

HOSPITAL FEDERAL DA LAGOA
Serviço de Dermatologia
Chefia: João Carlos Regazzi Avelleira

SÍNDROME DE BROOKE-SPIEGLER

Anna Clara dos Santos da Costa
Gabriela Menegol Bassani
Flavia de Freire Cassia
Juliana Marques da Costa

ANAMNESE

- **ID:** Feminino, 54 anos, natural de Japeri, RJ;
- **HDA:** Lesões sobrelevadas, assintomáticas, na face, tronco e membros superiores, iniciadas aos 14 anos de idade, com aumento progressivo no número e tamanho;
- **HPP:** Negou morbidades. Negou etilismo ou tabagismo;
- **HF:** Mãe, todas irmãs e filhas com lesões similares. Seu irmão não apresenta lesões.









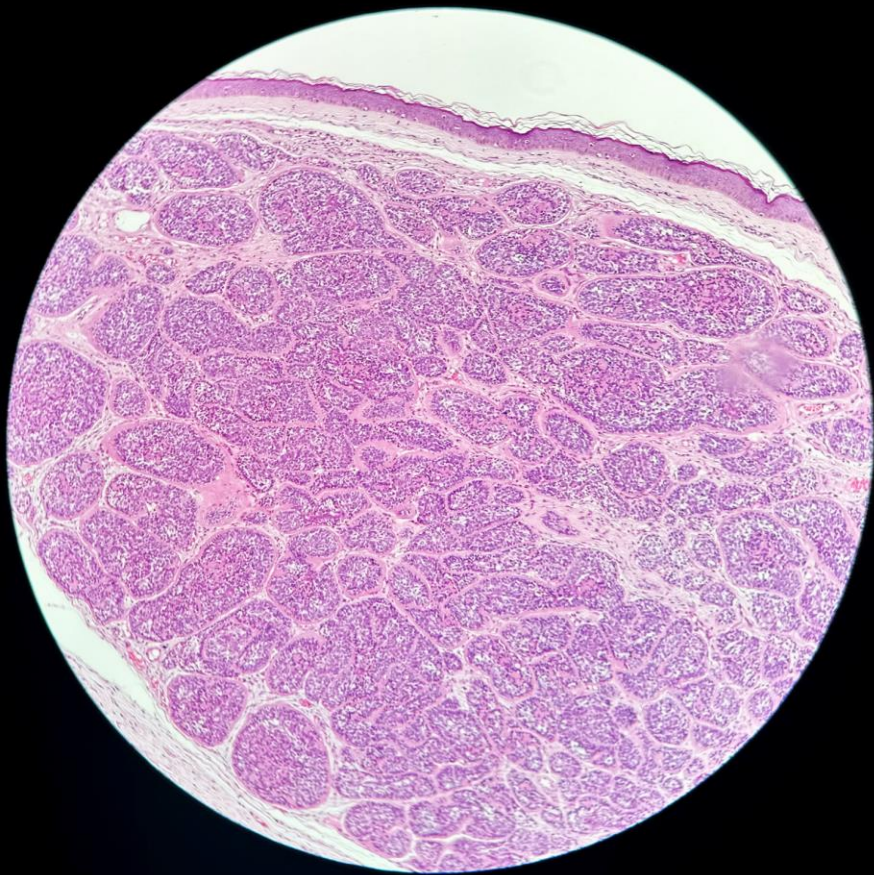
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Exérese

