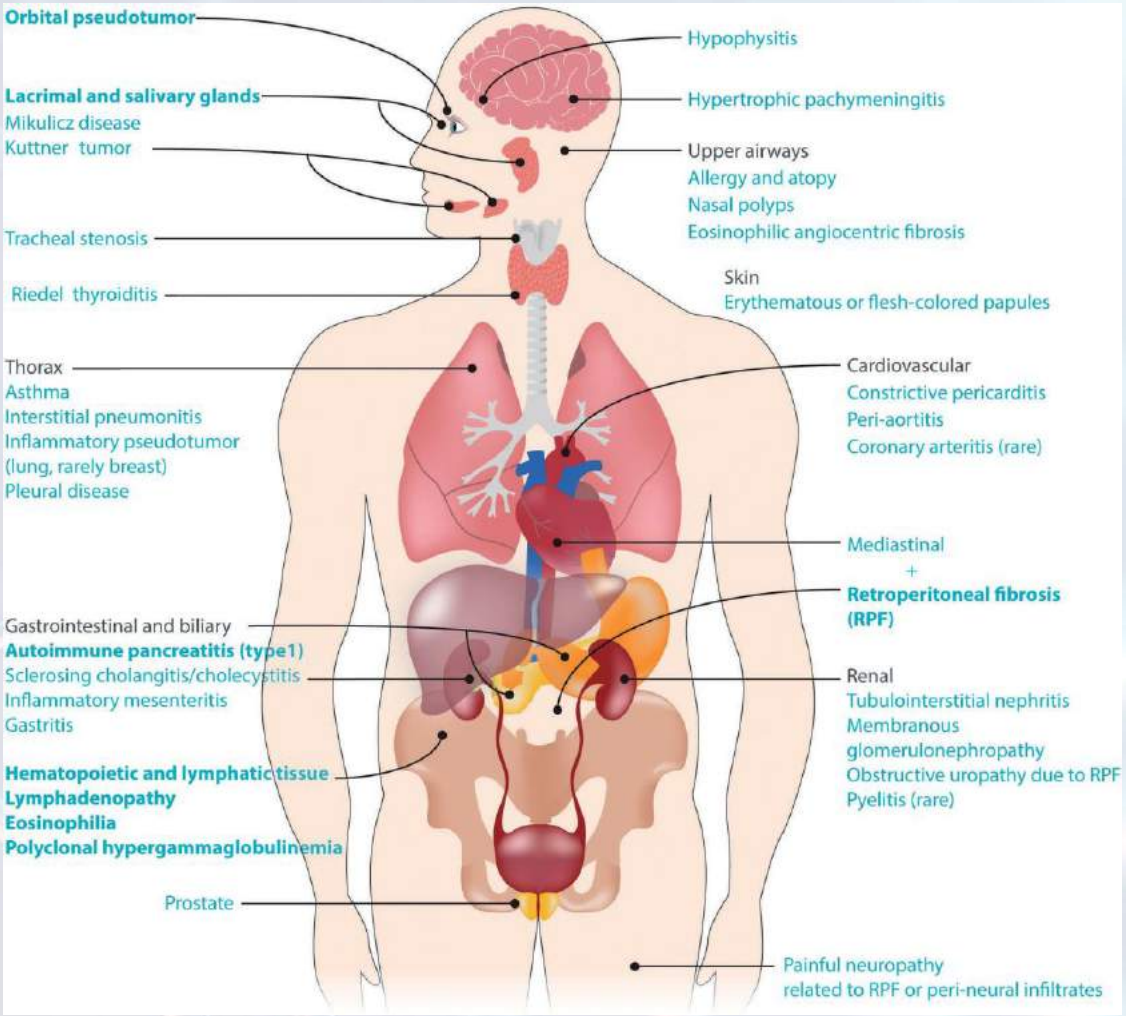
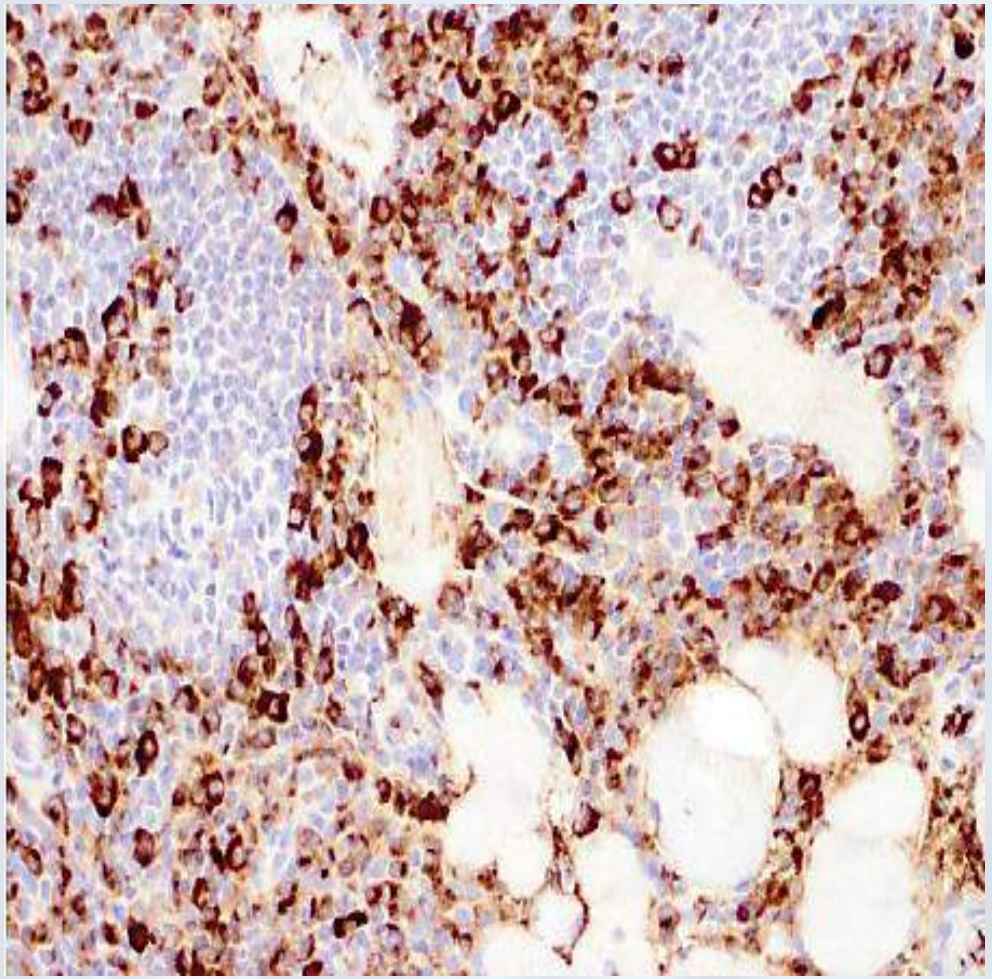


Figure 1. Manifestations of IgG4-related disease by organ system. The most common primary disease features are indicated in bold.

CASO CLINICO

- Se lleva el caso a comité multidisciplinario con Anatomía patológica y es solicitado IgG4 en IHQ.
- IgG4 sérico fue hallado normal

