Sociedade Brasileira de Patologia

Caso do mês – Fevereiro/2016

Dr. Igor Santos Costa

CDERM
CENTRO DE DERMATOLOGIA DONA LIBÂNIA

ARGOS PATOLOGIA

Um quadro confuso

- Paciente masculino, 77 anos;
- Há 1 mês e ½ com tontura, adinamia, anemia, referindo perda de peso de 7 kg nesse período;
- História de lesões em placa na pele do tronco, MMSS e MMII há 20 dias, sendo interrogado MHBT, Farmacodermia e LCCT, porém quando foi para a biópsia não haviam mais lesões;
- Paciente hipertenso, dislipidêmico e revascularizado;
- Investigado pela neurologia.

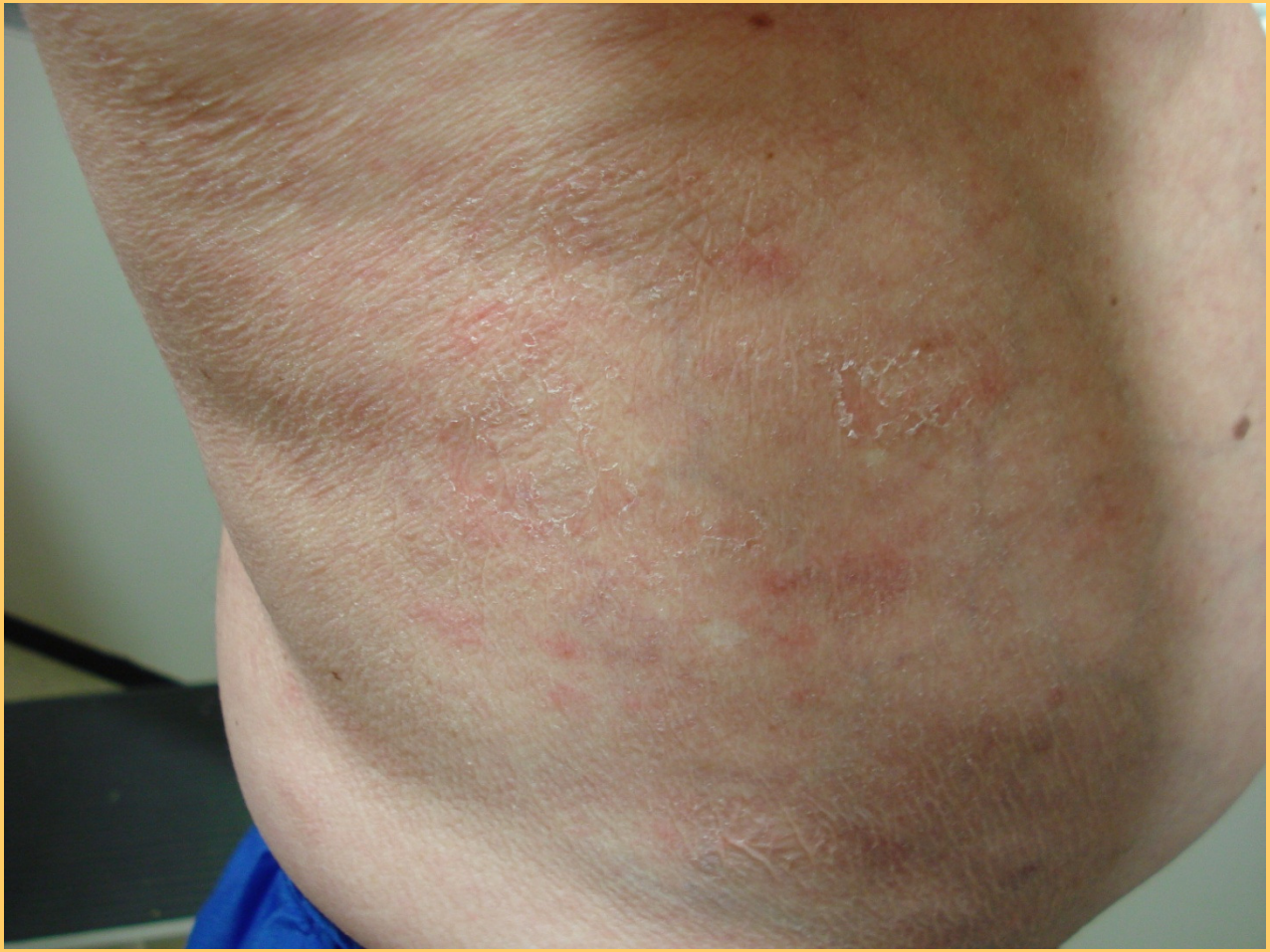
Em uso de vários medicamentos

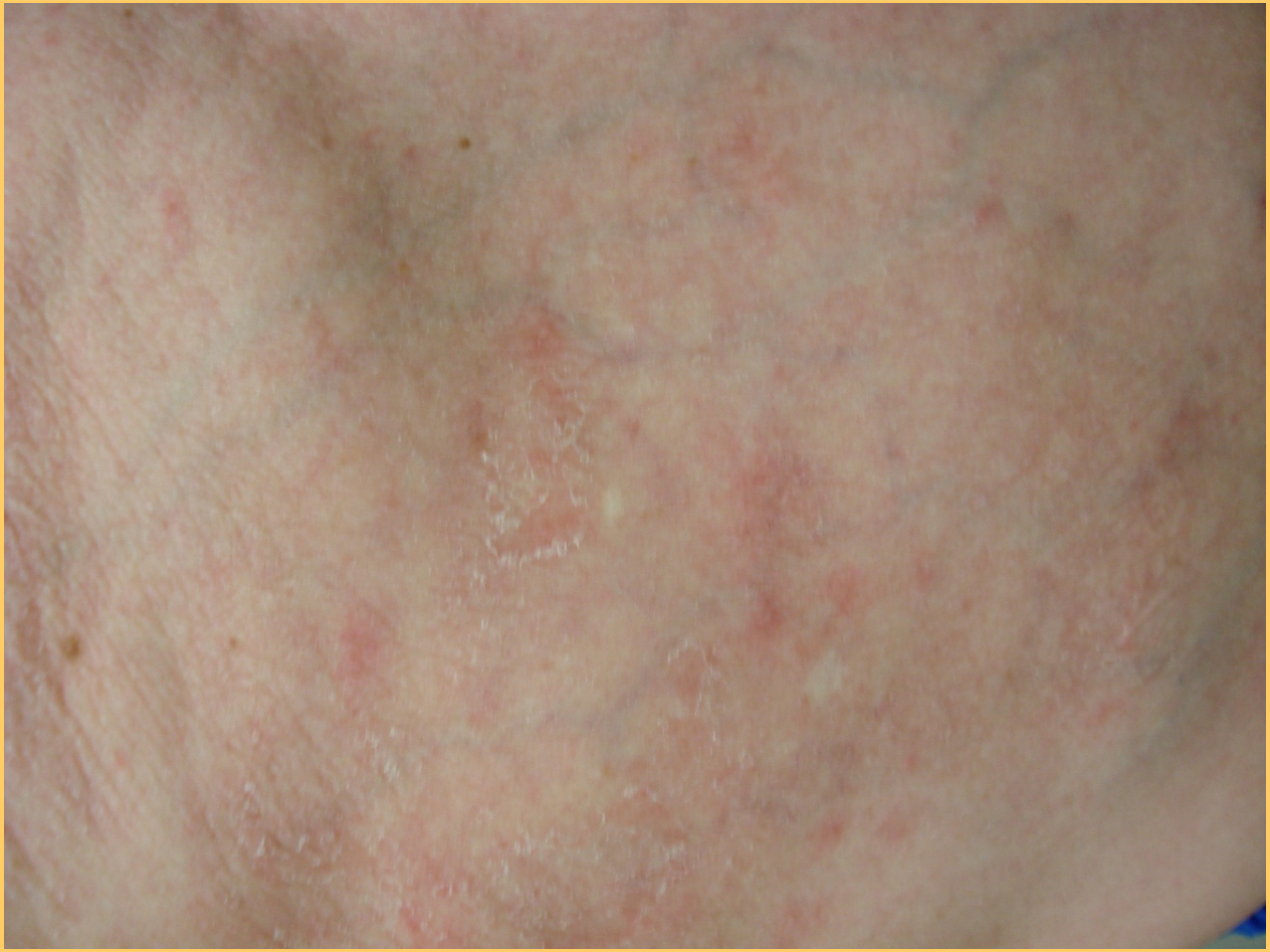
- Pantoprazol;
- Carbamazepina (crises convulsivas);
- Prolopa (parkinson);
- Marevan (TVP de repetição);
- Furosemida e Somalgin (hipertensão e cardiopatia).