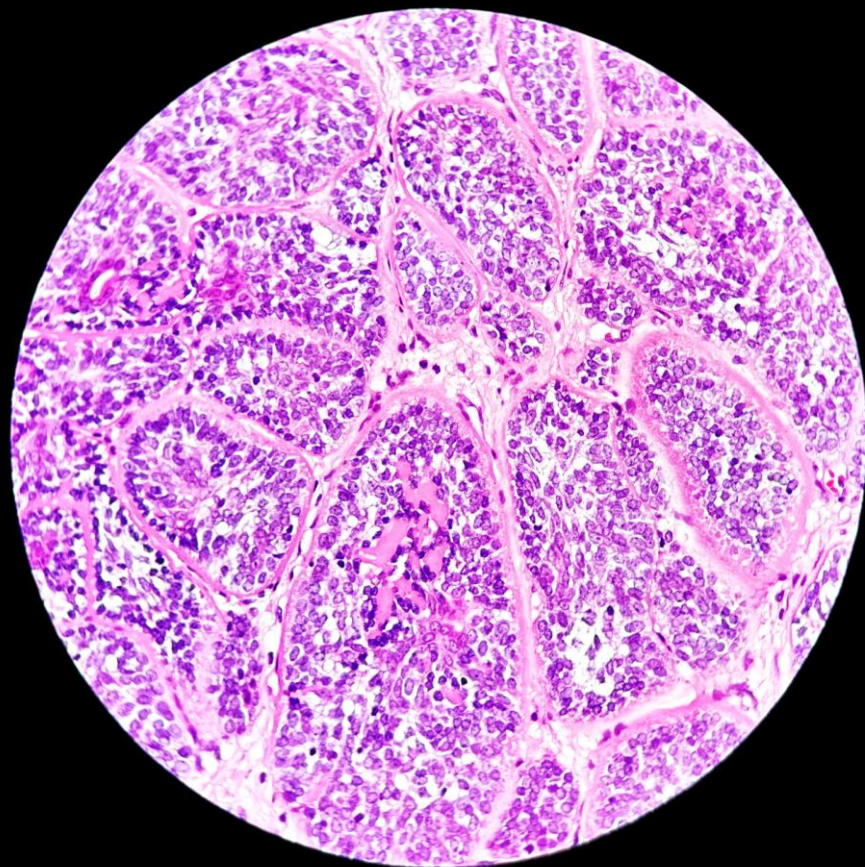


Nódulo eritematoso com
telangiectasias na superfície

HE, 100X



HE, 400X



CONDUTA

Exérese



CILINDROMA

Nódulo eritematoso com telangiectasias na superfície

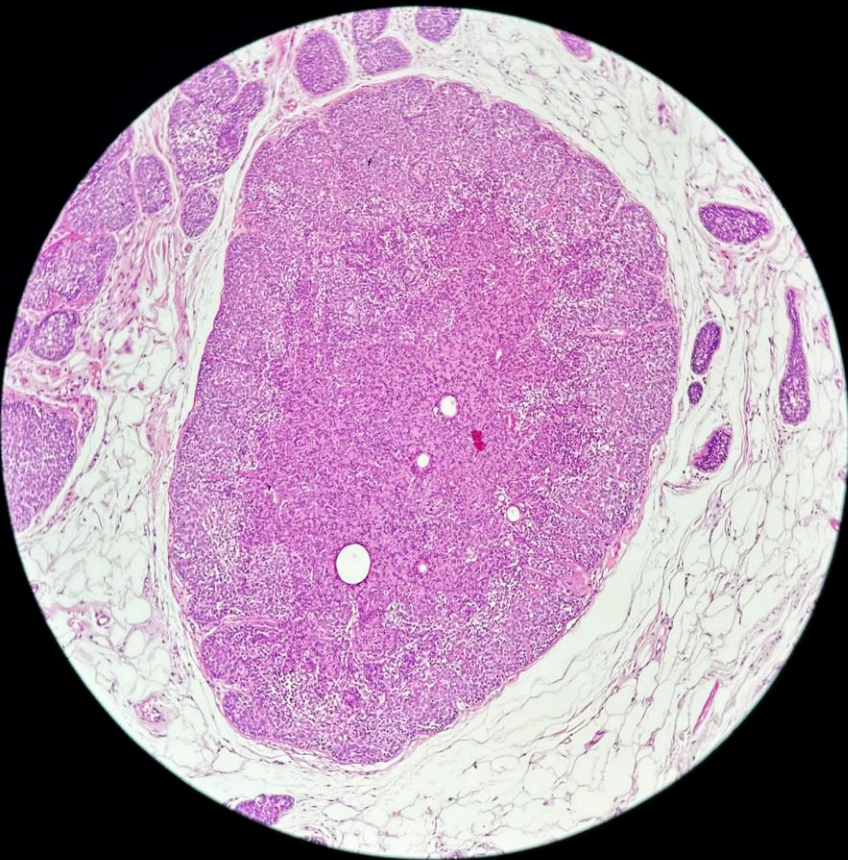
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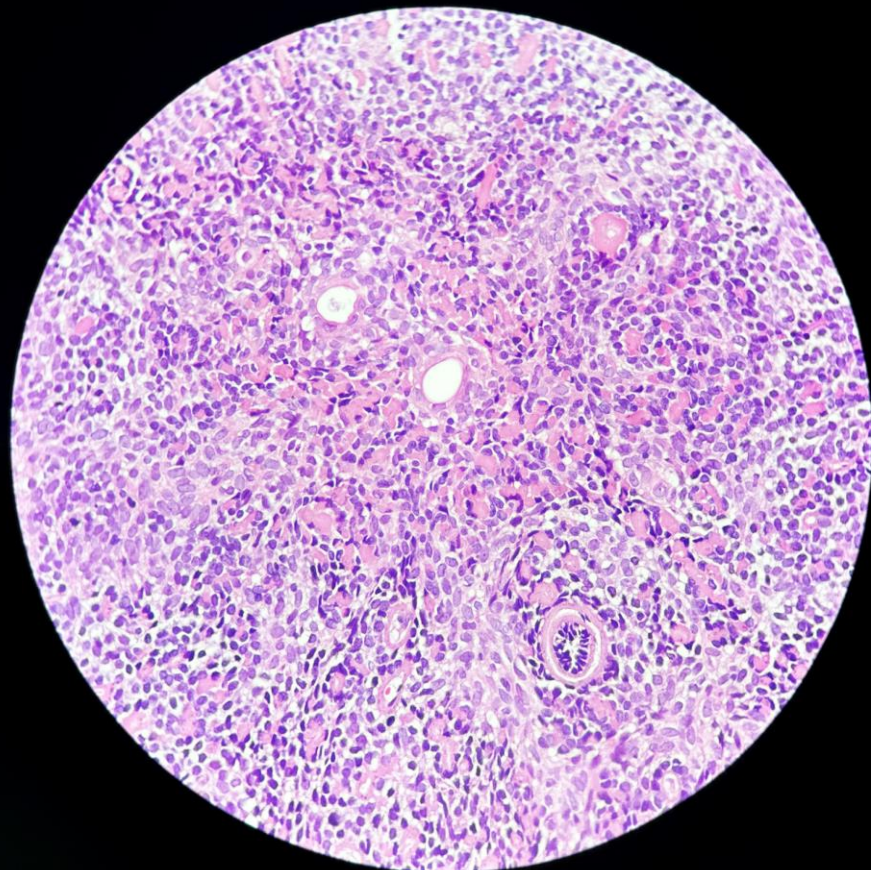