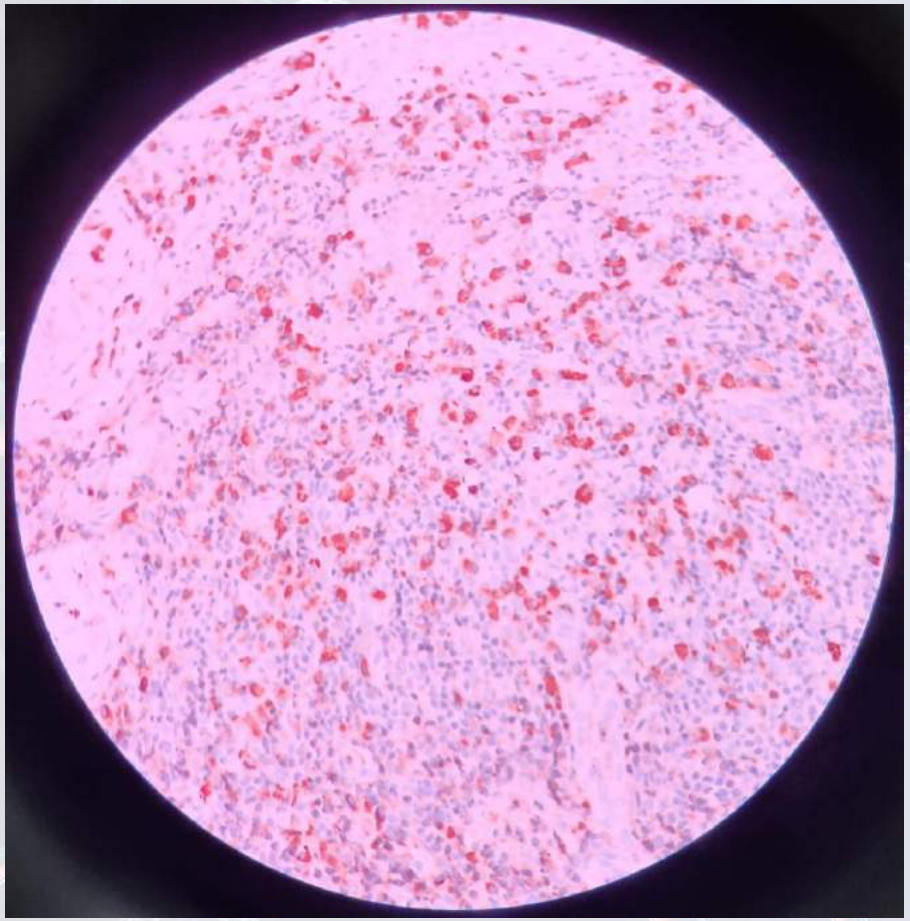


IgG4

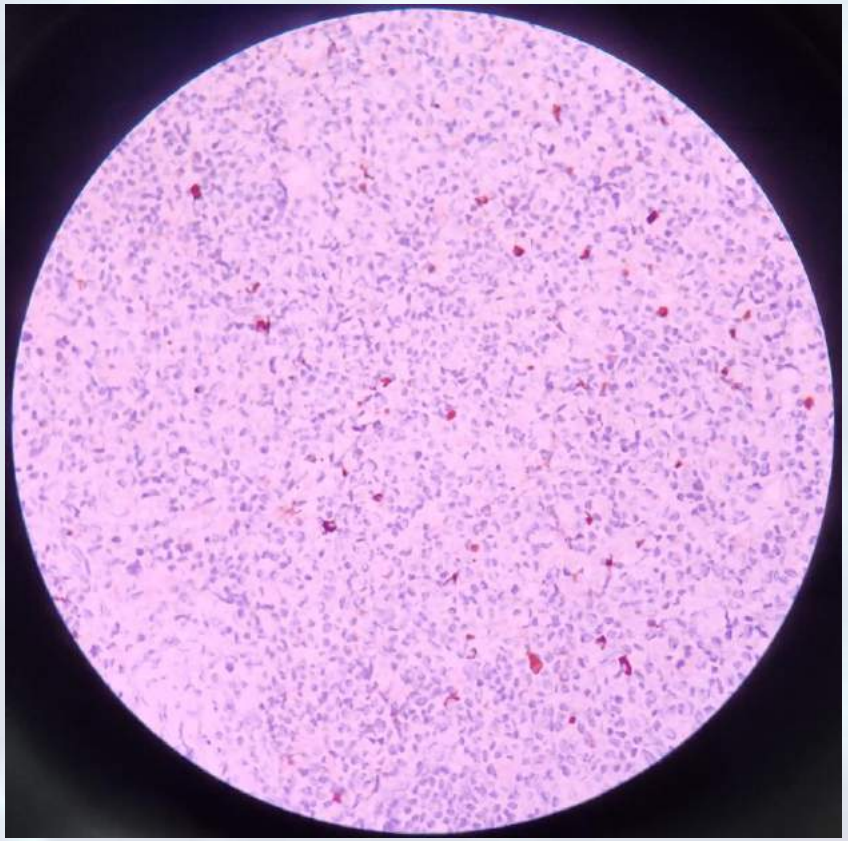
CASO CLINICO

- **DX: Linfoma de la zona marginal 1° anexo ocular EC IVB x comp. Partes blandas asociado a enfermedad IgG4 relacionada. (2016).**

NO TODO Ig G+ en IHQ, es Ig G4 +



IgG en tejido de glándula submaxilar



IgG4 en tejido de glándula submaxilar con ratio IgG4/IgG menos de 40%

CRITERIOS DIAGNÓSTICOS ENF. IGG4 RELACIONADA bajo OKAZAKI Y UMEHARA

Table 5: Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011 [15].

[Concept]

IgG4-related disease (IgG4-RD) shows organ enlargement or nodular/hyperplastic lesions in various organs concurrently or metachronously, due to marked infiltration of lymphocytes and IgG4-positive plasma cells, as well as fibrosis of unknown etiology. IgG4-RD affects various organs, including the pancreas, bile duct, lacrimal gland, salivary gland, central nervous system, thyroid, lung, liver, gastrointestinal tract, kidney, prostate, retroperitoneum, arteries, lymph nodes, skin, and breast. Although many patients with IgG4-RD have lesions in several organs, either synchronously or metachronously, others show involvement of a single organ. Clinical symptoms vary depending on the affected organ, and some patients may experience serious complications, such as obstruction or compression symptoms due to organomegaly or hypertrophy and organ dysfunction caused by cellular infiltration or fibrosis. Steroid therapy is often effective.

[Comprehensive clinical diagnostic criteria for IgG4-RD, 2011]

(1) Clinical examination shows characteristic diffuse/localized swelling or masses in single or multiple organs.

(2) Hematological examination shows elevated serum IgG4 concentrations (≥135 mg/dL).

(3) Histopathologic examination shows;

(1) marked lymphocyte and plasmacyte infiltration and fibrosis

(2) infiltration of IgG4-positive plasma cells: ratio of IgG4/IgG positive cells > 40% and > 10 IgG4-positive plasma cells/HPF.

Definite: (1) + (2) + (3), Probable: (1) + (3), Possible: (1) + (2)

However, it is important to differentiate IgG4-RD from malignant tumors of each organ (e.g. cancer, lymphoma) and similar diseases (e.g. Sjögren's syndrome, primary sclerosing cholangitis, Castleman's disease, secondary retroperitoneal fibrosis, Wegener's granulomatosis, sarcoidosis, and Churg-Strauss syndrome) by additional histopathological examination. Even when patients cannot be diagnosed using the CCD criteria, they may be diagnosed using organ-specific diagnostic criteria for IgG4RD.

Okazaki & Umehara, 2011

CRITERIOS HISTOLÓGICOS EN ENF. RELACIONADA A IGG4

MODERN PATHOLOGY (2012), 1–12

Consensus statement on the pathology of IgG4-related disease

Vikram Deshpande[1,31], Yoh Zen[2,31], John KC Chan[3], Eunhee E Yi[4], Yasuharu Sato[5], Tadashi Yoshino[5], Günter Klöppel[6], J Godfrey Heathcote[7], Arezou Khosroshahi[8], Judith A Ferry[1], Rob C Aalberse[9], Donald B Bloch[8], William R Brugge[10], Adrian C Bateman[11], Mollie N Carruthers[8], Suresh T Chari[12], Wah Cheuk[3], Lynn D Cornell[13], Carlos Fernandez-Del Castillo[14], David G Forcione[10], Daniel L Hamilos[15], Terumi Kamisawa[16], Satomi Kasashima[17], Shigeyuki Kawa[18], Mitsuhiro Kawano[19], Gregory Y Lauwers[1], Yasufumi Masaki[20], Yasuni Nakanuma[21], Kenji Notohara[22], Kazuichi Okazaki[23], Ji Kon Ryu[24], Takako Saeki[25], Dushyant V Sahani[26], Thomas C Smyrk[13], James R Stone[1], Masayuki Takahira[27], George J Webster[28], Motohisa Yamamoto[29], Giuseppe Zamboni[30], Hisanori Umehara[20] and John H Stone[8]

CRITERIOS HISTOLÓGICOS EN ENF. RELACIONADA A IGG4

All of the following criteria have to be satisfied:

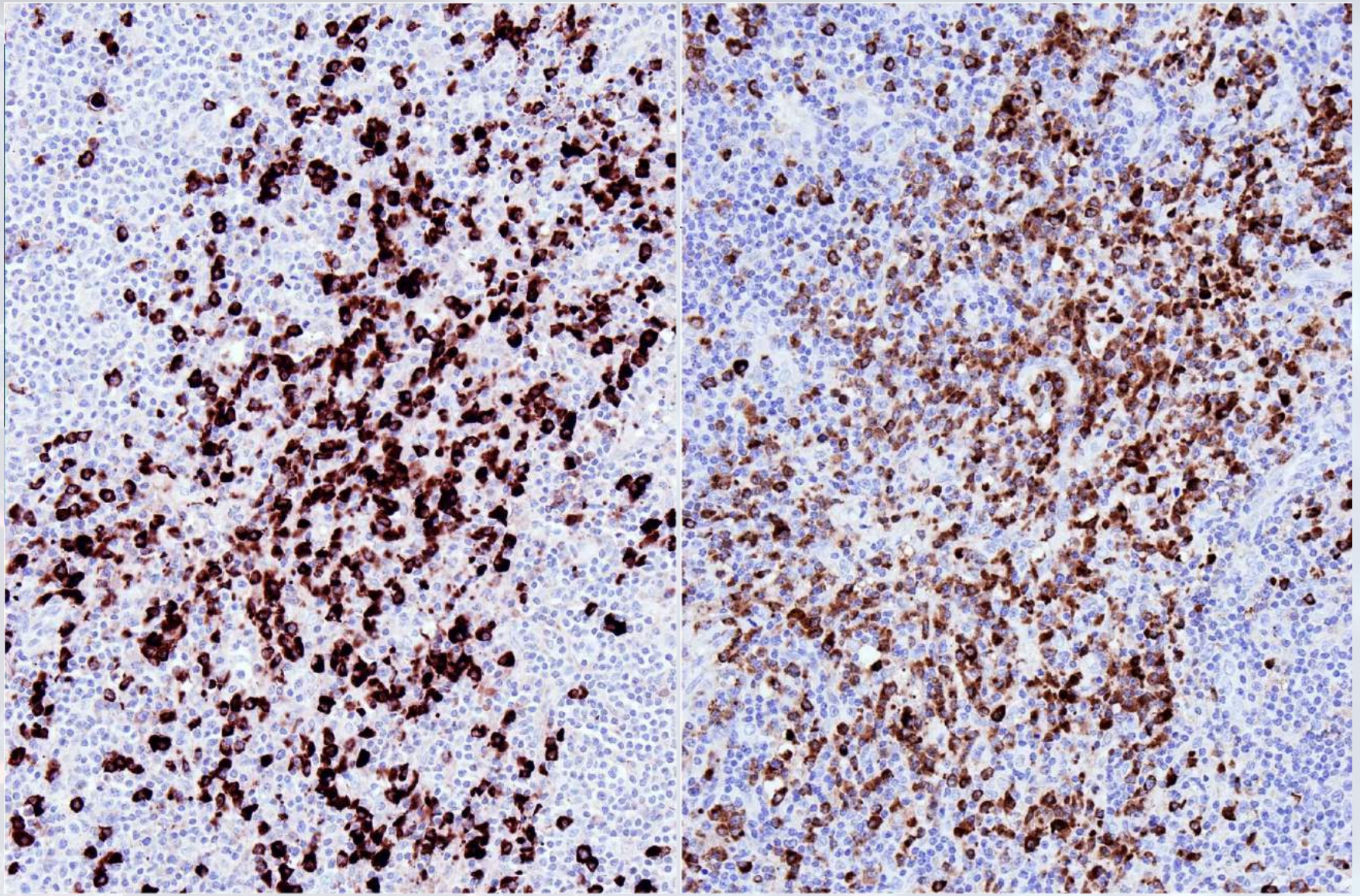
(1) Compatible morphology

(2) Absolute number of IgG4+ cells >10- 200/HPF (depending on anatomic location)

(3) Percentage of IgG4+/IgG+ cells >40%

Why essential?

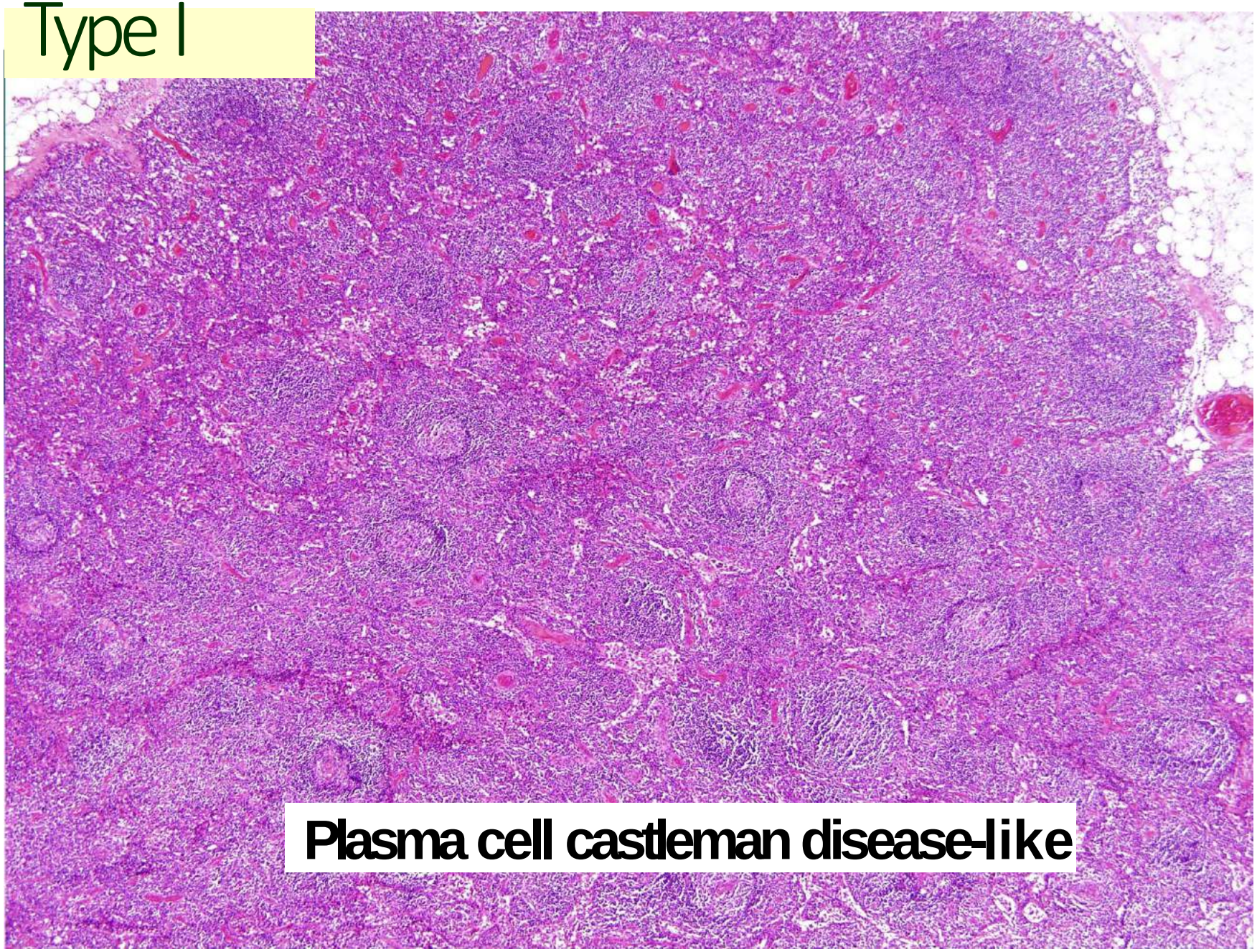
In any condition with significant plasmacytosis, absolute number of IgG4+ cells will be increased