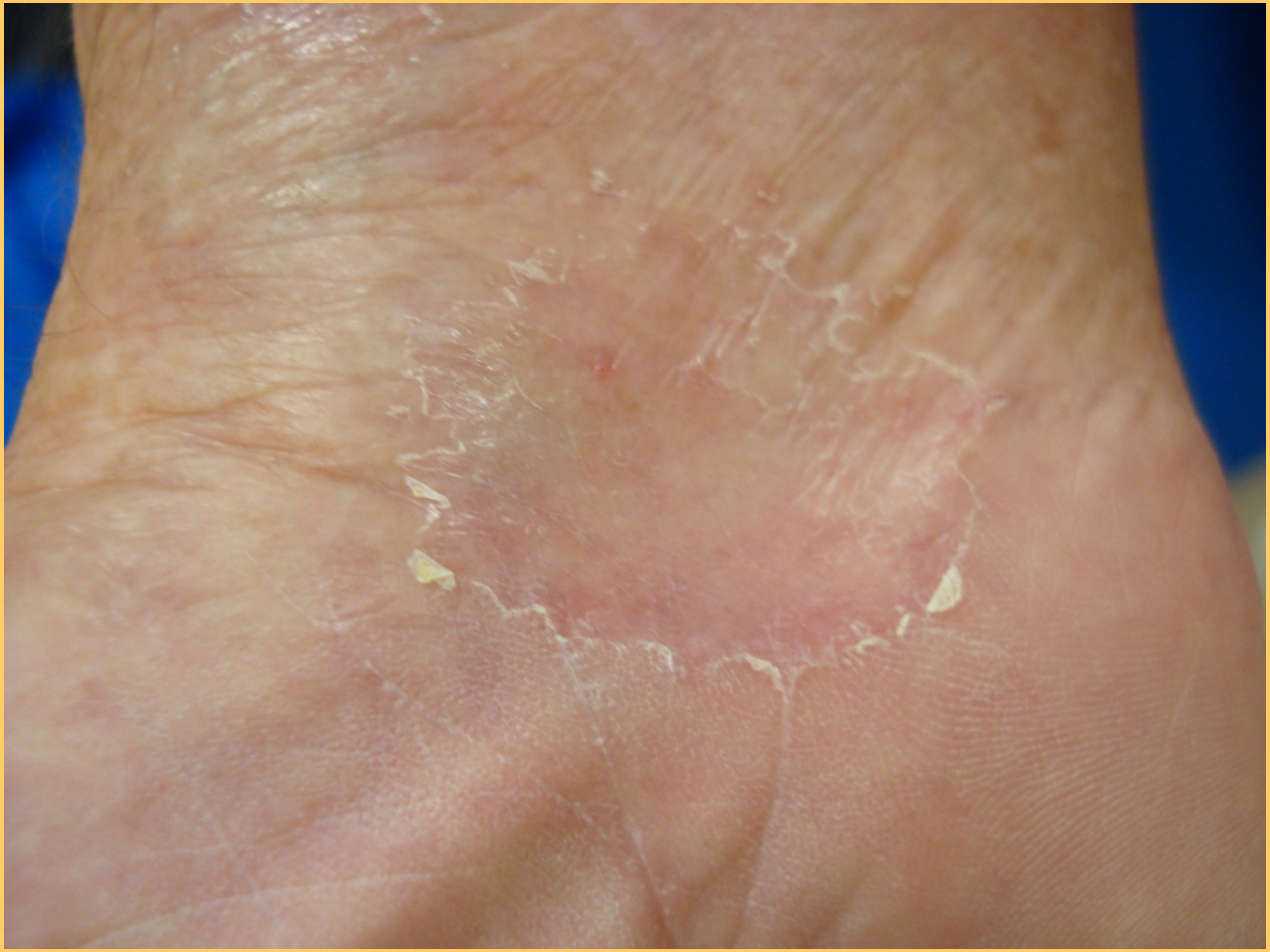
E com vários exames complementares

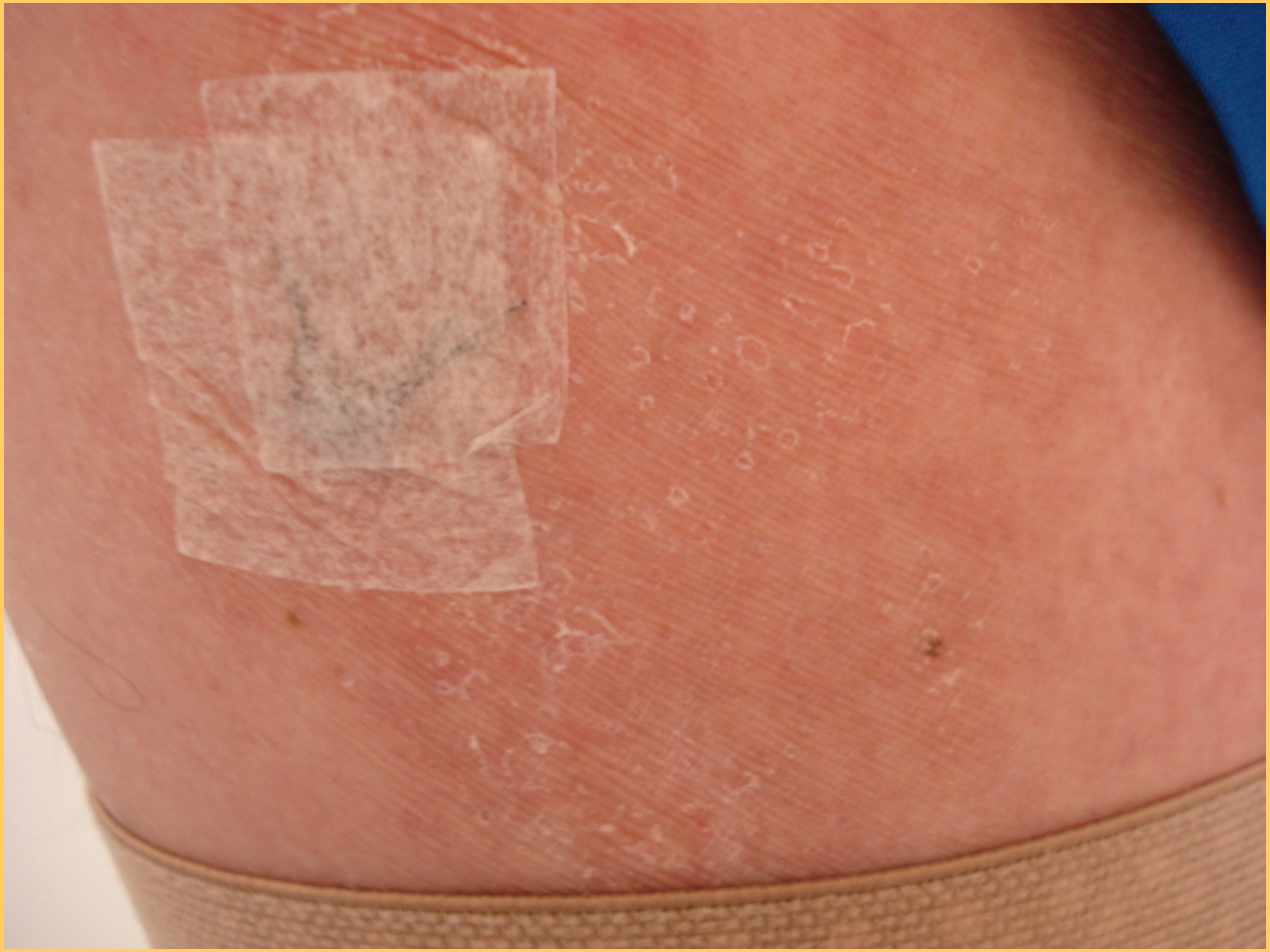
- US tireóide – nódulo no lobo inferior
- Ecocardio – insuficiência mitral e aórtica leves, hipocinesia anterior discreta
- US abdominal – hepatomegalia sem nódulos
- Rx e TC de tórax – atelectasias subsegmentares nos lobos inferiores, pequeno derrame pleural bilateral
- Hb 10,7 Leucocitos 10930 (9432 neu, 120 eos, 43 baso, 1040 linf e 295 mono).