Pápula normocrômica no
dorso

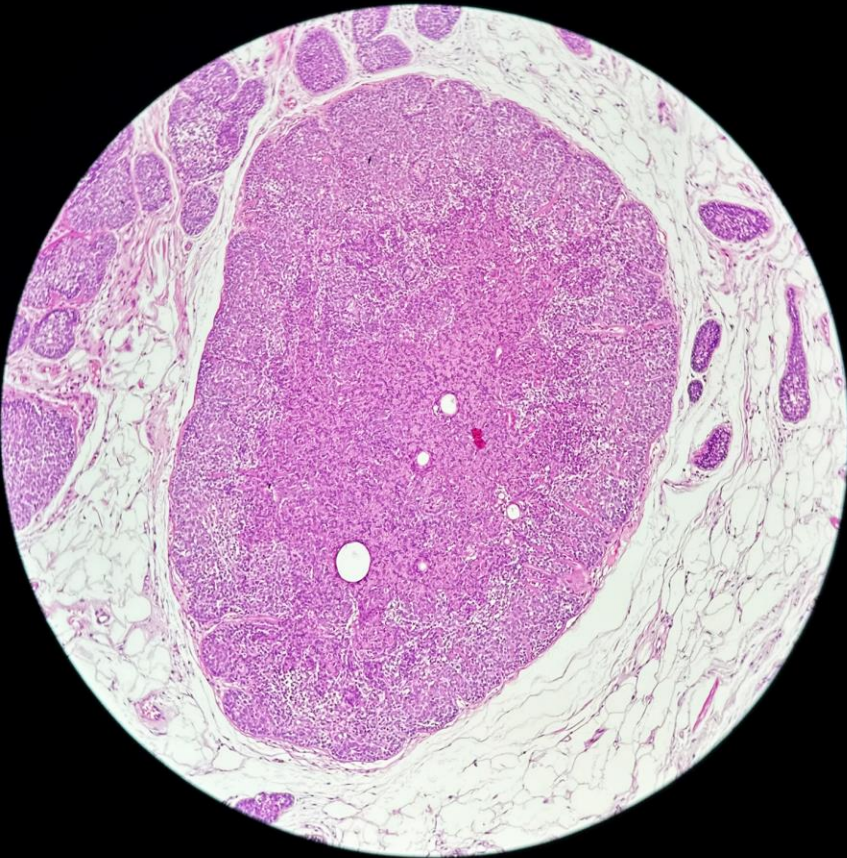
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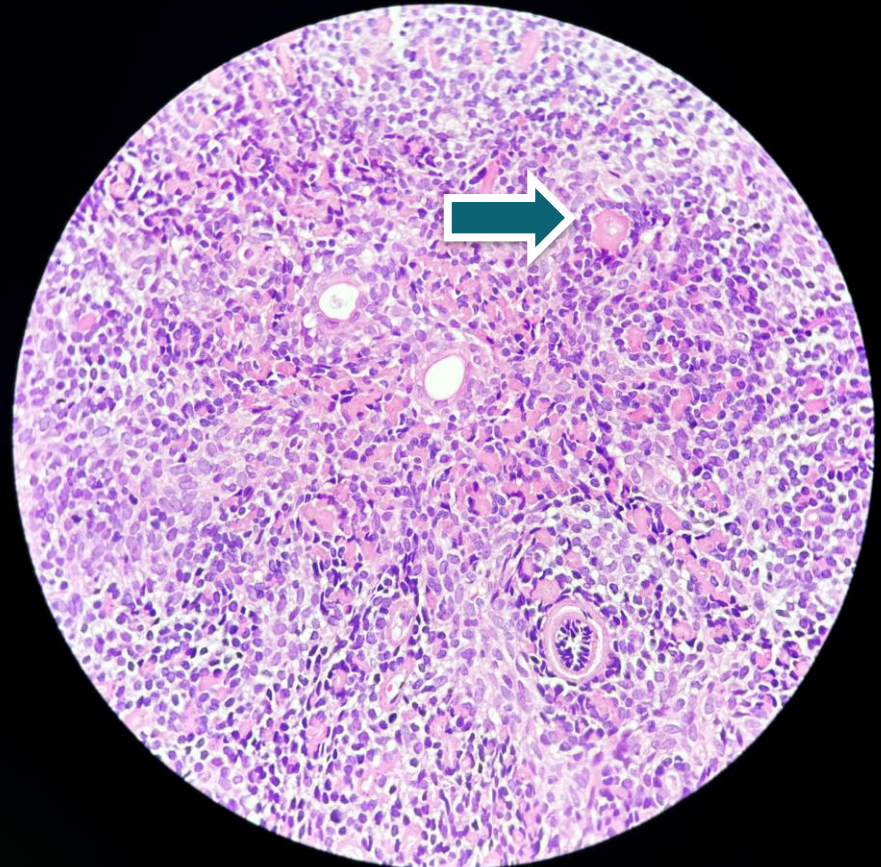
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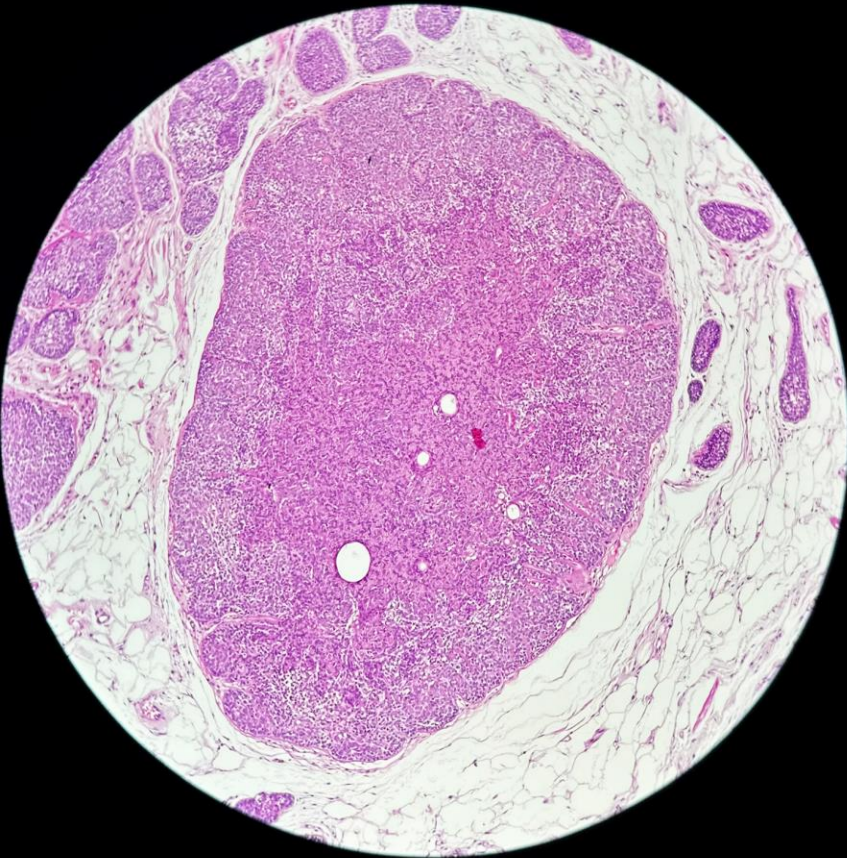
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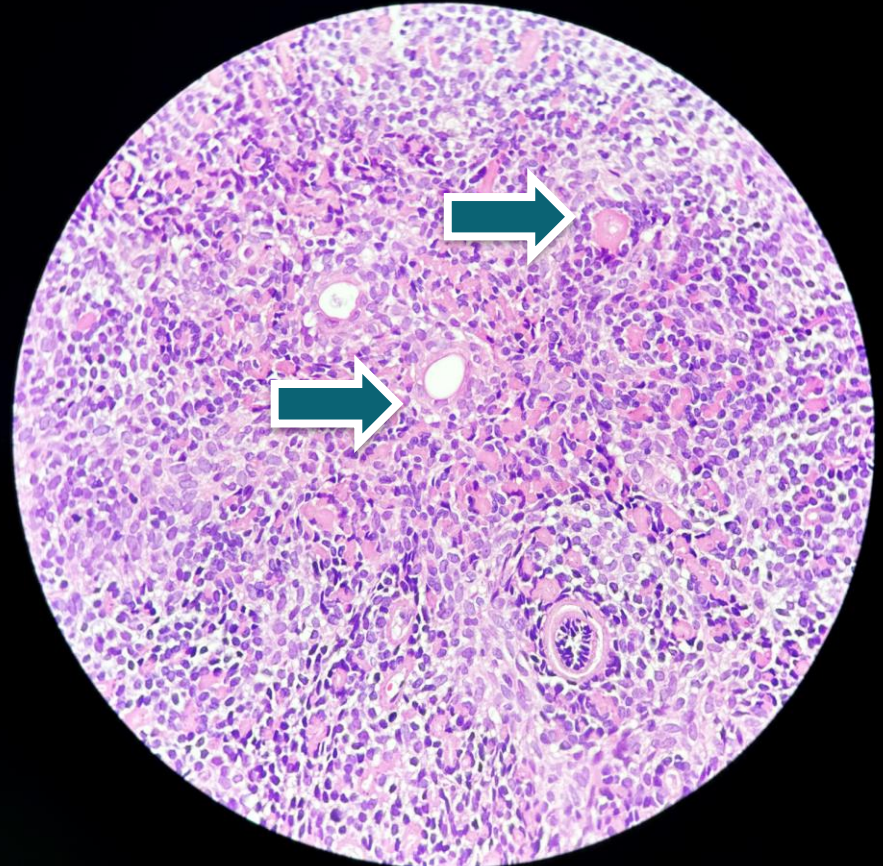
HE, 400X