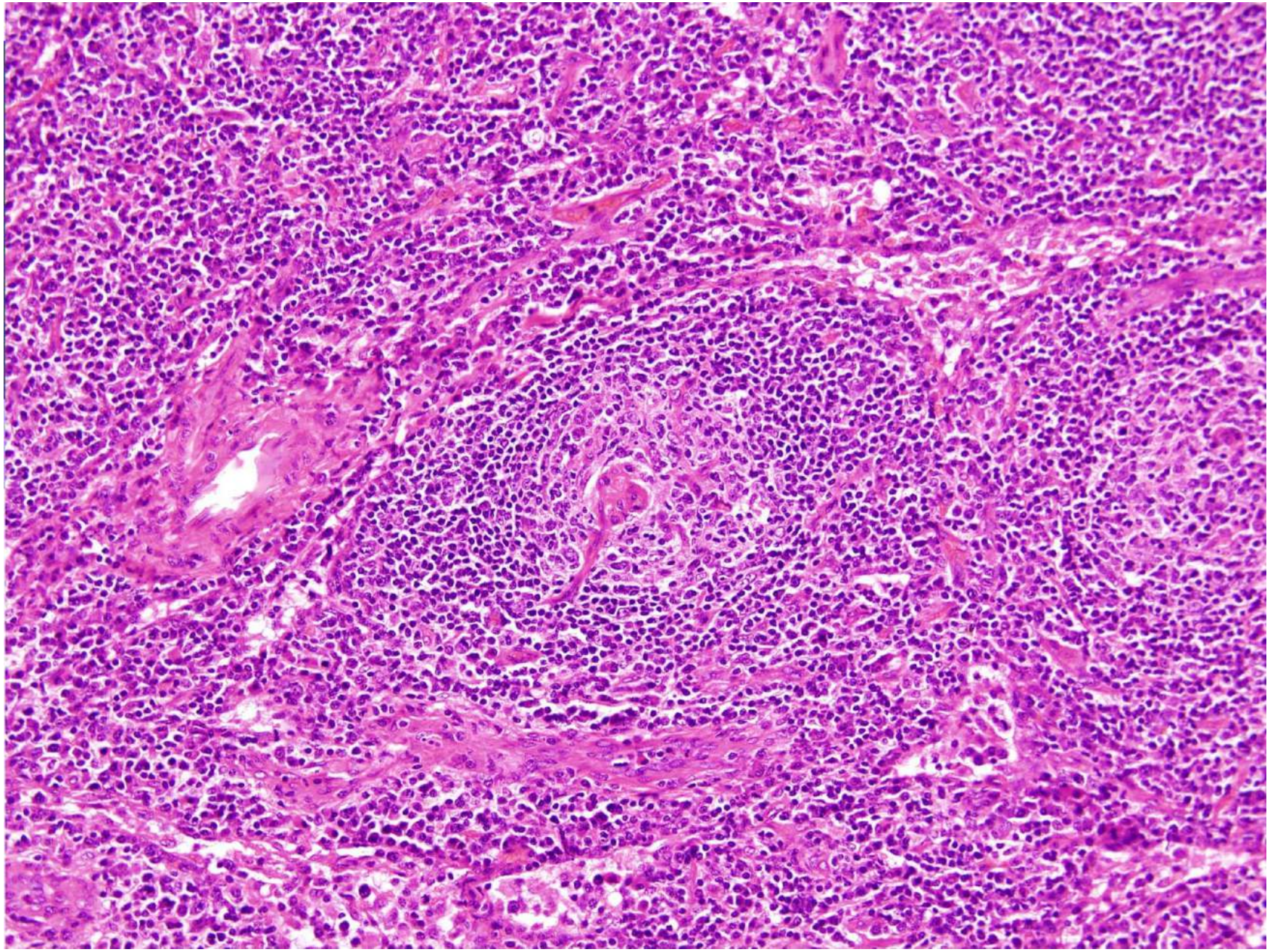
IgG4

IgG

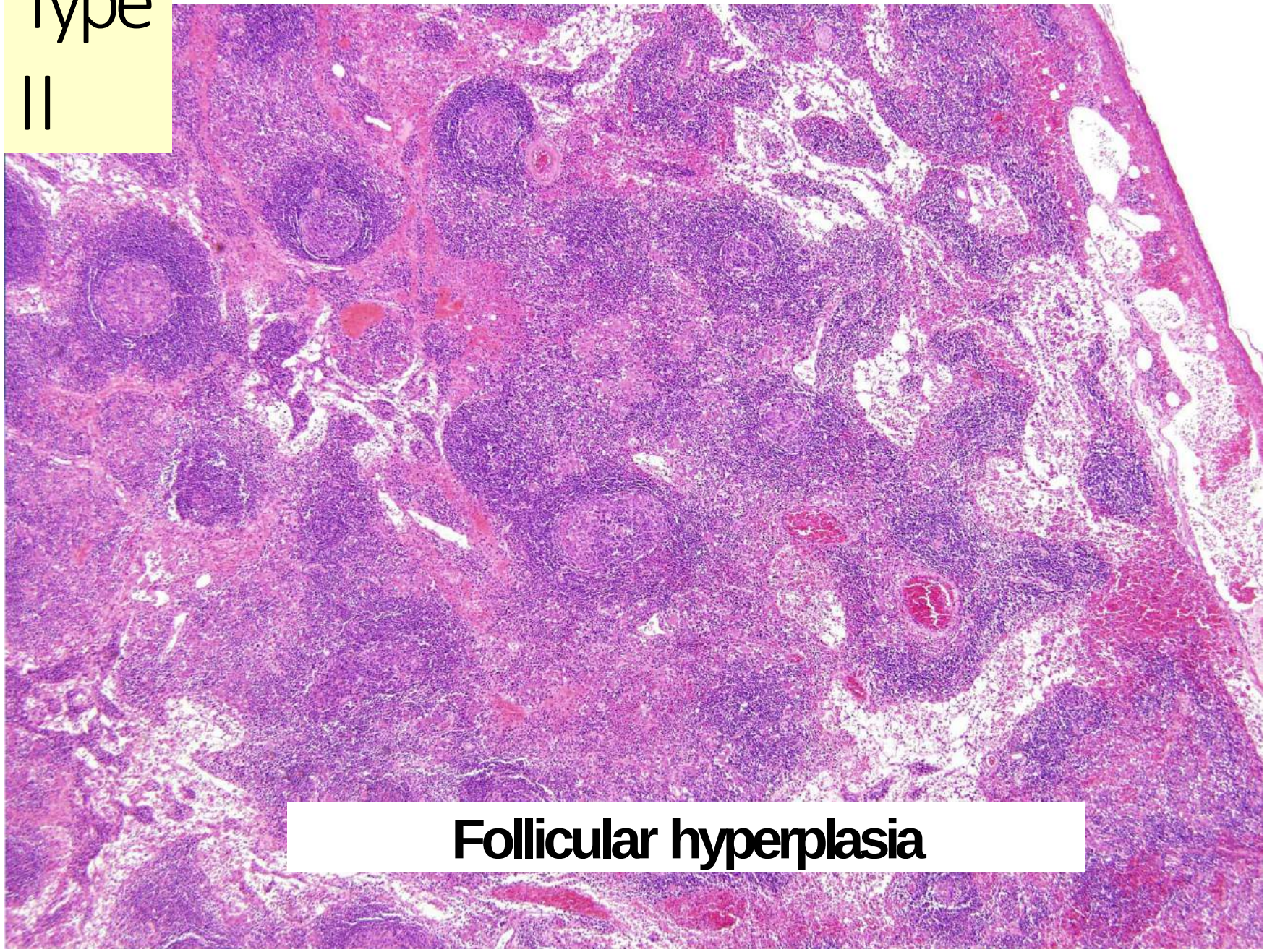
Type I



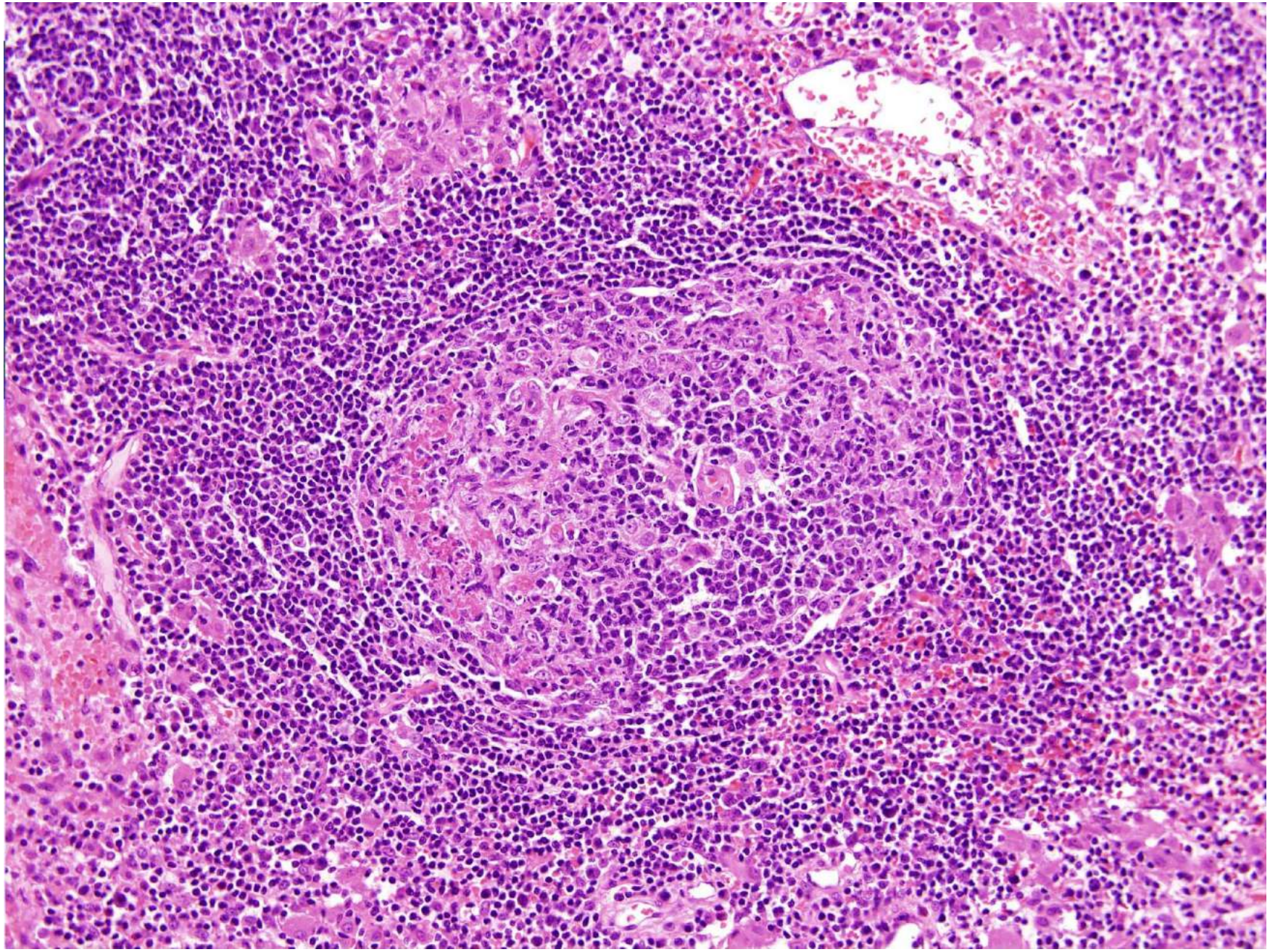
Plasma cell castleman disease-like



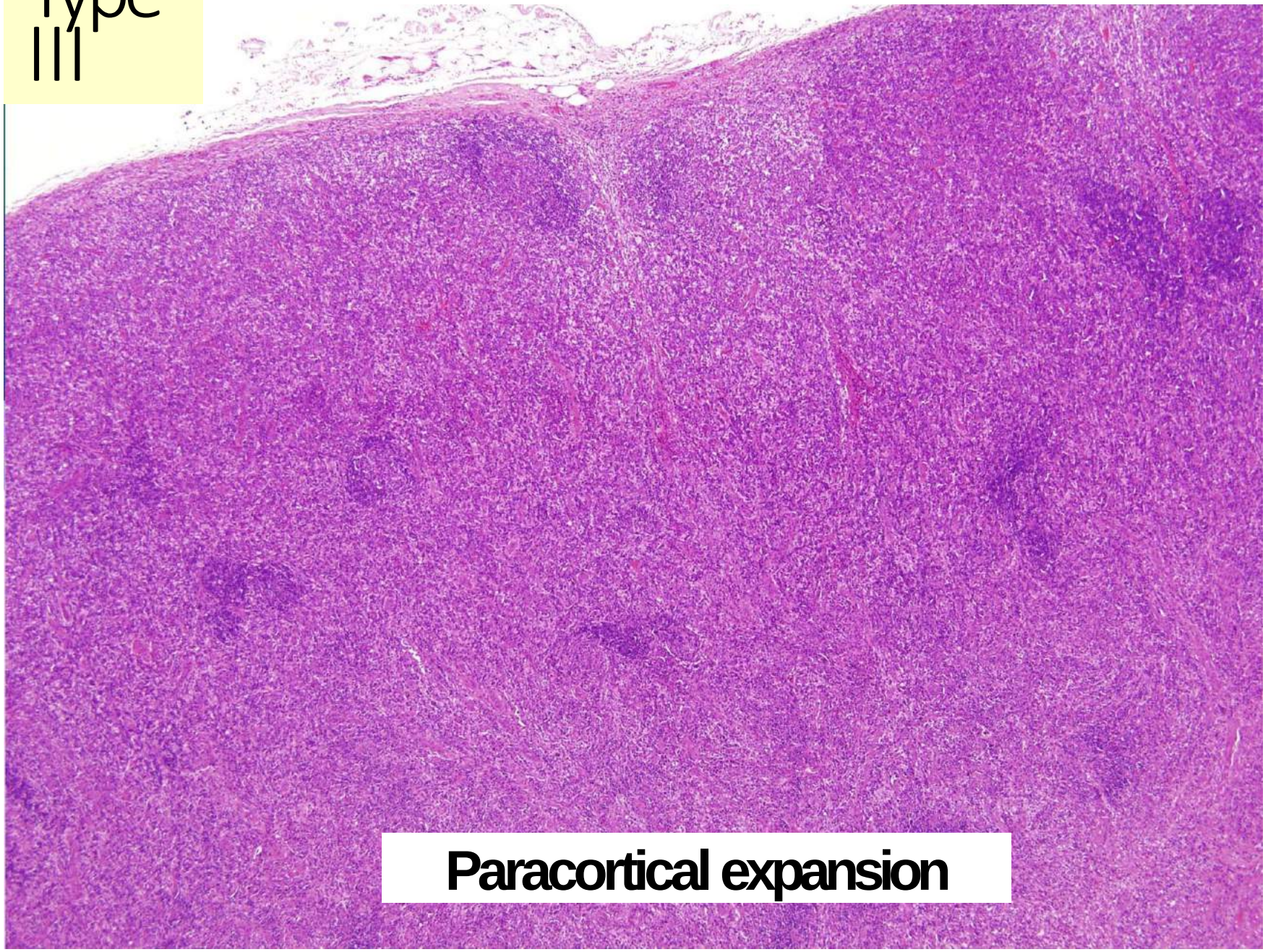
Type II



Follicular hyperplasia

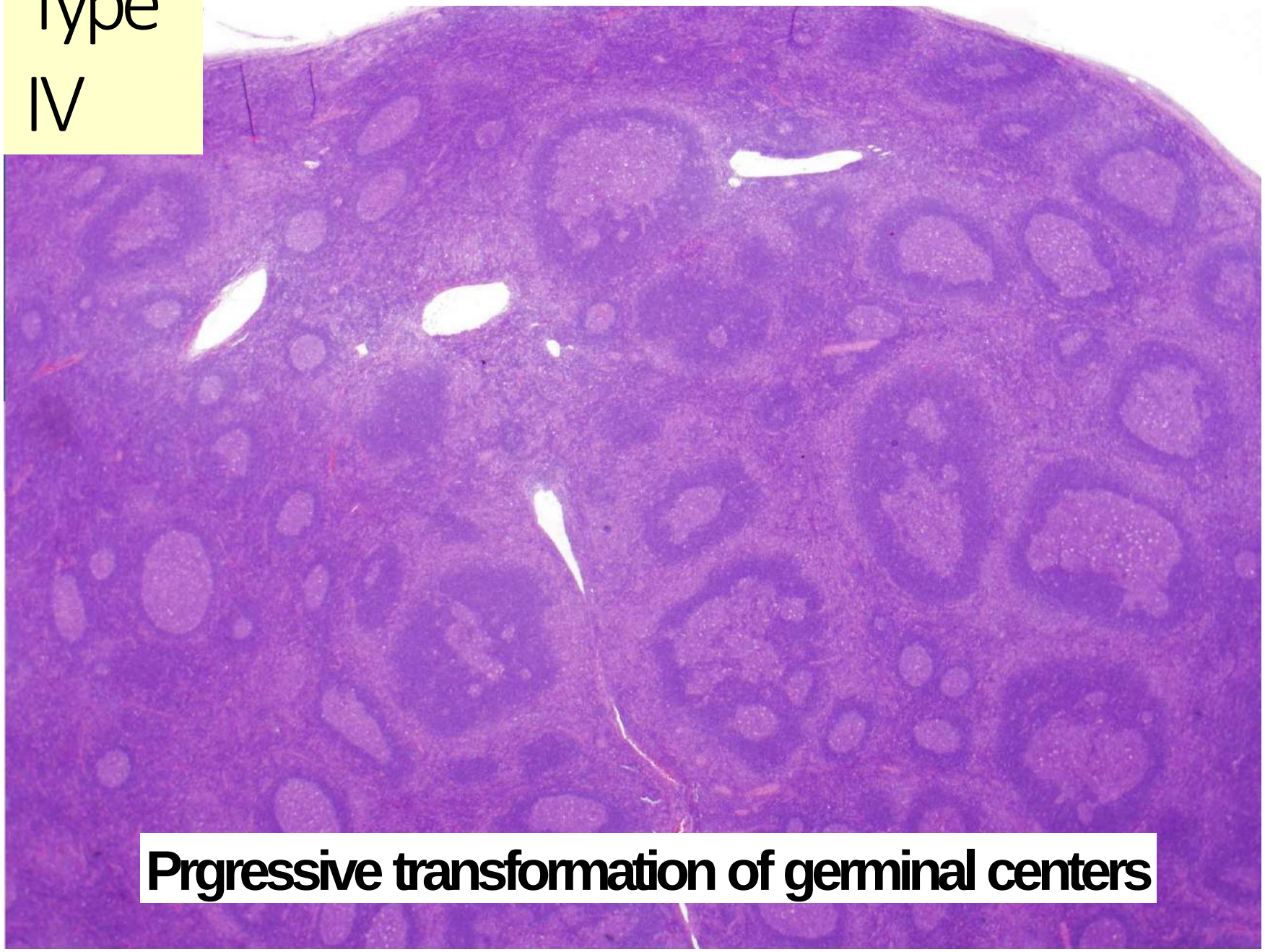


Type III

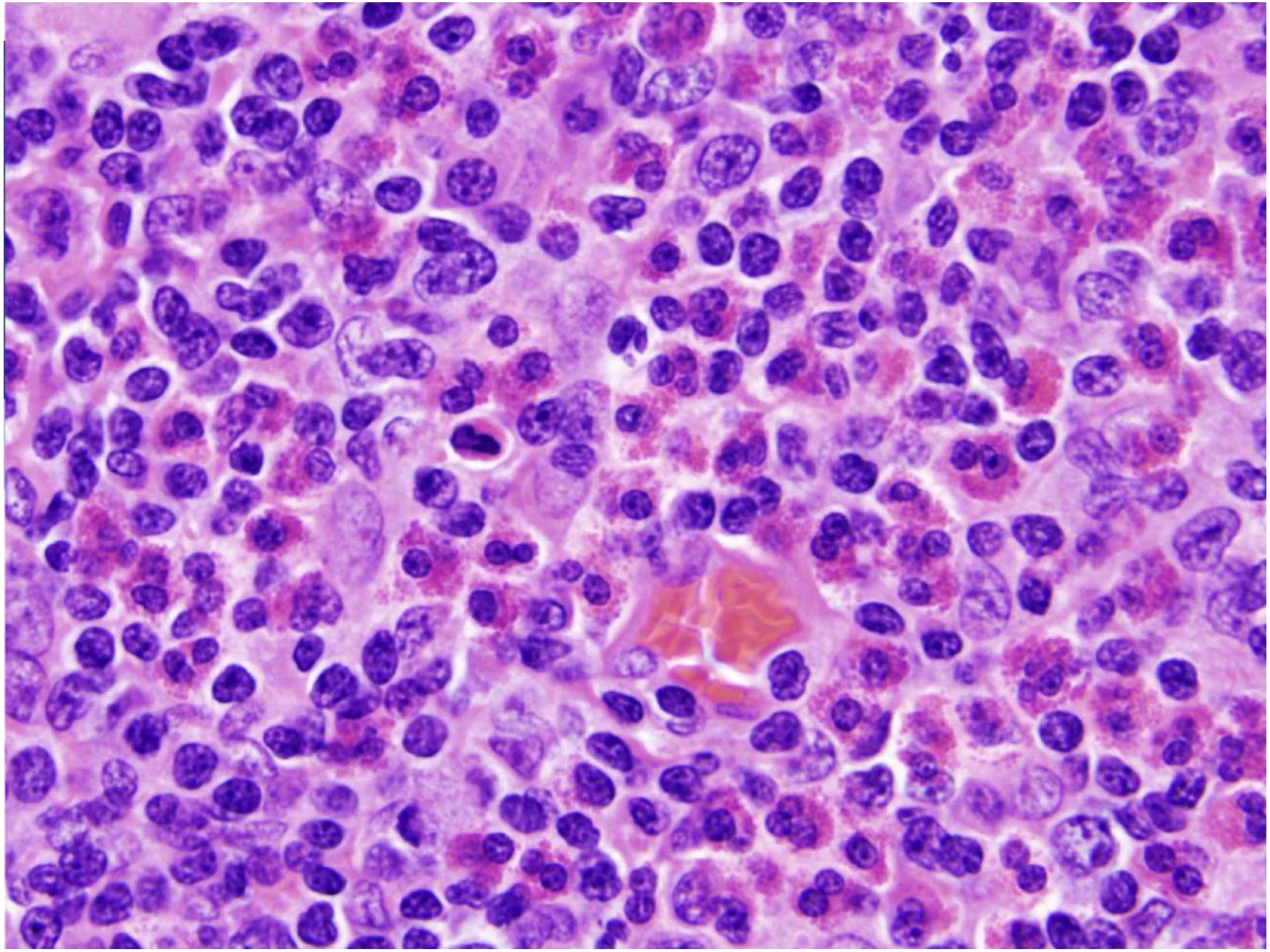


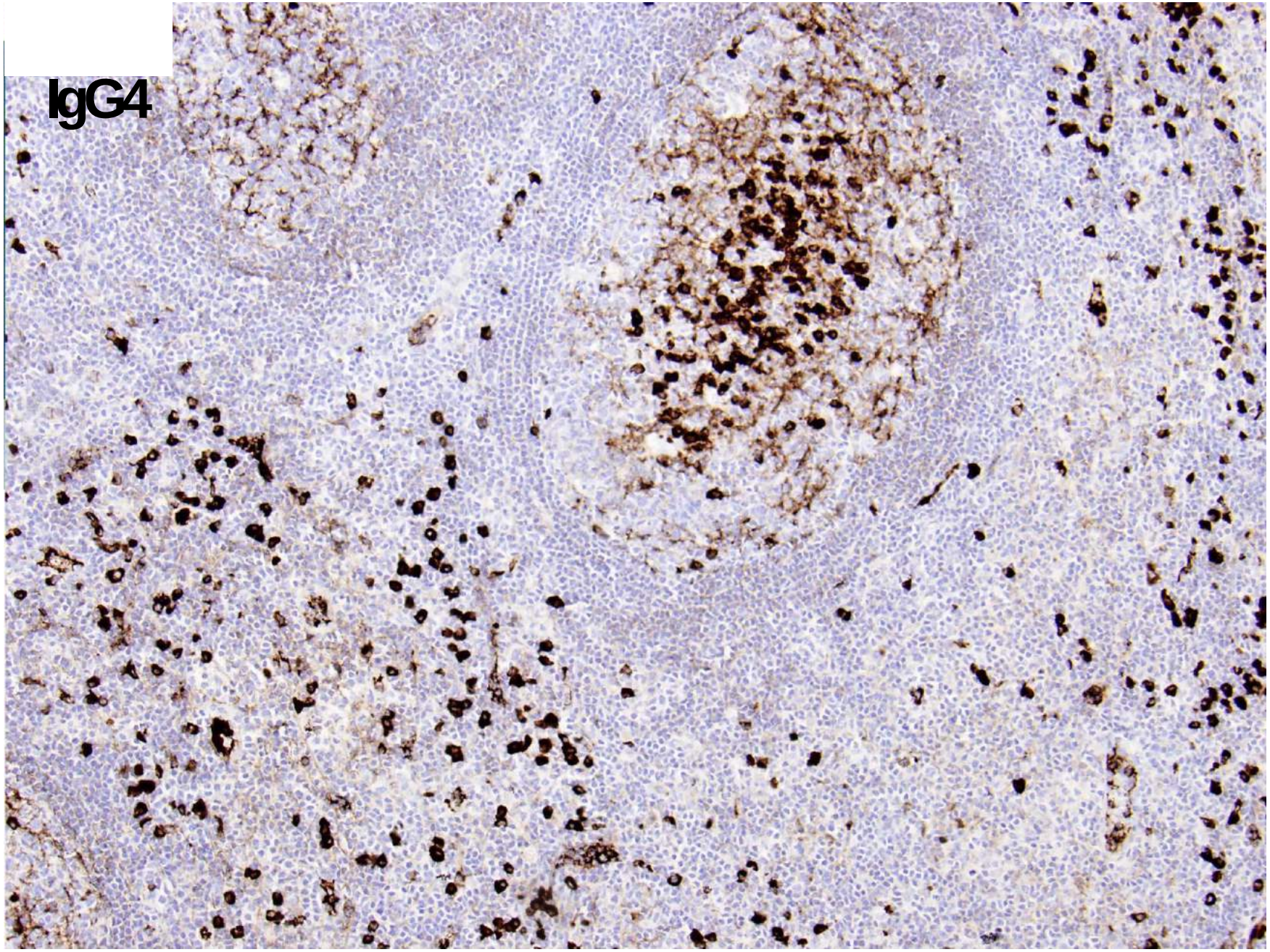
Paracortical expansion

Type IV

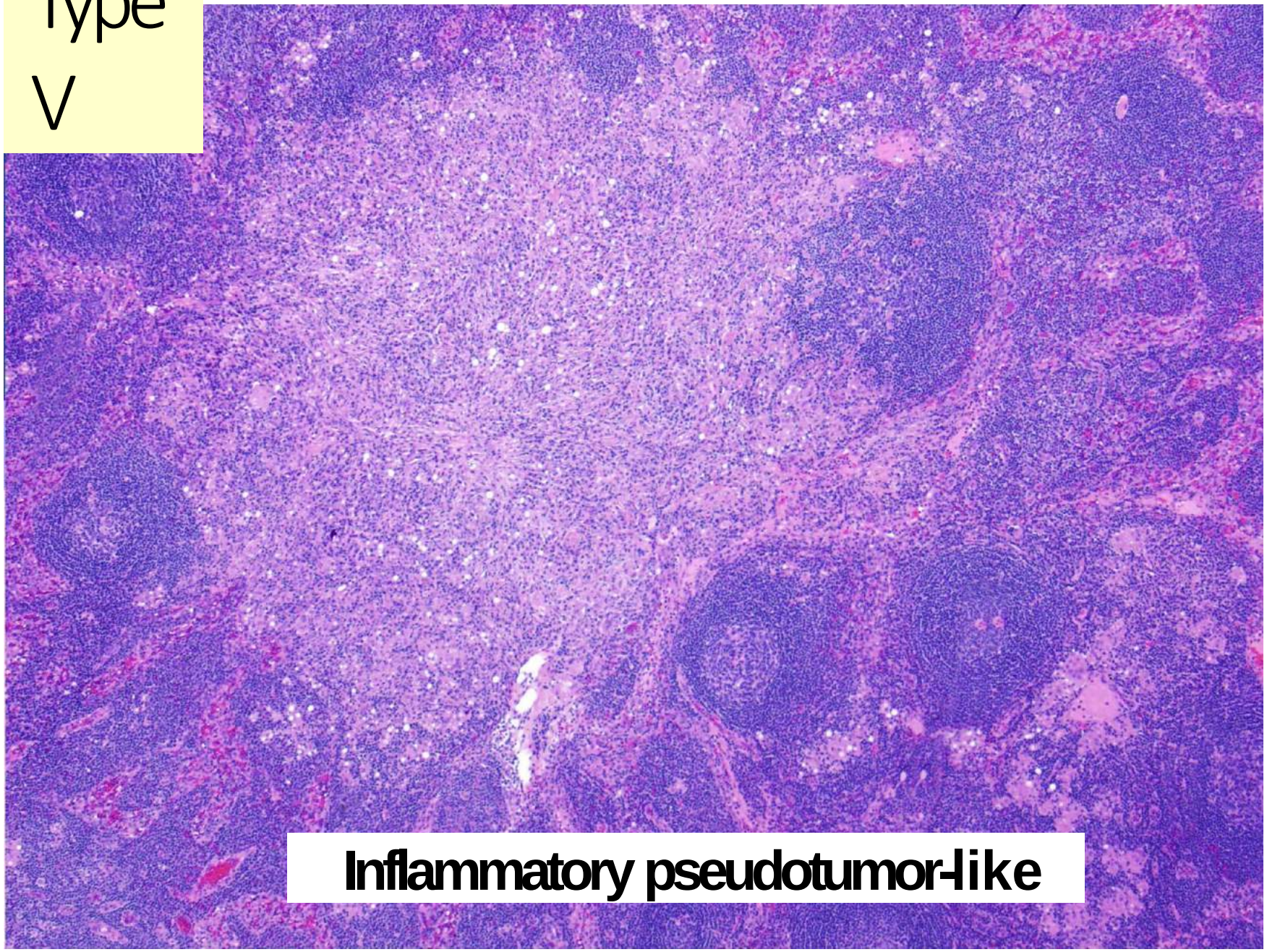


Prgressive transformation of germinal centers

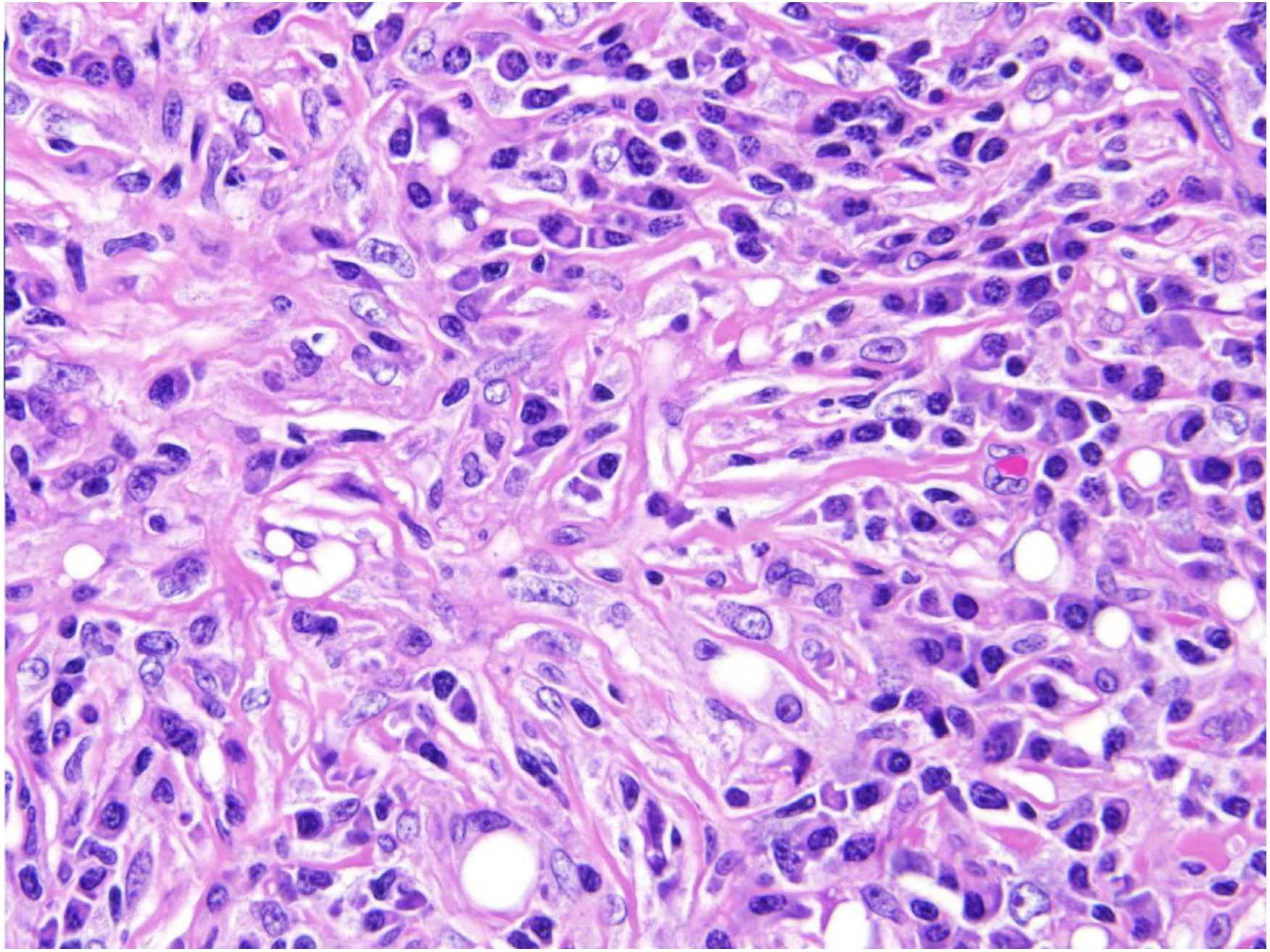



IgG4

Type V



Inflammatory pseudotumor-like



CRITERIOS DIAGNÓSTICOS ENF. IGG4 RELACIONADA bajo OKAZAKI Y UMEHARA

CRITERIO ESTRICTO DEBE SER USADO EN EL DIAGNÓSTICO

- **Sino enfermedad relacionada a IgG4 se convertiría en una “waste – basket”**

LAS TECNICAS DE IMAGEN NOVEDOSAS PUEDEN SERNOS DE AYUDA EN LINFOMA MALT Y EN IGG4?

LAS TECNICAS DE IMAGEN NOVEDOSAS PUEDEN SERNOS DE AYUDA EN LINFOMA MALT Y EN IGG4?

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Pictorial Review

Imaging manifestations of IgG4-related disease

M. Naik [a,1], S. Hesni [a,1], A. Tamimi [a], M. Hameed [a], J. Tomlinson [b], S. Poo [b], F. Tam [b], N. Strickland [a], T.D. Barwick [a], C.J. Harvey [a,*]