Solicitado parecer da dermatologia por novas lesões cutâneas





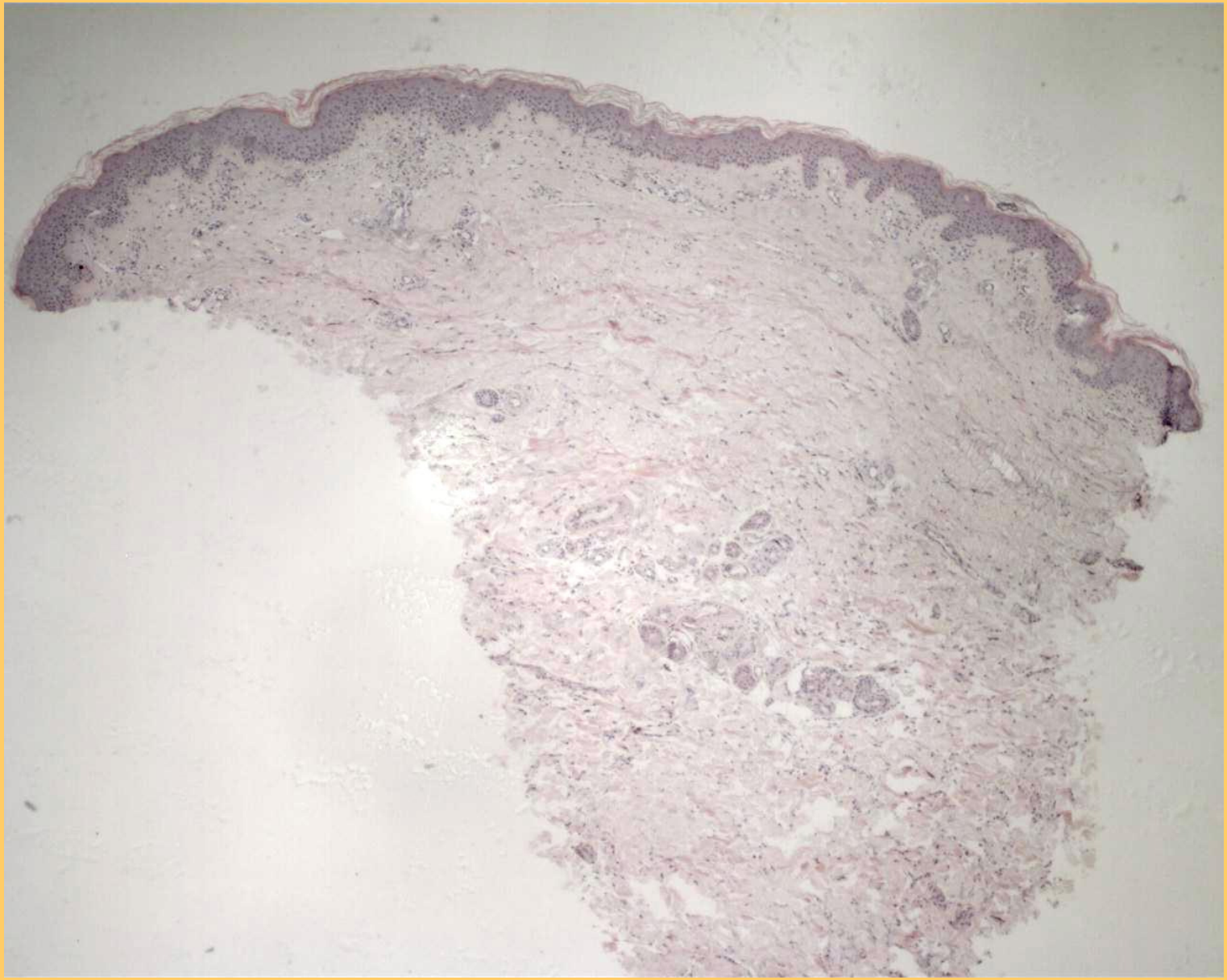


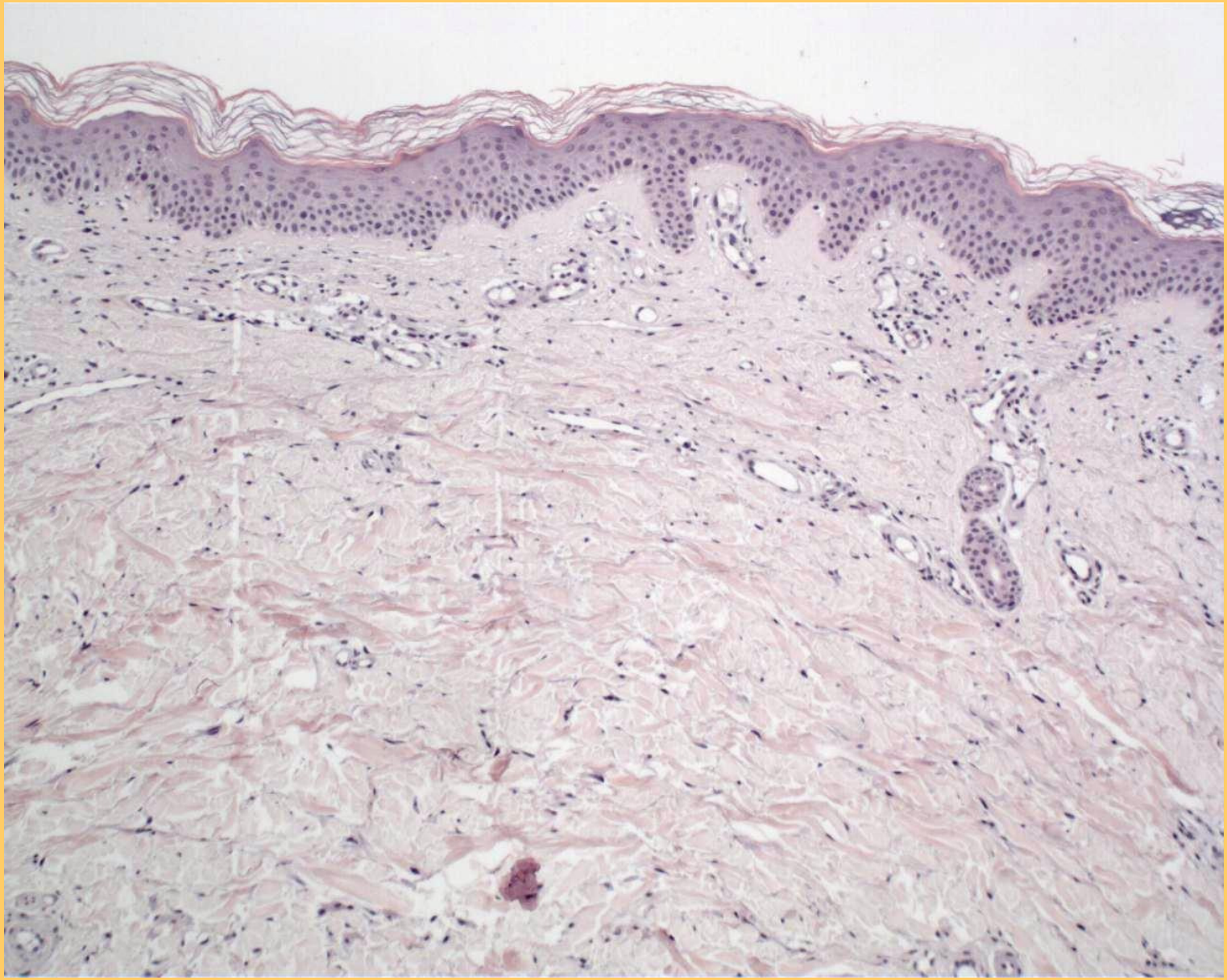


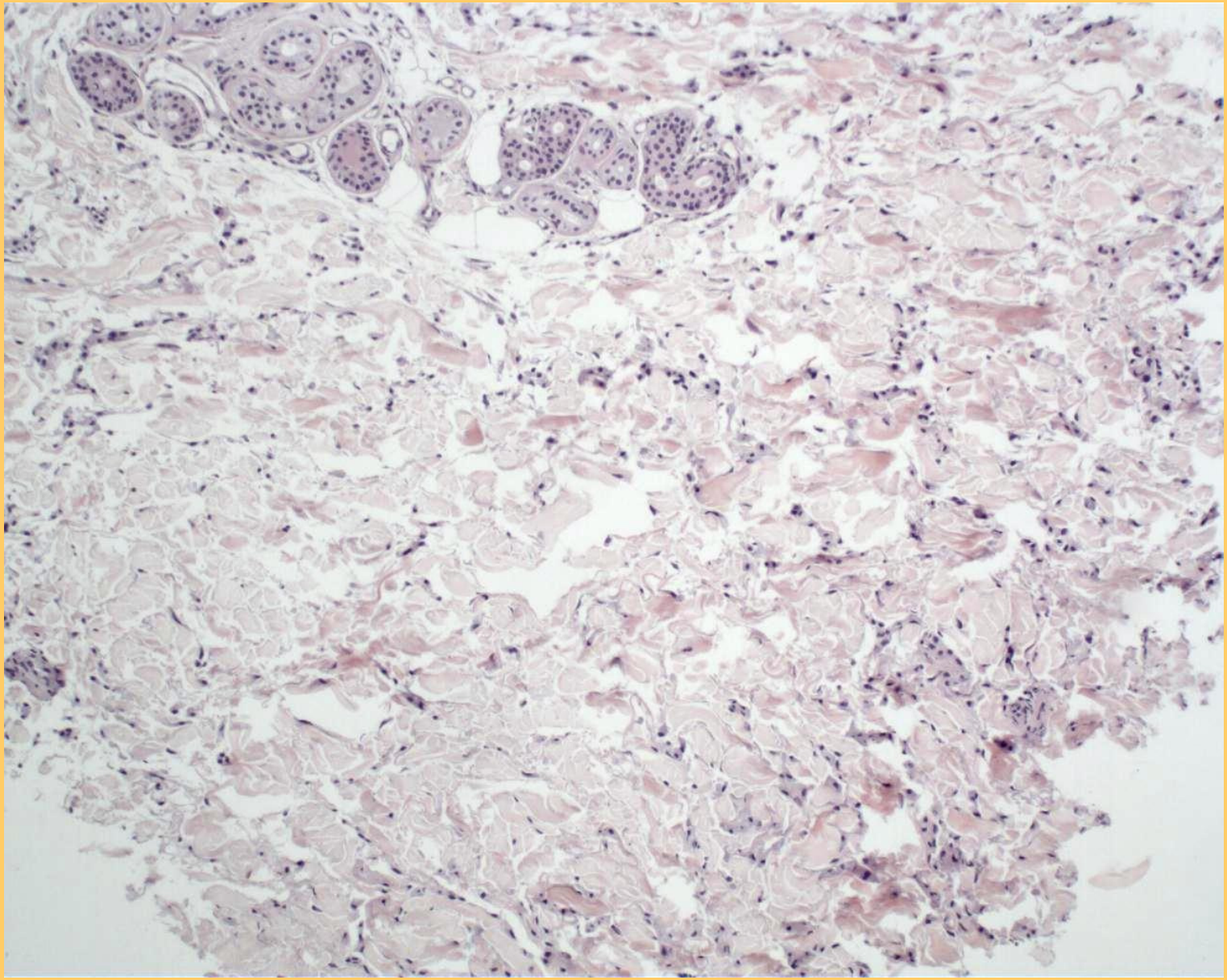
Hipóteses clínicas

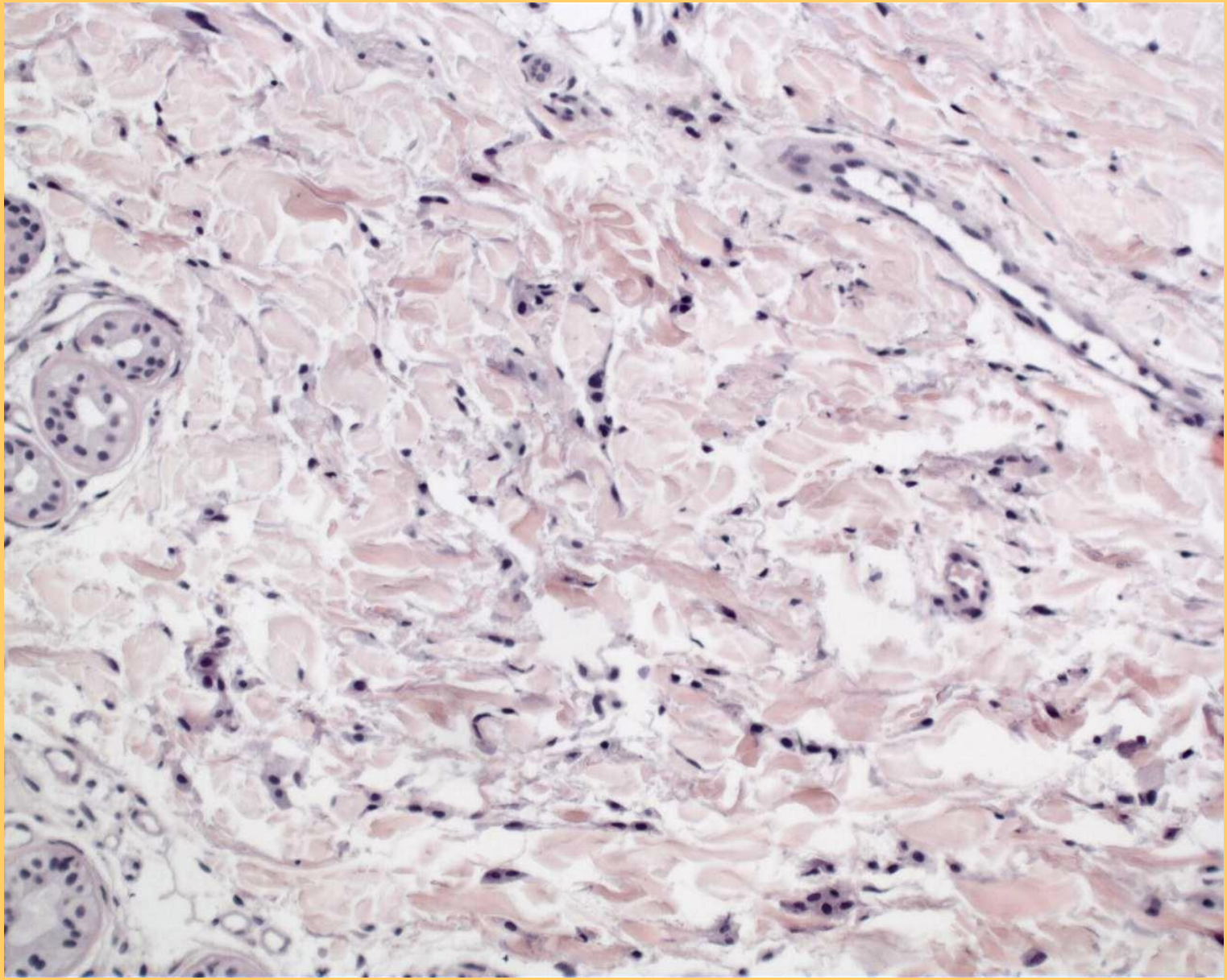
- A- Dermatite de contato
- B- Farmacodermia
- C- Eritema giratum repens
- D- LCCT
- E- Vasculite
- F- Tinha
- G- Outros