HE, 100X



HE, 400X



CONDUTA

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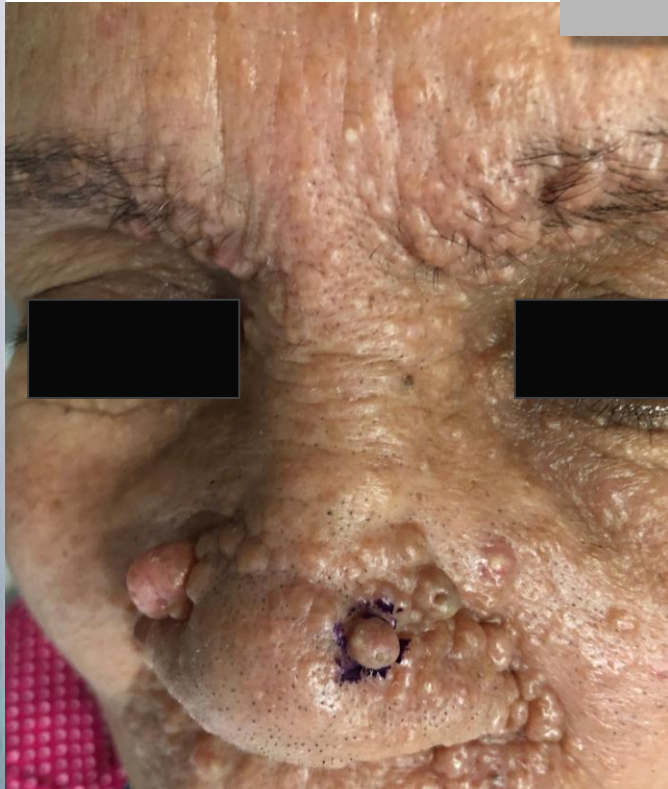