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LAS TECNICAS DE IMAGEN NOVEDOSAS PUEDEN SERNOS DE AYUDA EN LINFOMA MALT Y EN IGG4?

Table 2
Differential diagnosis of IgG4-related disease (IgG4-RD) and differentiating features.

Organ/region of involvement	Differential diagnoses	Differentiating features
Liver/biliary tree	PSC	PSC generally affects a younger age group and is associated with inflammatory bowel disease
	Cholangiocarcinoma	Co-existing autoimmune pancreatitis suggests IgG4-RD over cholangiocarcinoma
Pancreas	Pancreatic malignancy	Serological IgG4 levels >2x the upper limit of normal favour IgG4-RD over other pancreatic pathology
Kidneys	RCC	Multifocal or bilateral disease favours IgG4-RD
	Lymphoma	Narrowing of the collecting system is atypical of lymphoma, unlike IgG4-RD
	TCC	Renal tract obstruction is rare in IgG4-RD in contrast to TCC
Retroperitoneum	Retroperitoneal lymphoma	IgG4-RD does not tend to extend cranial to the renal hila, unlike retroperitoneal lymphoma
Lungs	Malignancy Infection Interstitial/organising pneumonia Sarcoidosis	Distinguishing IgG4-RD from other conditions typically relies on the presence of multisystem involvement