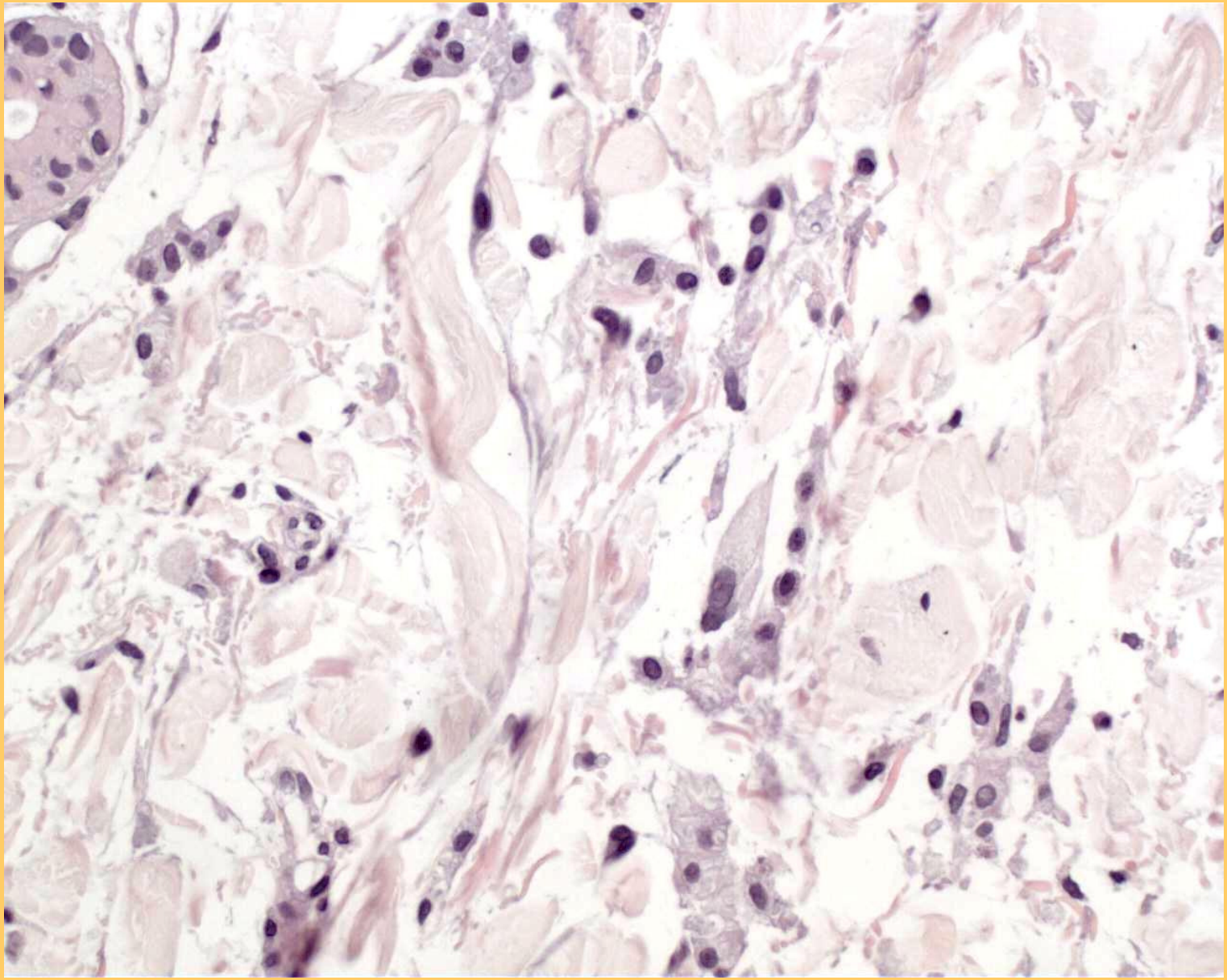
A biópsia











Hipóteses histopatológicas

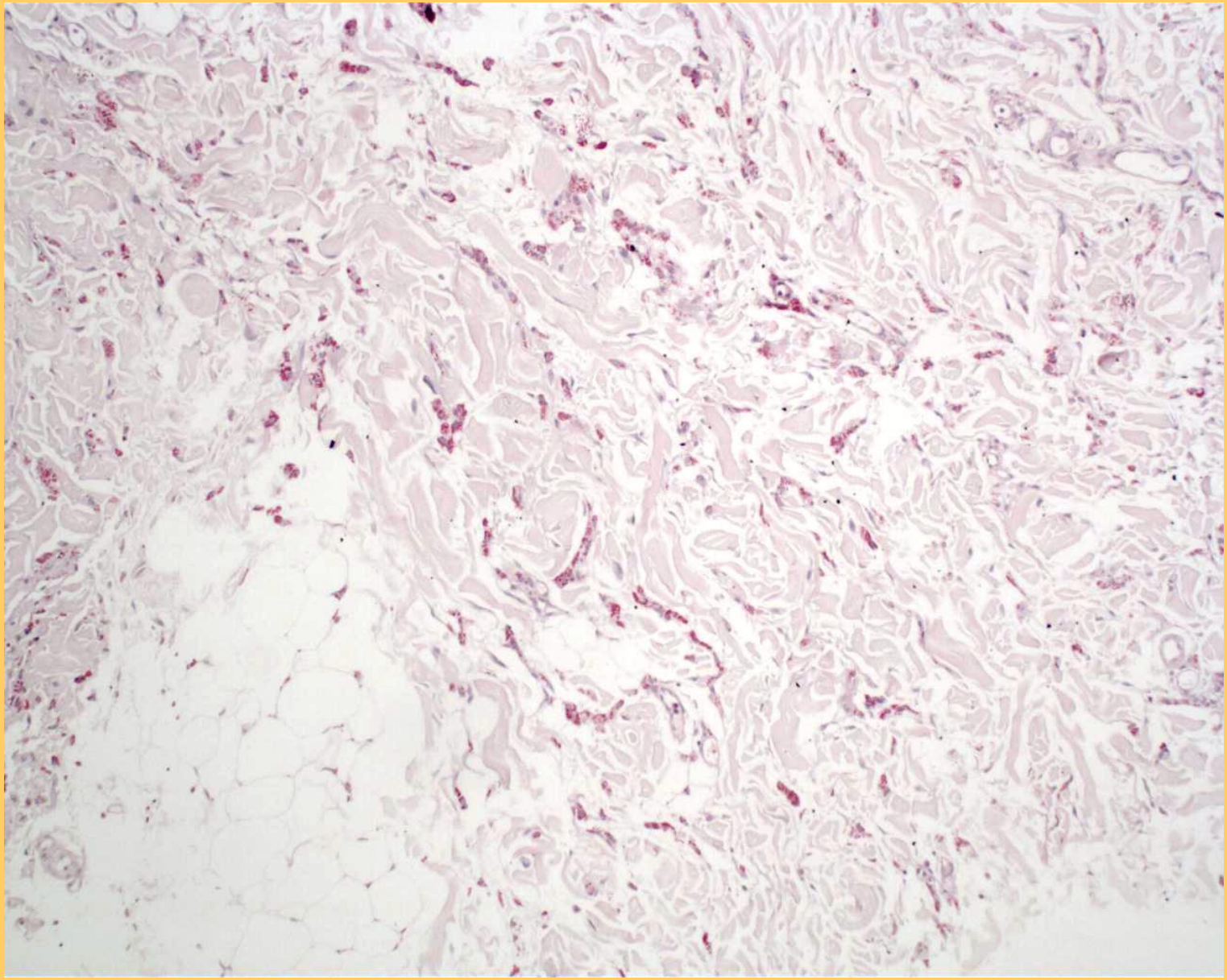
- A- Neoplasia primária x metastática
- B- Doença infecciosa
- C- Doença de depósito
- D- Outras causas

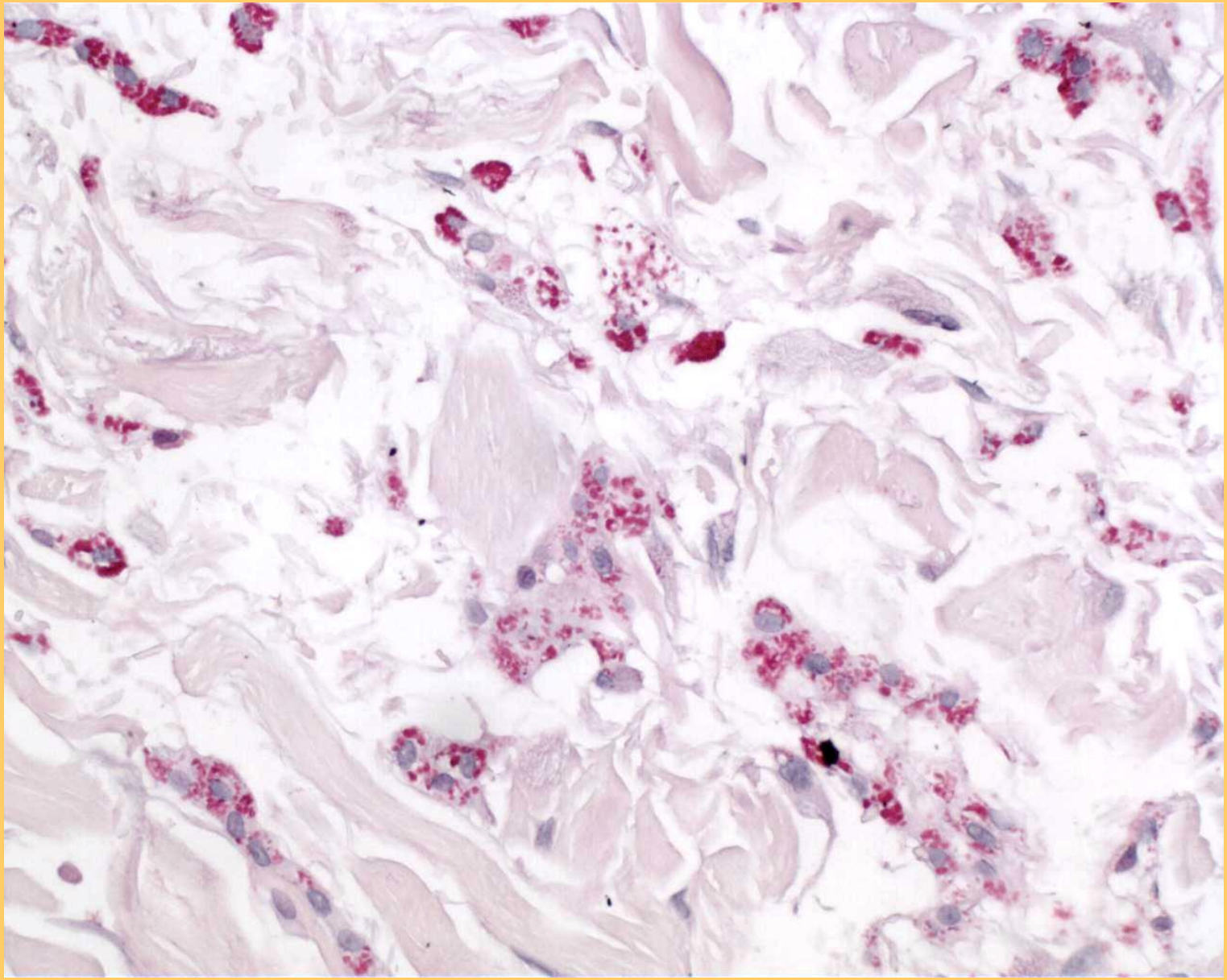
O padrão de “derme ocupada” (busy dermis)

- Granuloma anular, dermatofibroma, dermatite intersticial granulomatosa da DTC;
- Vasculite neutrofílica, Sweet, foliculite em resolução;
- Sarcoma de Kaposi (fase inicial), melanoma desmoplásico, metástase de carcinoma mamário, leucemia cutis, mastocitose, etc.

Madke B, Doshi B, Khopkar U, Dongre A. Appearances in dermatopathology: the diagnostic and the deceptive. Indian J Dermatol Venereol Leprol. 2013 May-Jun;79(3):338-48.