ESPIROADENOMA ÉCRINO

Pápula normocrômica no
dorso

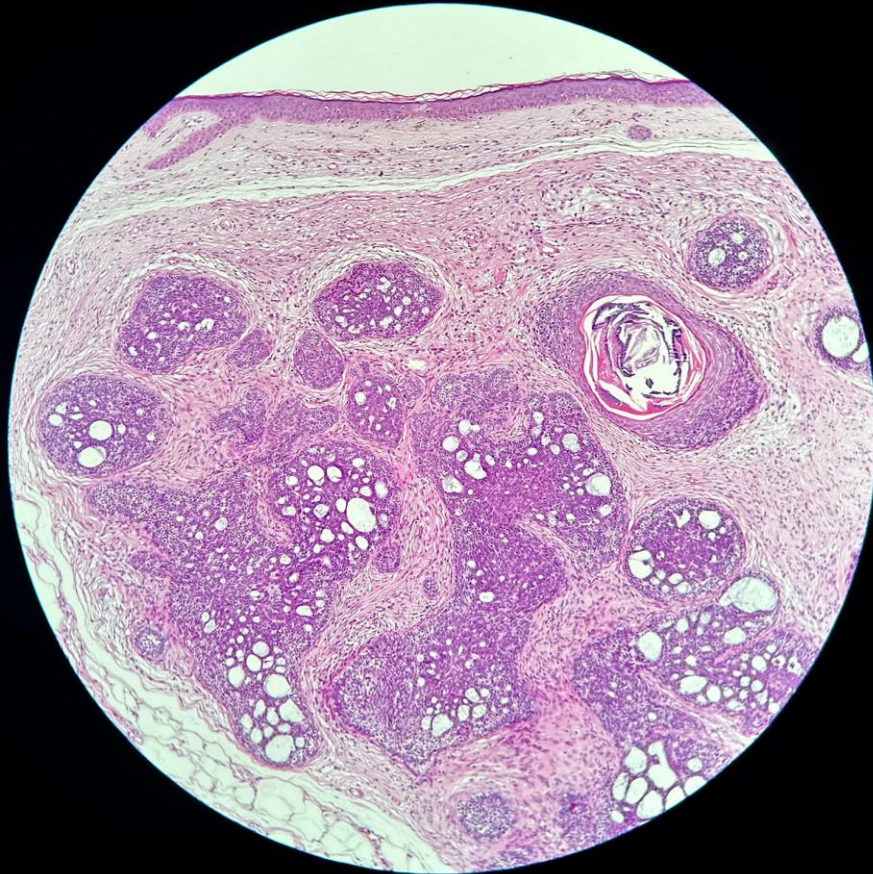
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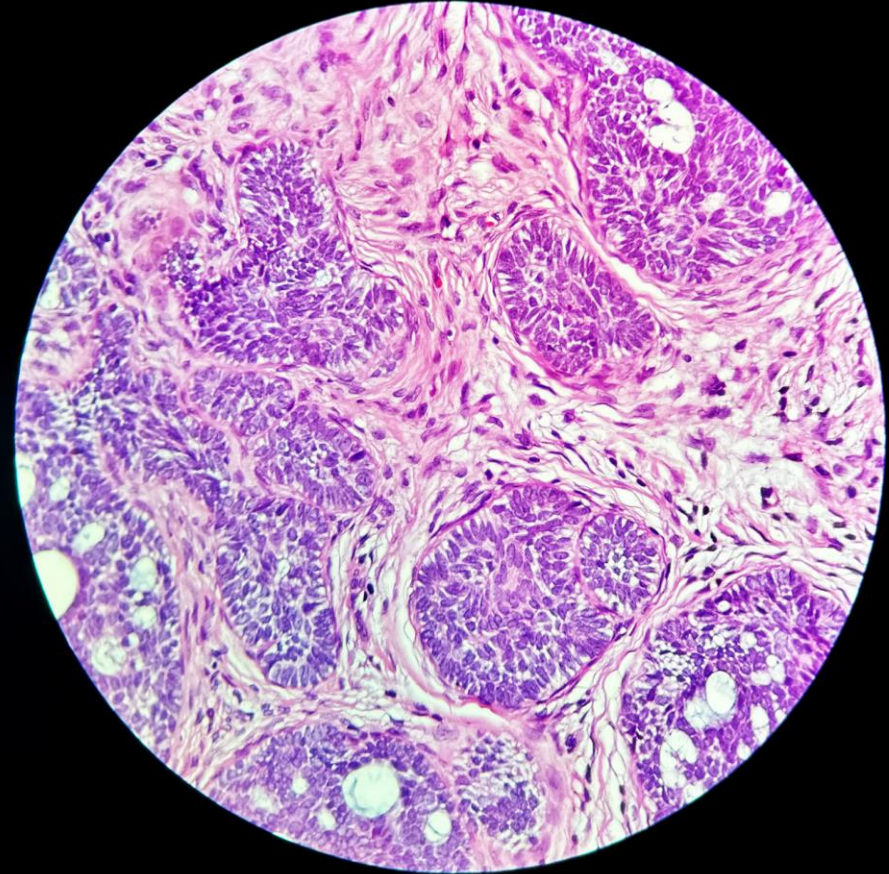