Heart/Mediastinum	Other vasculitides	Distinguishing other vasculitides from IgG4-RD typically relies on the presence of multisystem involvement
Salivary glands	Sjögren's-related sialadenitis	A highly vascular nodal pattern on ultrasound favours IgG4-RD compared to the dot-like vascular pattern seen in Sjögren's-related sialadenitis Absence of parotid involvement favours IgG4-RD
Pituitary	Malignancy Tuberculosis Sarcoidosis	Imaging is non-specific and biopsy/resection may be required if no multisystem involvement
Orbits	Thyroid eye disease	Sparing of tendons and preferential involvement of lateral rectus suggests IgG4-RD Infraorbital nerve enlargement, adjacent sinus mucosal thickening and infiltration of orbital fat favours IgG4-RD

RCC, renal cell carcinoma; TCC, transitional cell carcinoma; PSC, primary sclerosing cholangitis

CASO CLINICO

- **DX: Linfoma de la zona marginal 1° anexo ocular EC IVB x comp. Partes blandas asociado a enfermedad IgG4 relacionada. (2016).**

- **El paciente recibió R-CHOP x 6 cursos con adecuada rpta clínica. Continua vivo al seguimiento.**

CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

PB2264 CLINICAL FEATURES, TREATMENT PATTERNS, AND OUTCOMES AMONG 120 PATIENTS WITH MALT LYMPHOMA IN LATIN AMERICA: A STUDY FROM THE GRUPO DE ESTUDIO LATINOAMERICANO DE LINFOPROLIFERATIVOS (GELL)