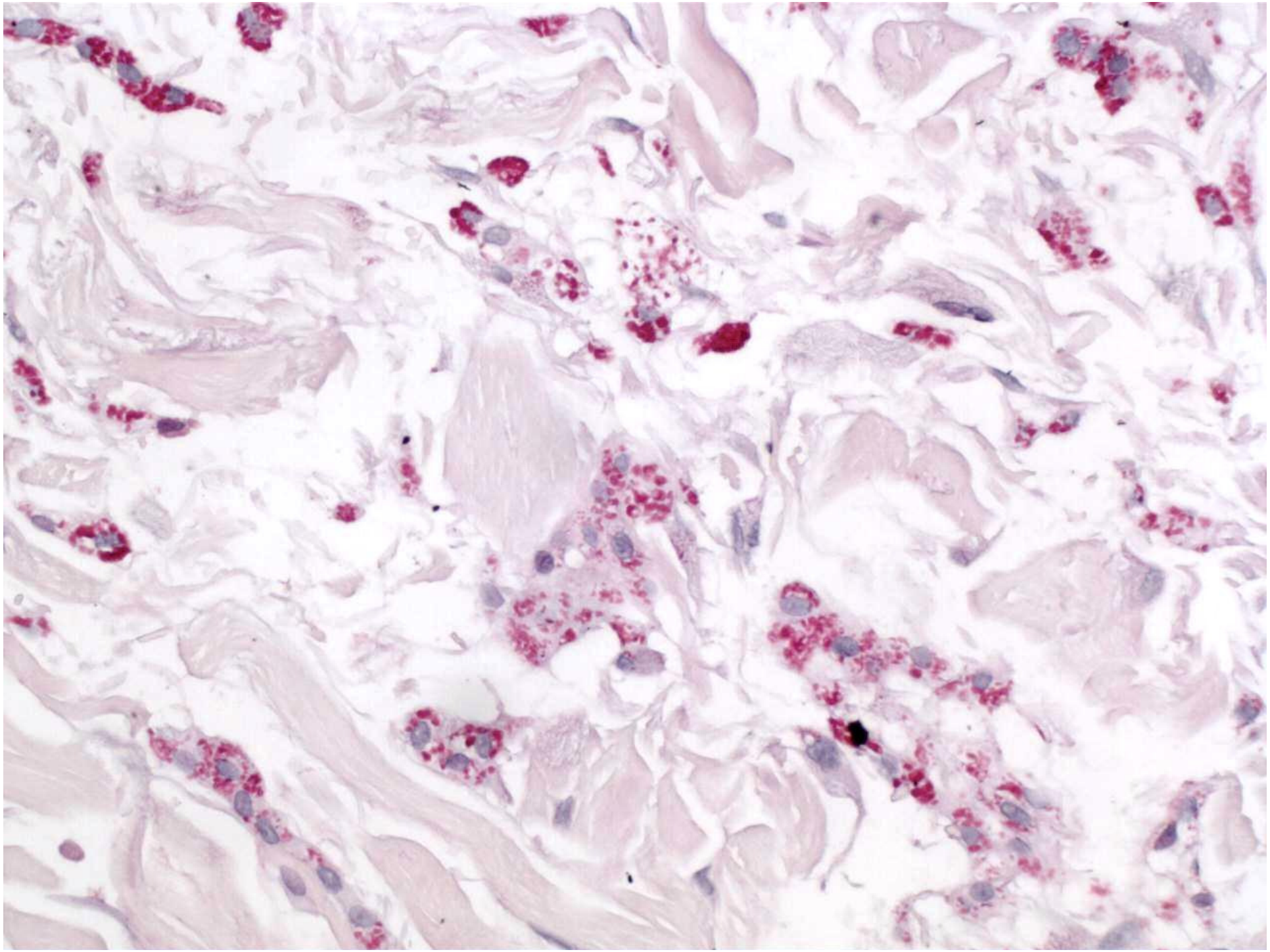
O PAS



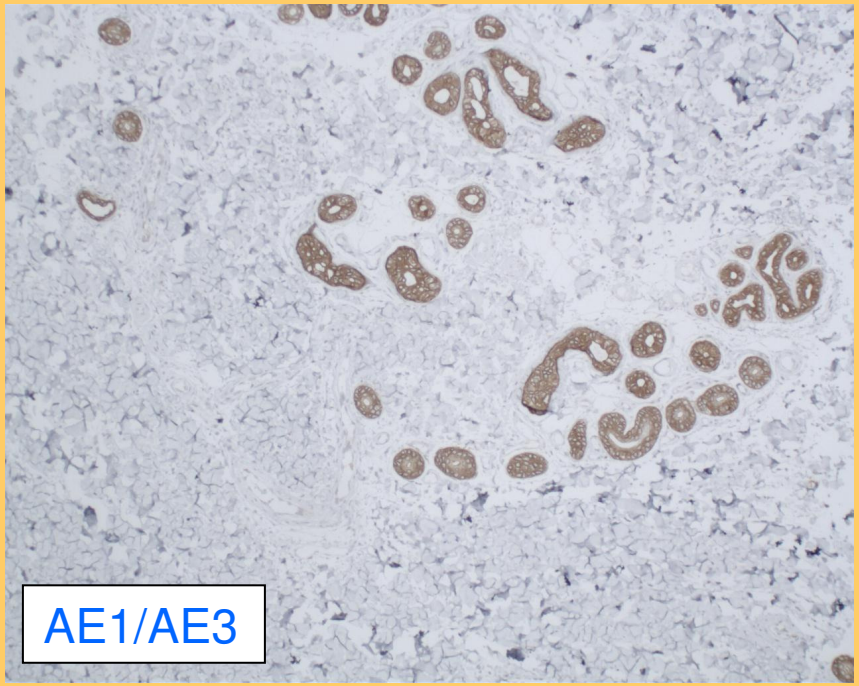


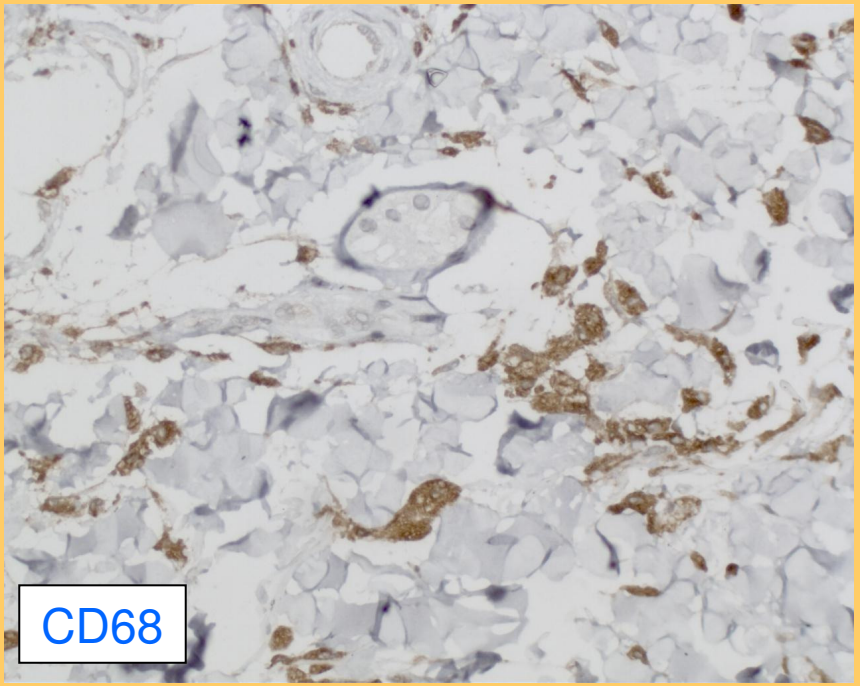
Hipóteses histopatológicas

- Células PAS-positivo e Diastase-resistente
- Neoplasia?
- Doença infecciosa?
- Doenças de depósito?
- Outras doenças – mastocitose? Leucemia cutis? Melanoma?



Imunoistoquímica





Com base nos achados morfológicos, histoquímicos e imunohistoquímicos, qual o seu diagnóstico?

Diagnóstico:

Manifestação cutânea da Doença de Whipple

XXXIII Symposium of the International Society of Dermatopathology 2012

Am J Dermatopathol. 2012 Apr;34(2):182-7. doi: 10.1097/DAD.0b013e318221ba55.

Treated Whipple disease with erythema nodosum leprosum-like lesions: cutaneous PAS-positive macrophages slowly decrease with time and are associated with lymphangiectases: a case report.

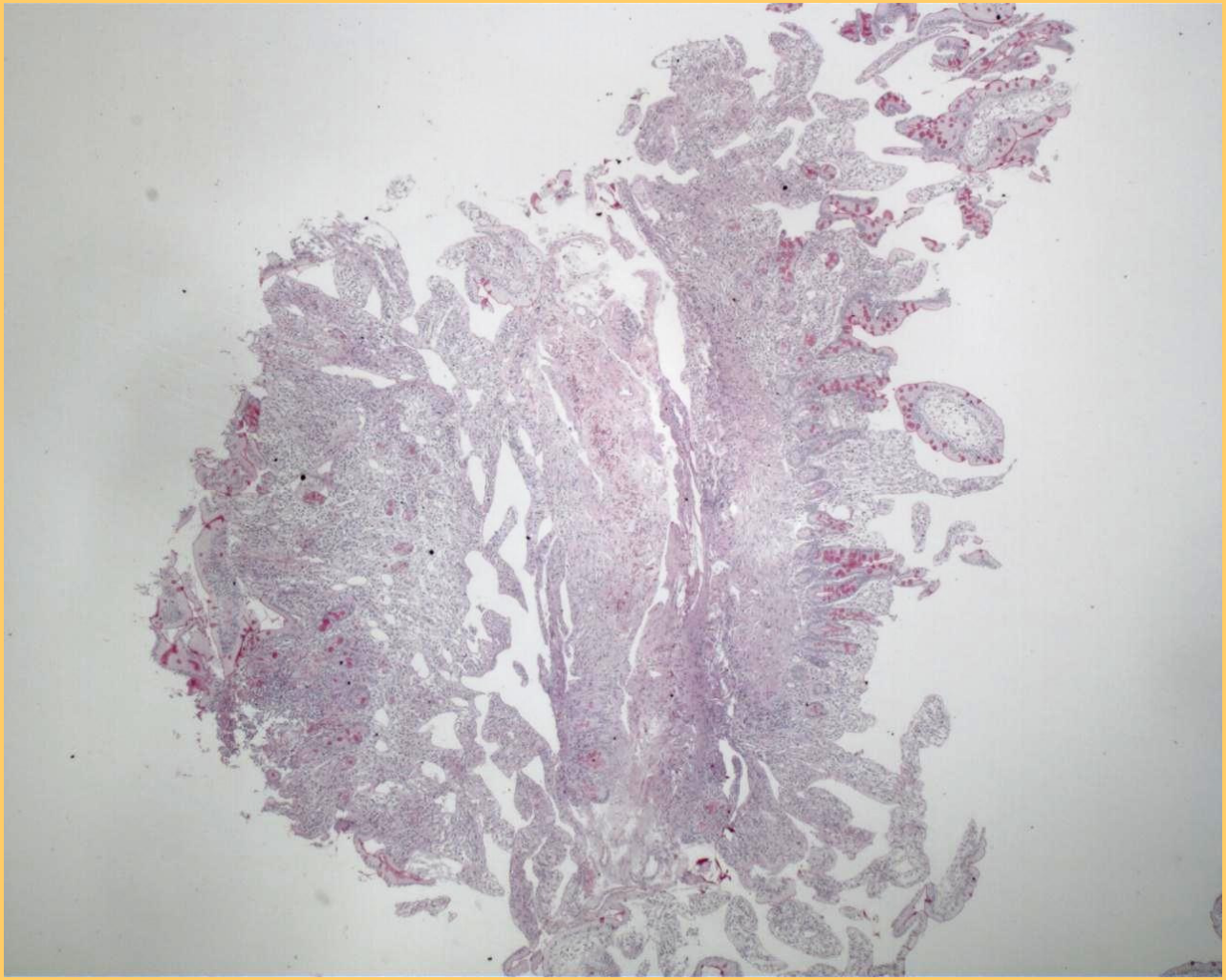
Paul J, Schaller J, Rohwedder A, Carlson JA.

Divisions of Dermatopathology and Dermatology, Department of Pathology, Albany Medical College, Albany, NY, USA.

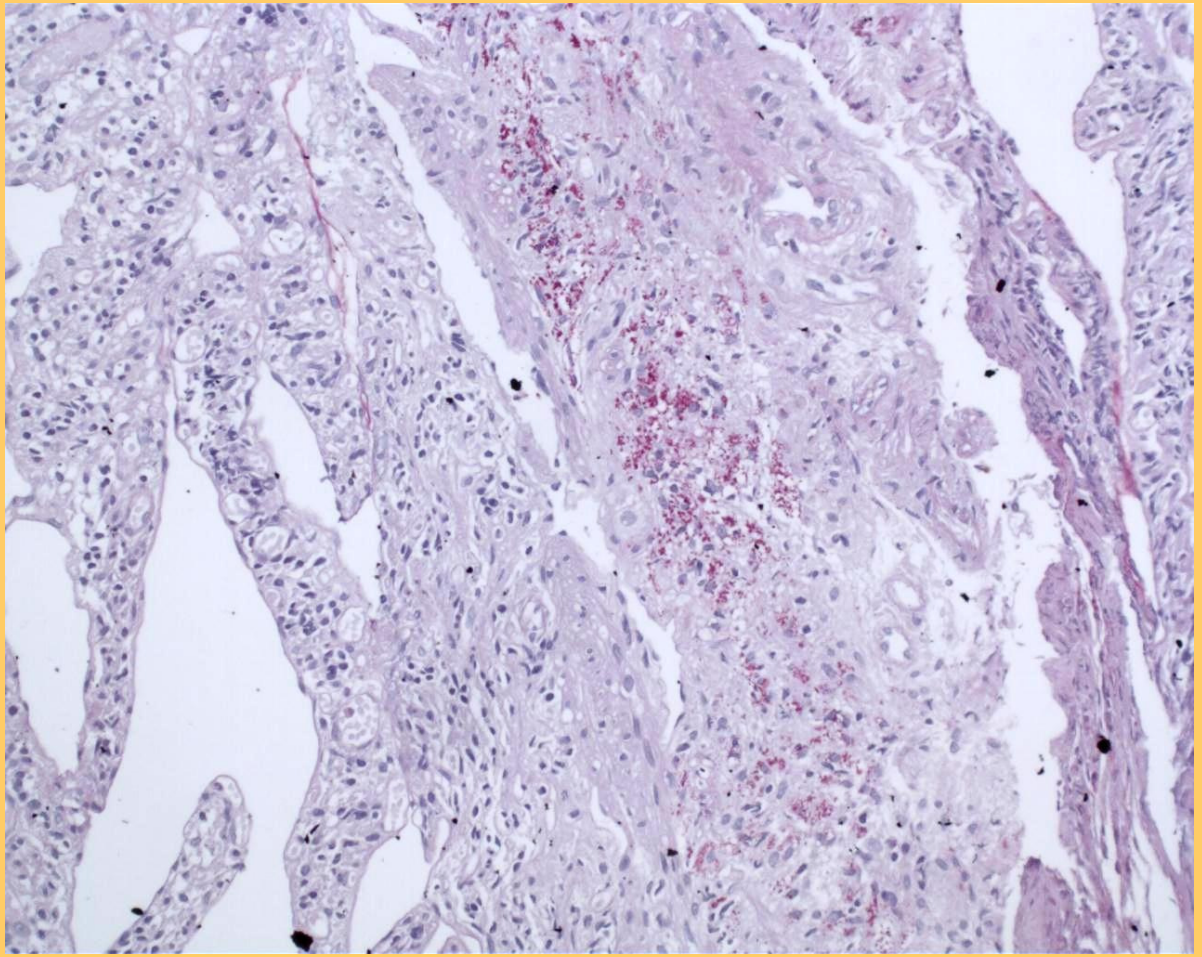
Conduta

- Sugerida a realização de EDA com biópsias duodenais para pesquisar Doença de Whipple.