Pápula normocrômica na
região nasal

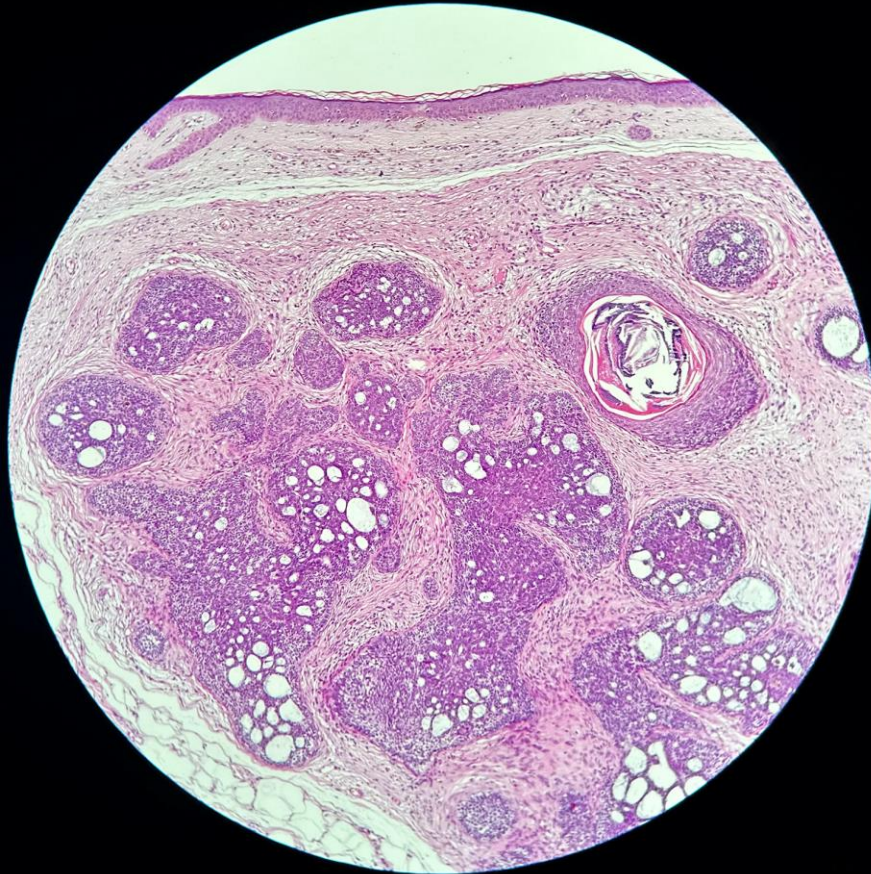
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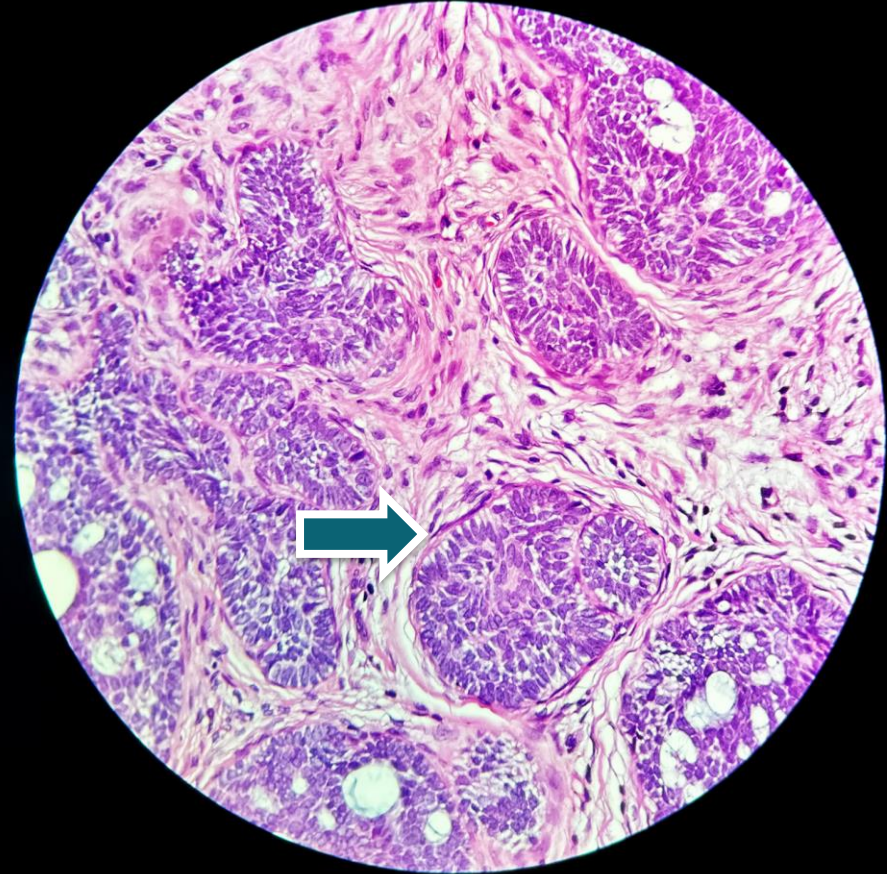
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