Topic: 18. Indolent and mantle-cell non-Hodgkin lymphoma - Clinical

Seisha Alana Von Glasenapp Gossen*[1], Denisse Castro Uriol[2], Brady Beltran Garate[2], Sally Paredes[2], Victoria Otero[3], Sofia Rivarola[4], César Camilo Carías Alvarado[5], Juan Carlos Barrios Menéndez, Ana Carolina Oliver[6], Victoria Irigoin[7], Julio Fernández Aguia[8], Fabiola Valvert, Henry Idrobo Quintero[9], Yudy Lorena Delgado Pascuaza[9], Gloria Caterine Peréz Mingan[9], Humberto Martinez[10], Bryan Valcarcel[11], Luis Malpica[12]

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GELL Grupo de Estudio Latinoamericano de Linfoproliferativos

CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

- 120 pts were identified
  - 47 (39%) Peru
  - 25 (21%) Argentina
  - 21 (18%) Guatemala
  - 11 (9%) Cuba
  - 7 (6%) Uruguay
  - 5 (4%) Paraguay
  - 4 (3%) Colombia
- Median age at diagnosis was 63 years (27-93) with slight female predominance (51%).
- Localized stage (I-II) at diagnosis was presented in 93 (78%) pts.
- Gastric and ocular adnexal were the most common disease sites (n=63, 52% and n=25, 21%, respectively).
- Primary cutaneous MALT was found in 8 (7%) pts.

CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

- Infectious etiology with Hp, Chlamydia psittaci and Borrelia burgdorferi were identified in 65% (41/63), 4% (1/25) and 13% (1/8) of gastric, ocular adnexal and cutaneous MALT lymphoma, respectively.
- Other MALT cases had Hep C (n=2), IgG4 disease (n=3), Hashimoto thyroiditis (n=13) and Sjogren's syndrome (n=9).
- Therapy patterns were heterogeneous.
  - In localized stage, watch-and-wait (n=29, 31%) and single-agent rituximab (n=22, 24%) were the most common frontline approaches whereas R-CHOP (n=17, 63%) was common in advanced disease.
  - Frontline RT alone was given to 14 (15%) pts with localized disease. Median RT dose was 24 Gy (4-40).

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CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

- **In the entire cohort, the overall response rate (ORR) to frontline was 95% (CR 76%). With a median follow up of 45 months (37-55) the 4-year and median OS and PFS for the entire cohort was 84% and not reached (NR), and 60% and NR, respectively.**

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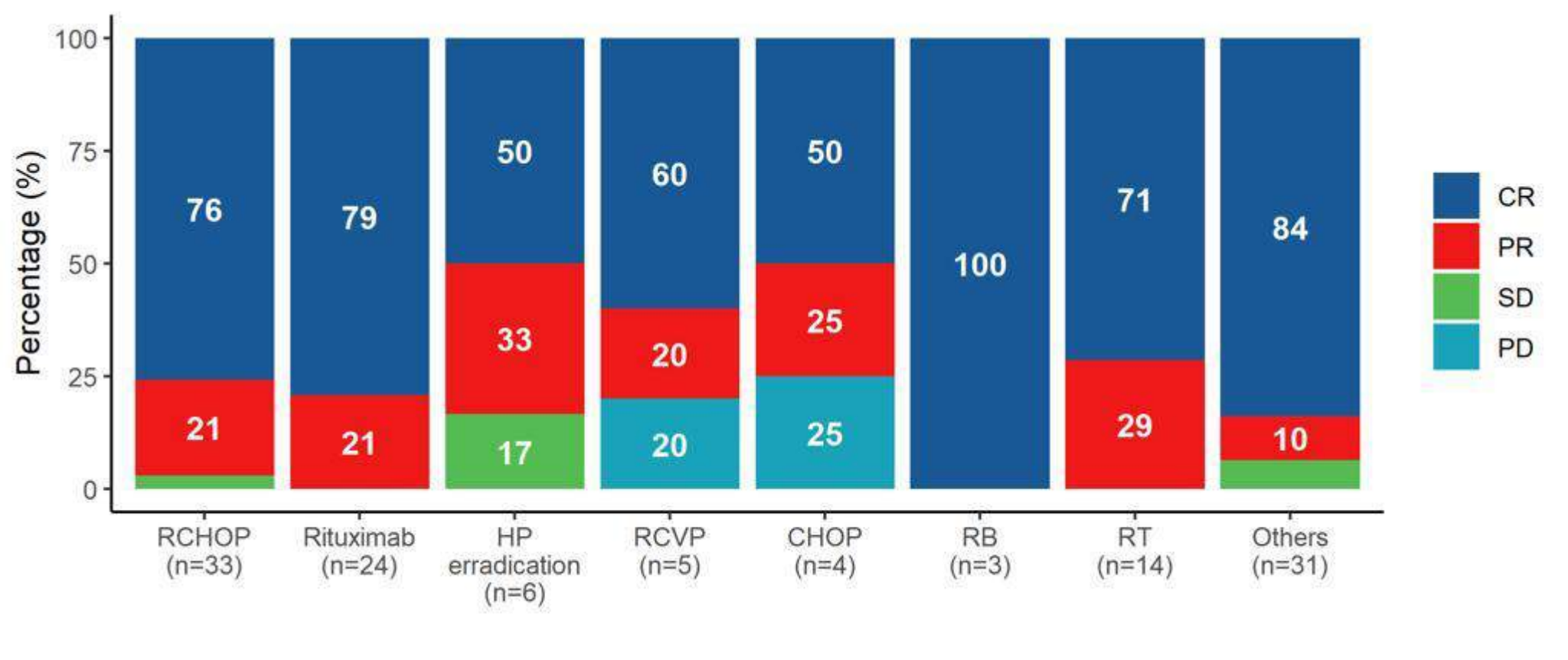
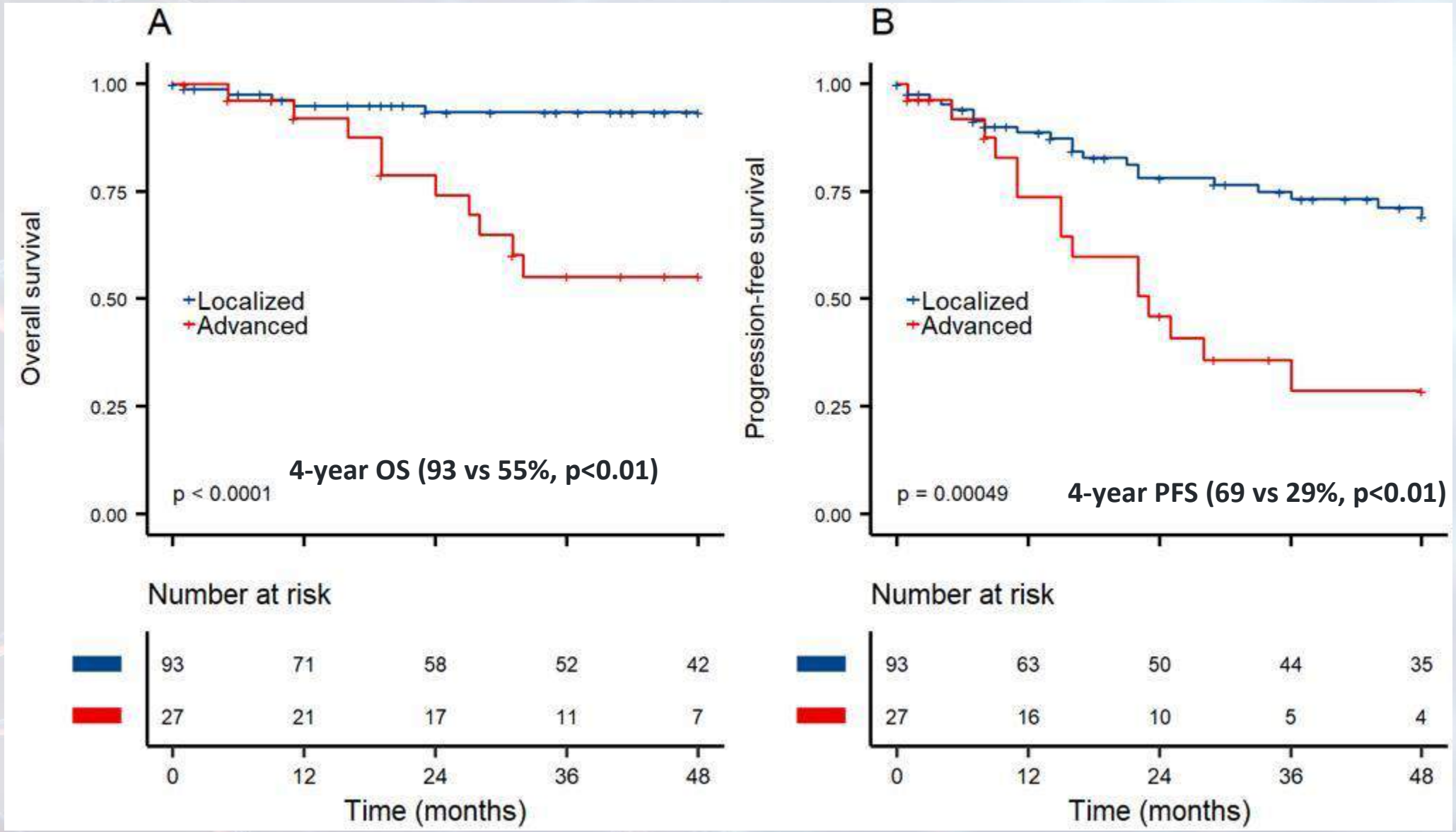
Cómo nos ido a LATAM en términos de manejo de Linfoma MALT?



Cómo nos ido a LATAM en términos de manejo de Linfoma MALT?



CÓMO NOS IDO A LATAM EN TÉRMINOS DE MANEJO DE LINFOMA MALT?

- **Second line therapy was used in 35 (39%) pts. ORR to second line were 97% (CR 74%). The 4-year and median OS in relapsed pts were 71% and NR.**
- **No patients received novel agents during their treatment course (ie BTK, clinical trial).**

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CONCLUSIONES

- IgG4 es una entidad que fue subdiagnosticada y el objetivo es tenerla presente y como sospecha de diagnóstico clínico.
- 3 factores son preponderantes en el diagnóstico: eosinofilia, infiltrados plasmocitarios y linfadenopatías.
- La unidad clínica con Anatomopatología y la discusión multidisciplinaria son pilares en el diagnóstico y tratamiento.
- La rpta en IgG4 es excelente a corticoides, no obstante, debe ser valorada la coexistencia con linfomas u otras entidades.

CONCLUSIONES

- Continúa siendo un reto en LATAM la disponibilidad de exámenes, la uniformidad de los tratamientos, el acceso a los mismos y la oportunidad y participación en ensayos clínicos.

GRACIAS…


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