Biópsia duodenal - PAS



Biópsia duodenal - PAS



Biópsia duodenal - PAS

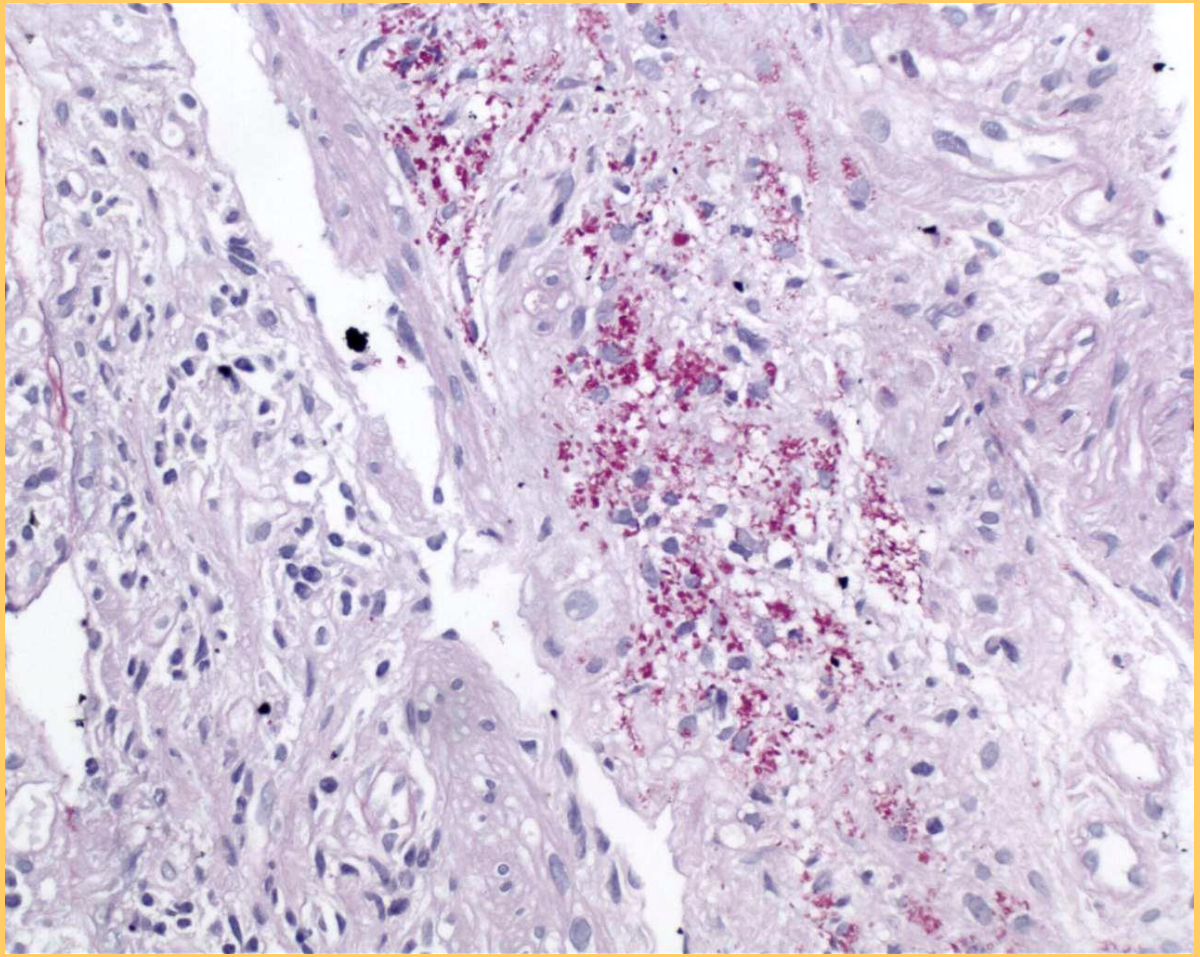


Table 1. Milestones in the History of Whipple's Disease and *Tropheryma whipplei*.

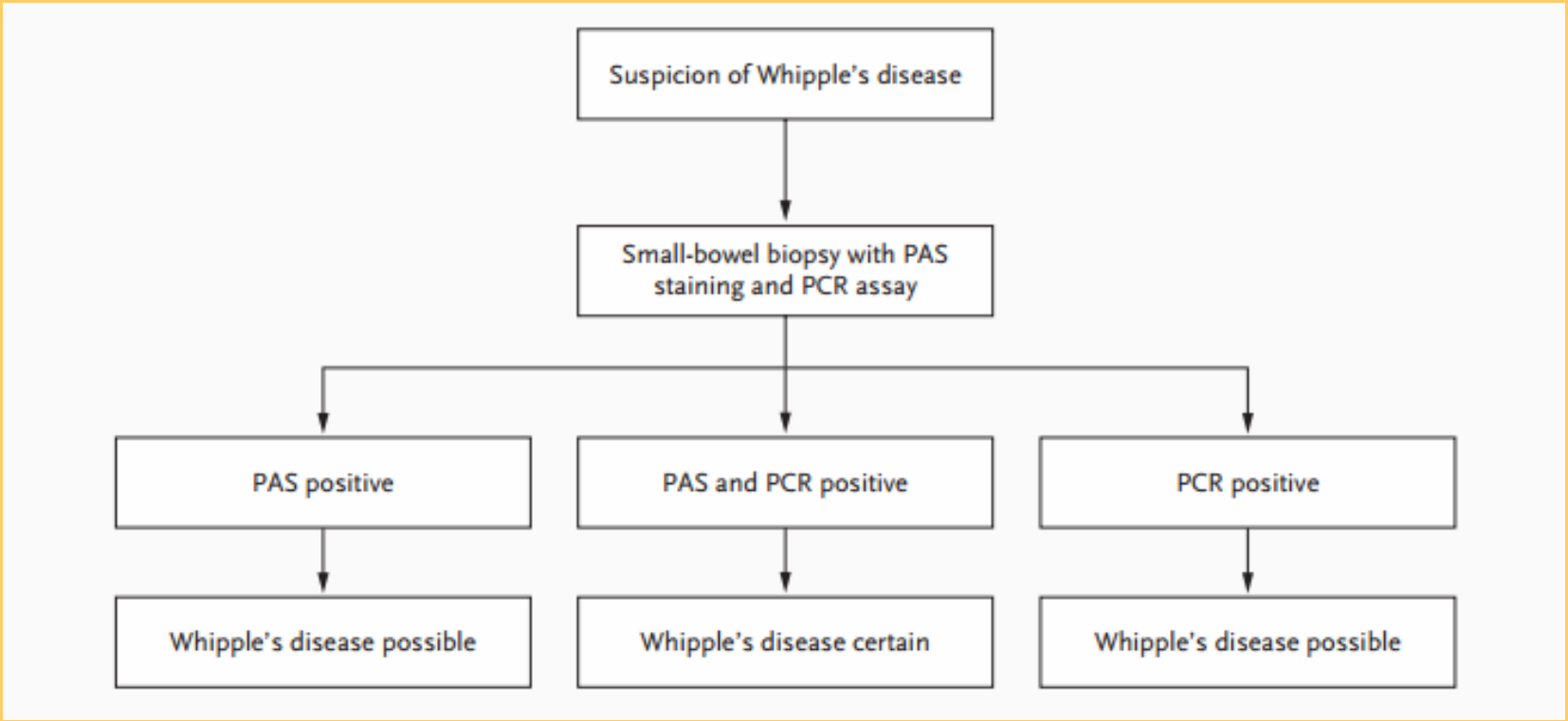
Date	Investigators	Advance
1907	Whipple[1]	First description of the disease
1947	Oliver-Pascual et al.[2]	First diagnosis before the death of a patient
1949	Black-Schaffer[3]	Development of periodic acid–Schiff staining for diagnosis
1952	Paulley[4]	First reported efficacy of antibiotic treatment
1961	Chears and Ashworth,[5] Yardley and Hendrix[6]	Detection of bacteria in macrophages by electron microscopy
1991	Wilson et al.[7]	Partial sequencing of 16S rRNA of an unknown bacterium
1992	Relman et al.[8]	Confirmation and extension of the 16S rRNA sequence; first naming of the bacterium: *T. whippelii*
2000	Raoult et al.[9]	First cultivation of the Whipple bacillus
2001	La Scola et al.[10]	First phenotypic characterization of the Whipple bacillus; renaming of the bacterium: *T. whipplei*
2003	Bentley et al.,[11] Raoult et al.[12]	Full sequencing of two genomes from two different strains of *T. whipplei*

Fenollar, F., Puéchal, X., Raoult, D. (January 2007). "Whipple's disease". New England Journal of Medicine 356 (1): 55–66

Lesões de pele na doença de Whipple

- Dois tipos: não específica e específica

1. Não específica: relacionadas a quadro de malnutrição e deficiências nutricionais (hiperpigmentação cutânea, petéquias, púrpura e hiperqueratose);
2. Específica: devido à ação direta dos microorganismos ou à reação secundária aos antígenos bacterianos (tipo eczematoso, lúpus-símile, dermatomiosite-símile, urticariforme, vasculítico, EN-símile e tipo nódulo reumatóide).