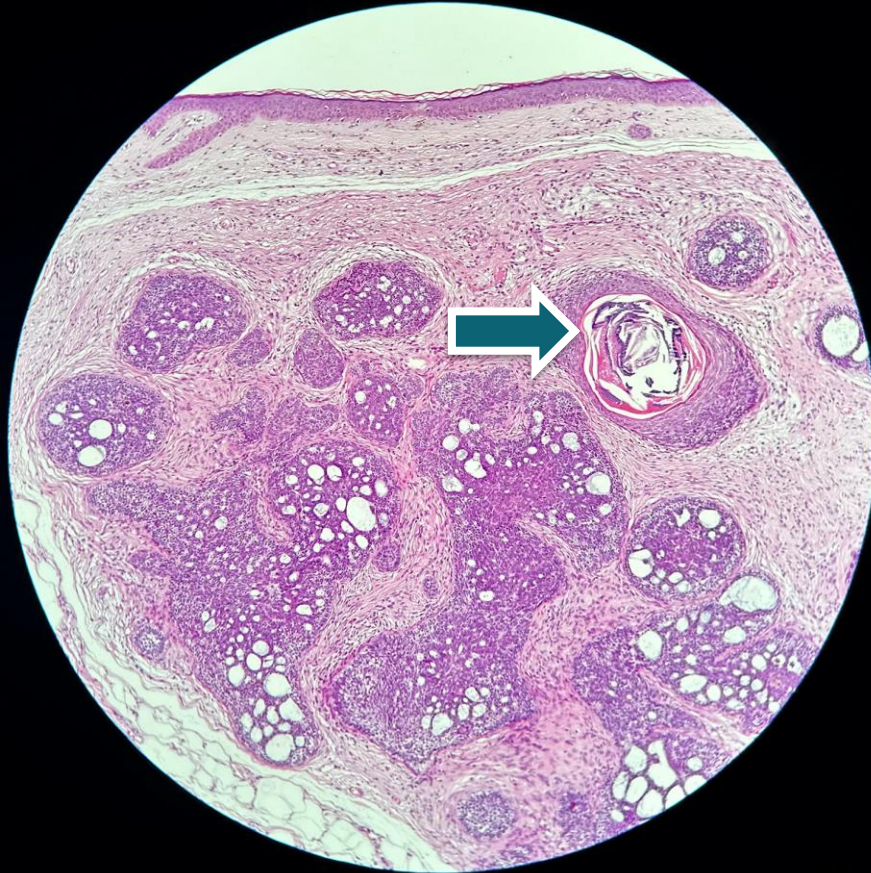
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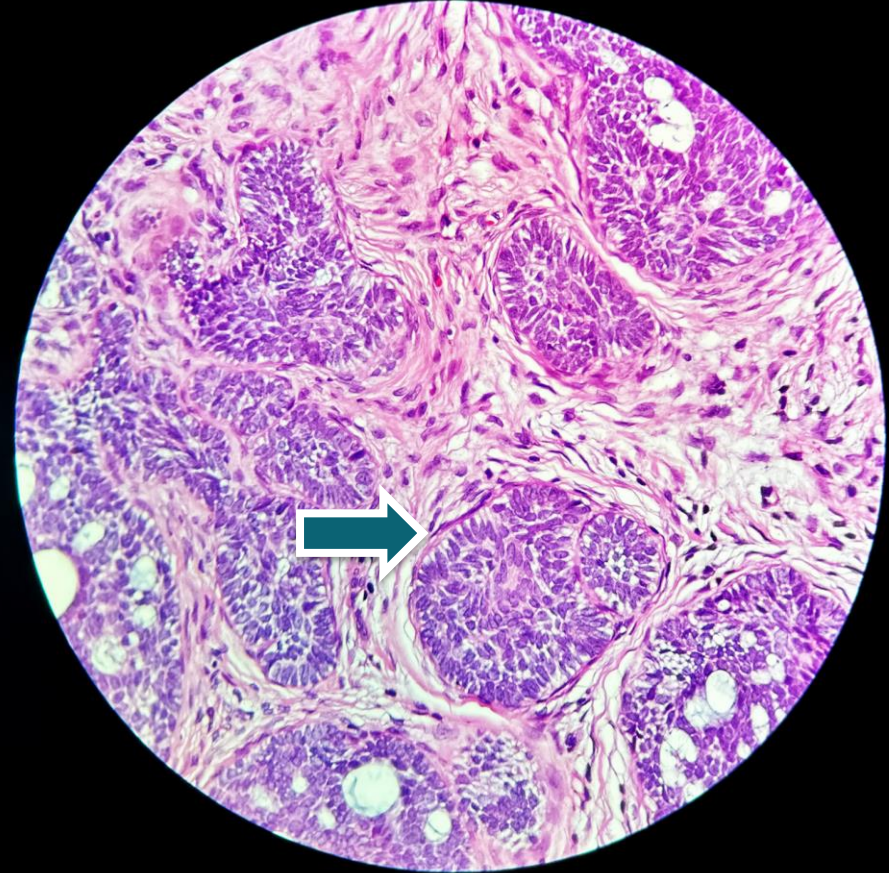
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HE, 100X