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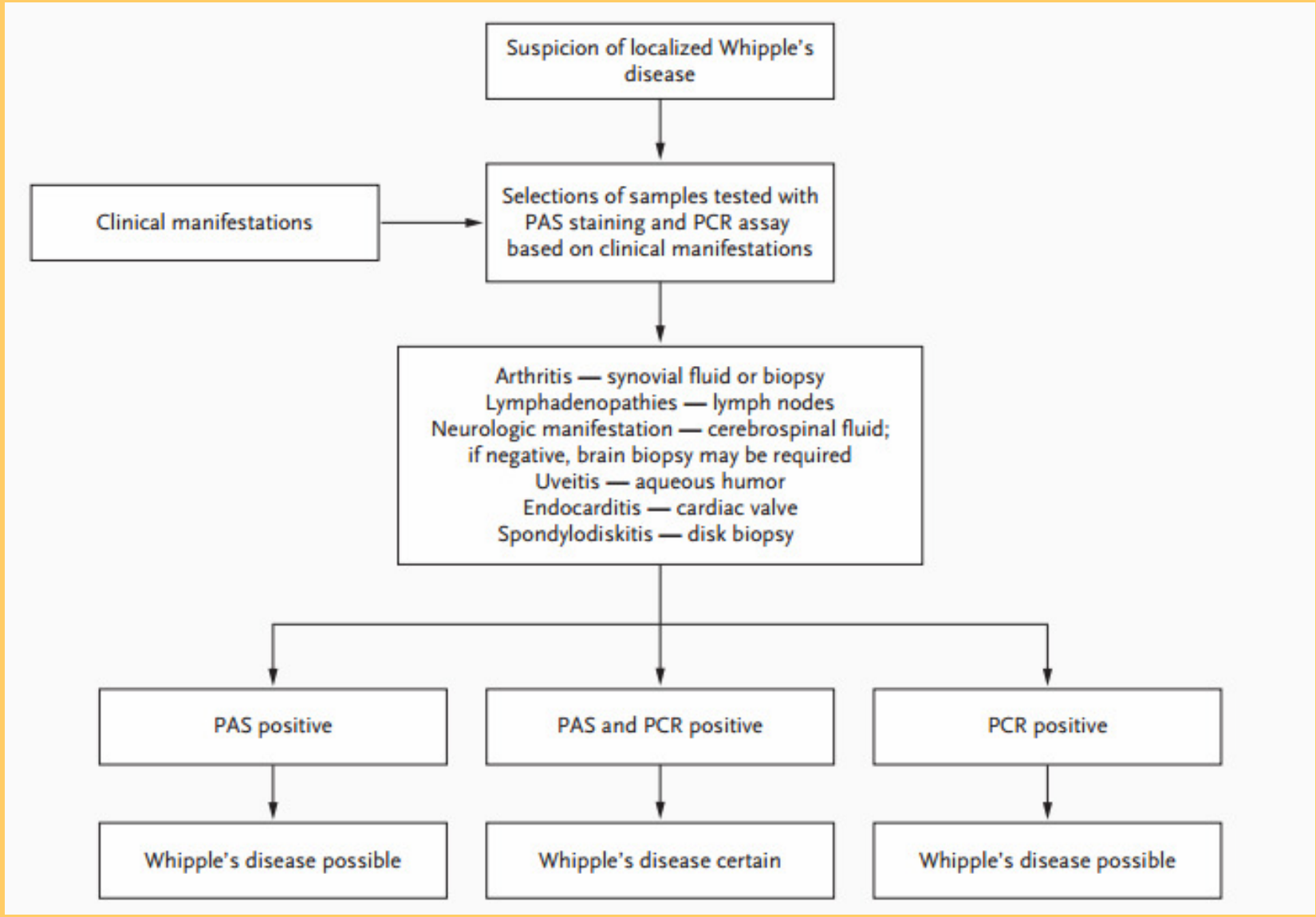
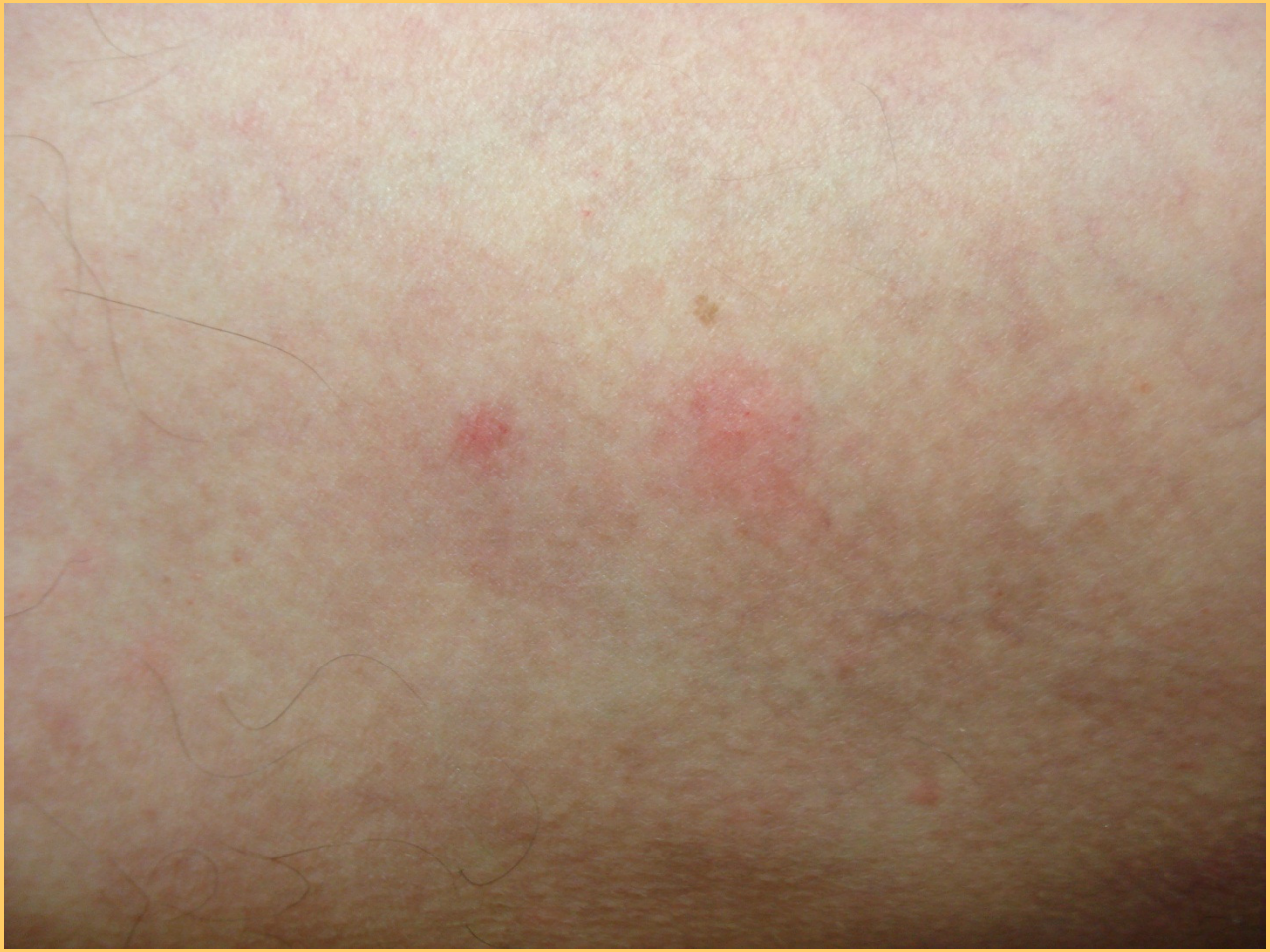
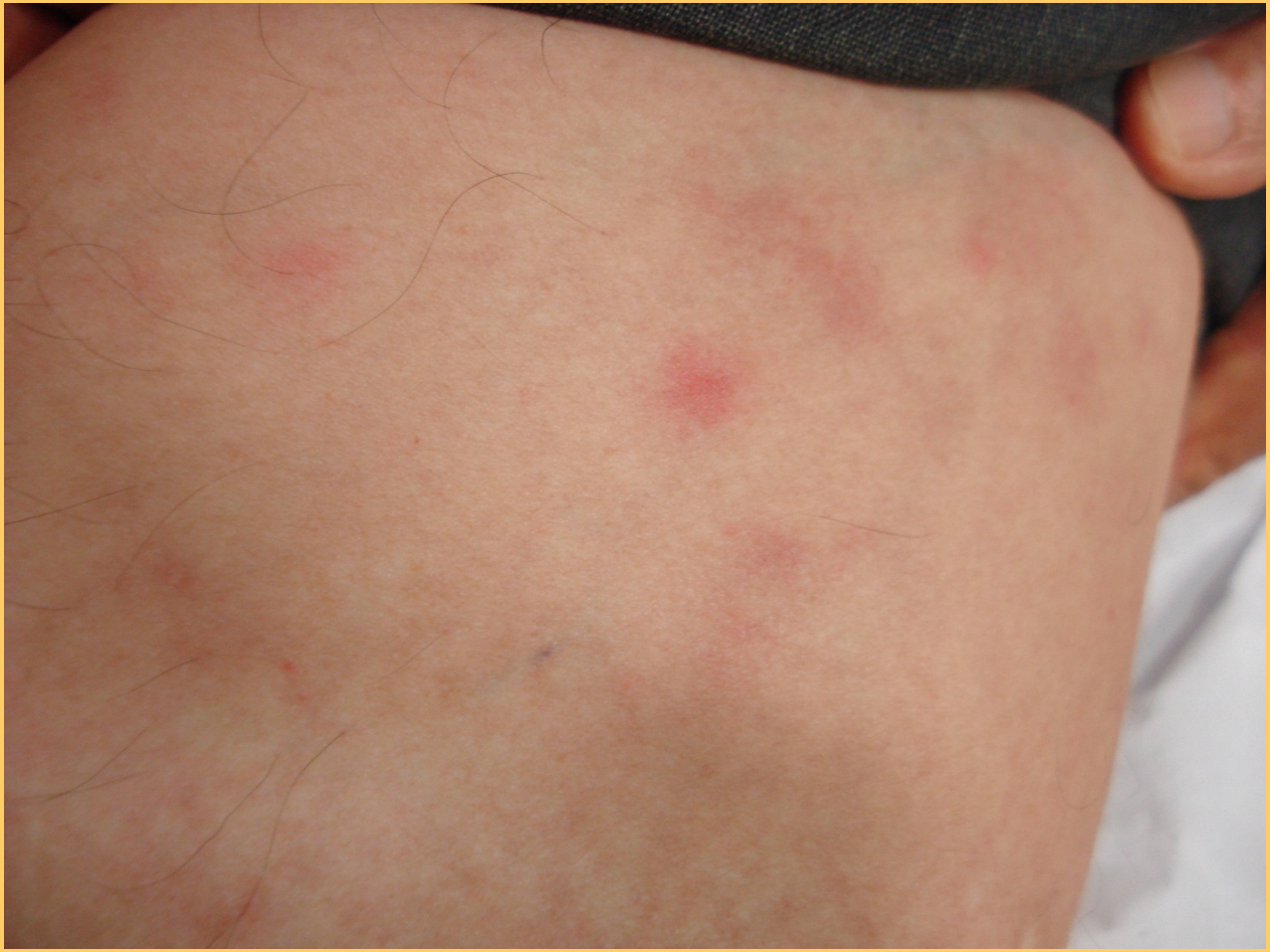


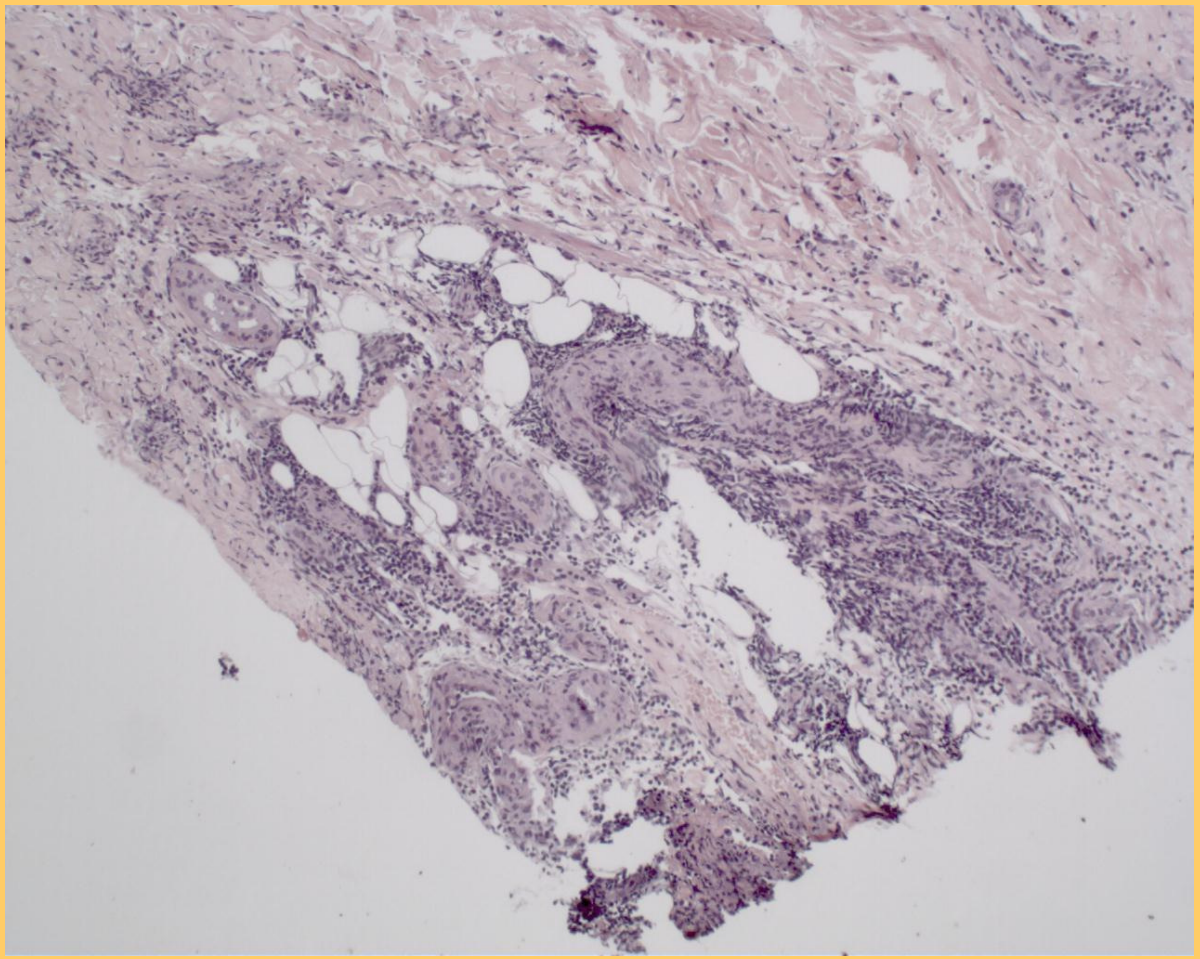
Fenollar, F., Puéchal, X., Raoult, D. (January 2007). "Whipple's disease". New England Journal of Medicine 356 (1): 55–66

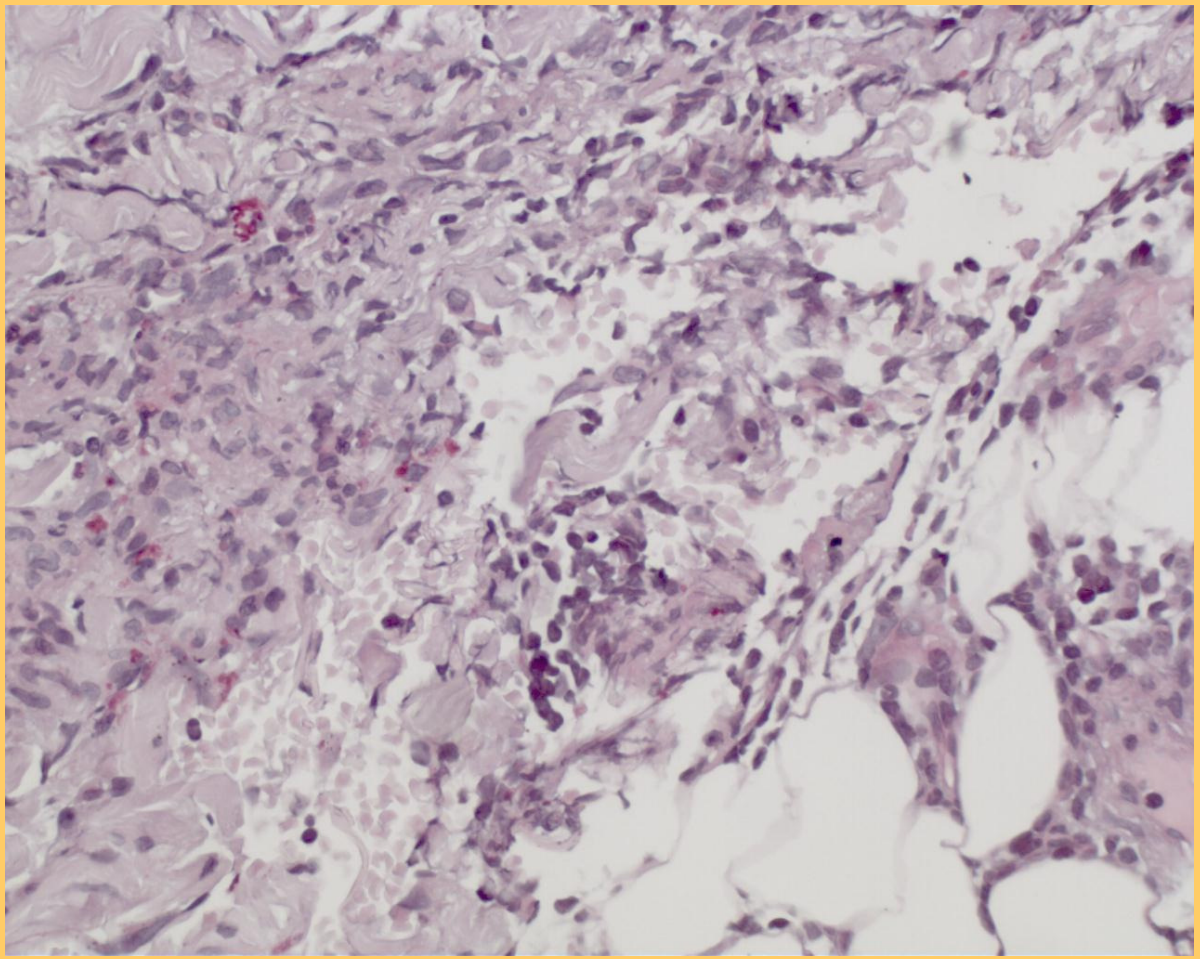
Evolução

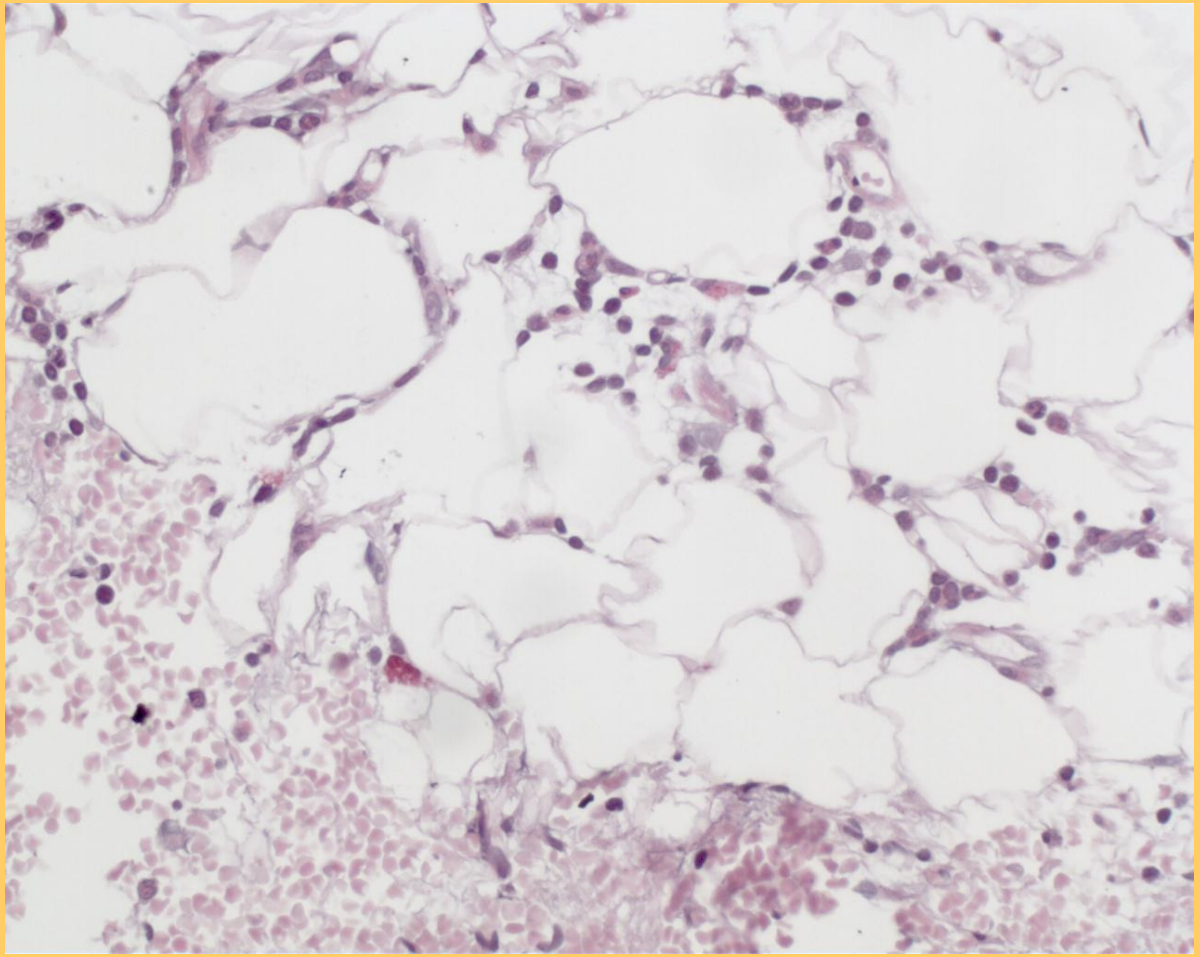
- Fez tratamento com sulfametoxazol + trimetoprima (160 mg e 800mg) 2x dia por 1 a 2 anos;
- Apresentou melhora significativa com ganho ponderal (6 kg em 4 meses);
- Apareceu após 4 meses de tratamento, pápulas e nódulos eritematosos, pouco dolorosos e não pruriginosos, localizados nas coxas, perna, braço e tronco.

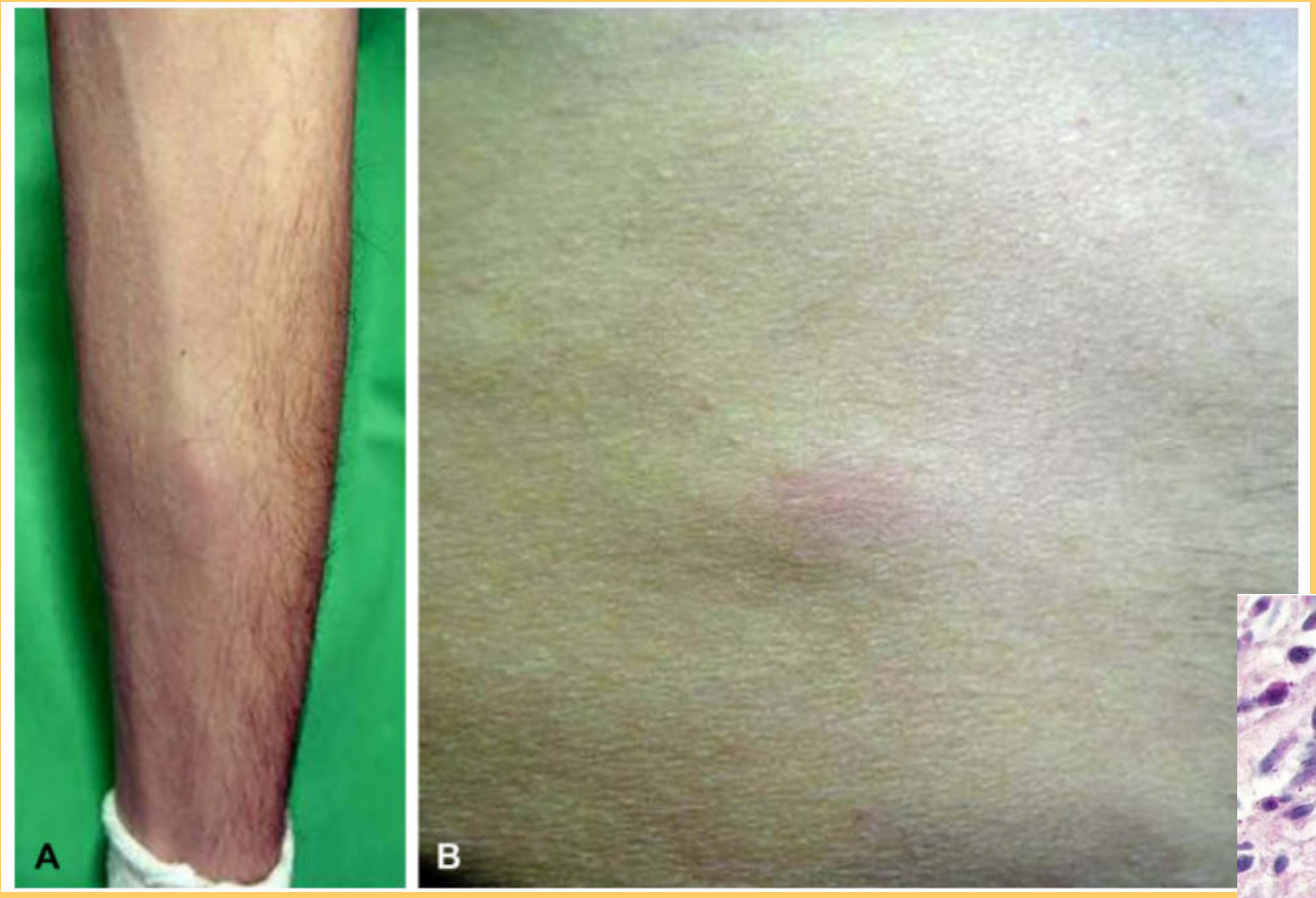


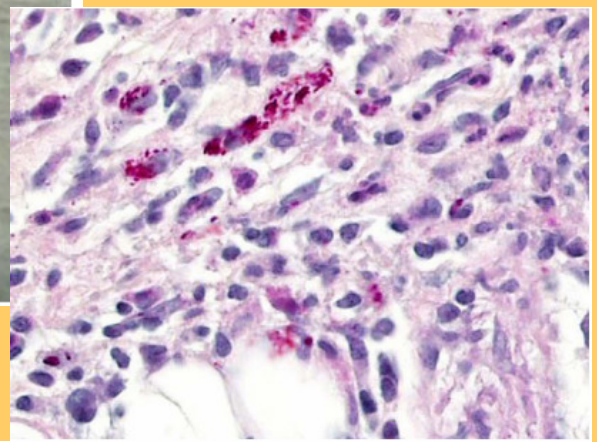












J Am Acad Dermatol. 2009 Feb;60(2):277-88. doi: 10.1016/j.jaad.2008.09.024.

Erythema nodosum-like lesions in treated Whipple's disease: signs of immune reconstitution inflammatory syndrome.

Schaller J, Carlson JA.

Department of Dermatohistology, Catholic Clinics, Duisburg, Germany.

Am J Dermatopathol. 2012 Apr;34(2):182-7. doi: 10.1097/DAD.0b013e318221ba55.

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J Infect. 2010 Sep;61(3):266-9. doi: 10.1016/j.jinf.2010.06.007. Epub 2010 Jun 19.

Tropheryma whipplei in the skin of patients with classic Whipple's disease.

Angelakis E, Fenollar F, Lepidi H, Birg ML, Raoult D.

Université de la Méditerranée, Unité des Rickettsies, URMITE CNRS-IRD UMR 6236, Faculté de Médecine, 27 Bd Jean Moulin, 13385 Marseille Cedex 05, France.

J Cutan Pathol. 2001 Aug;28(7):368-70.

Subcutaneous nodules in Whipple's disease.

Tarroch X, Vives P, Salas A, Moré J.

Servicio de Anatoía Patológica, Hospital Mútua de Terrassa, Terrassa, Barcelona, Spain. 2415xts@comb.es

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Obrigado!

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