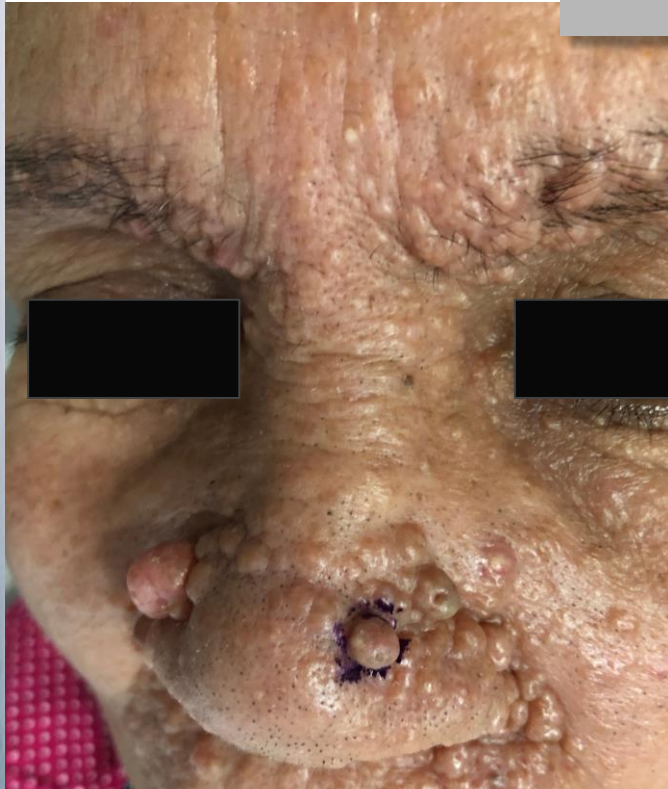


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CONDUTA

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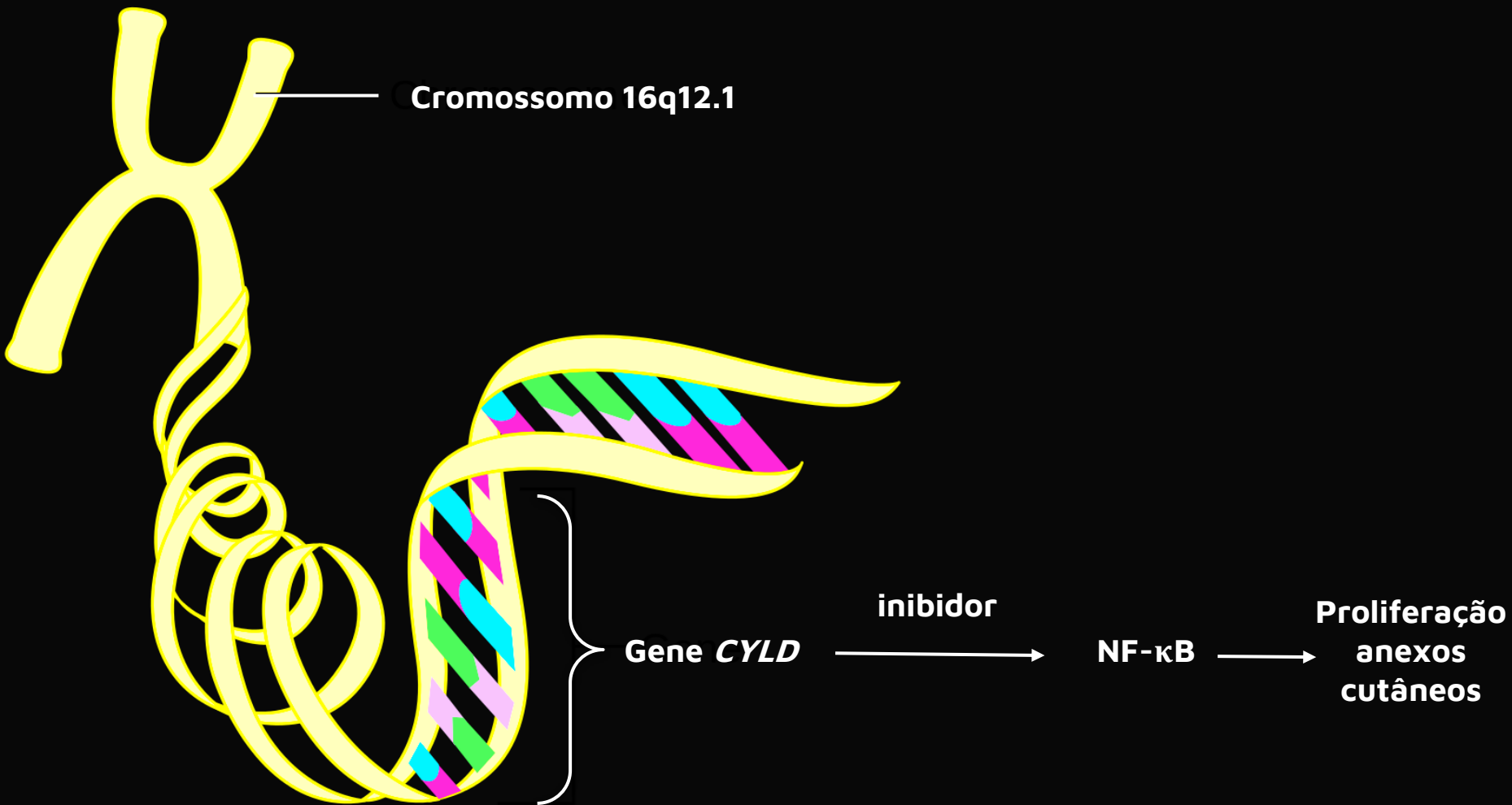


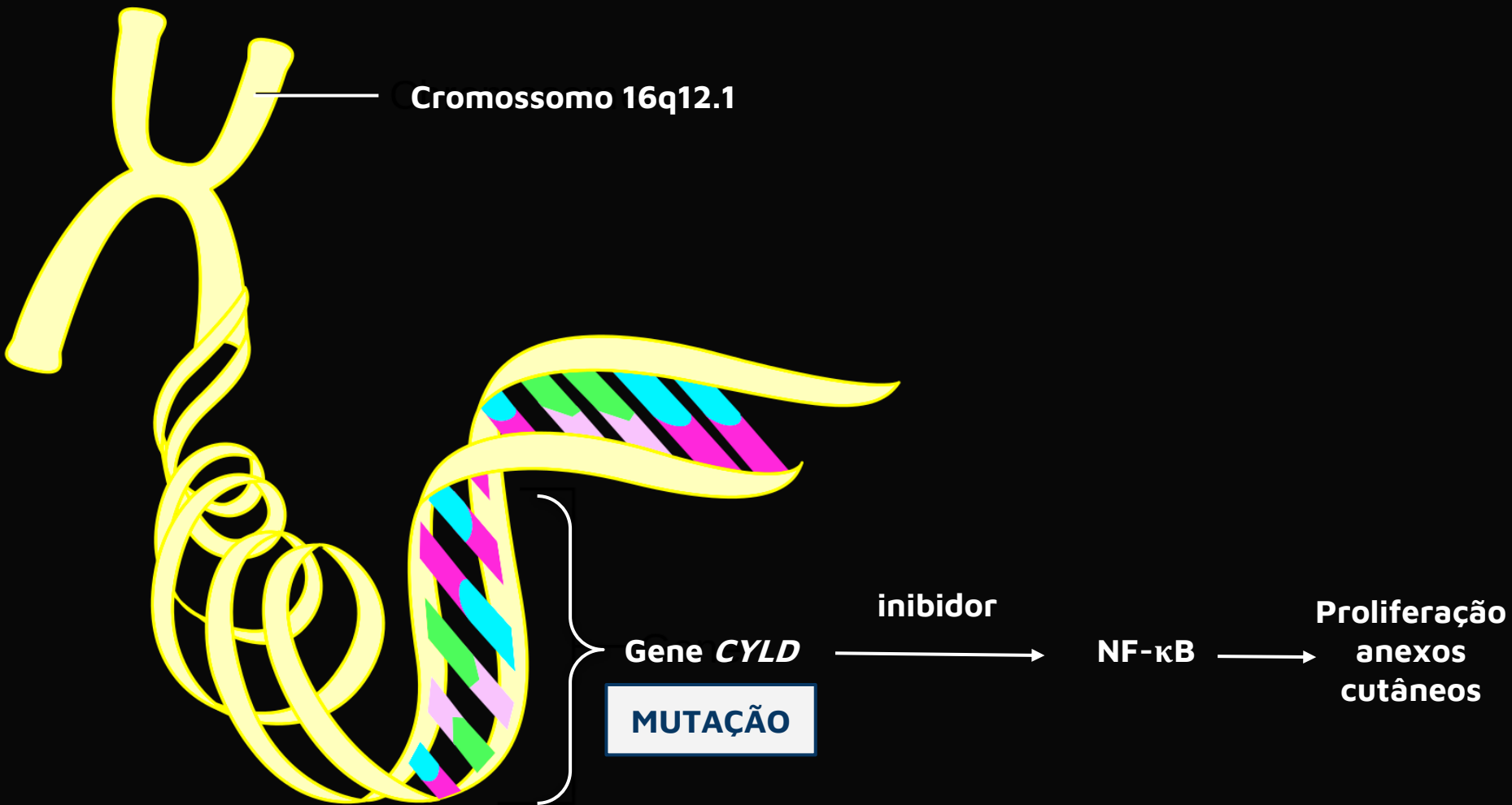
TRICOEPITELIOMA

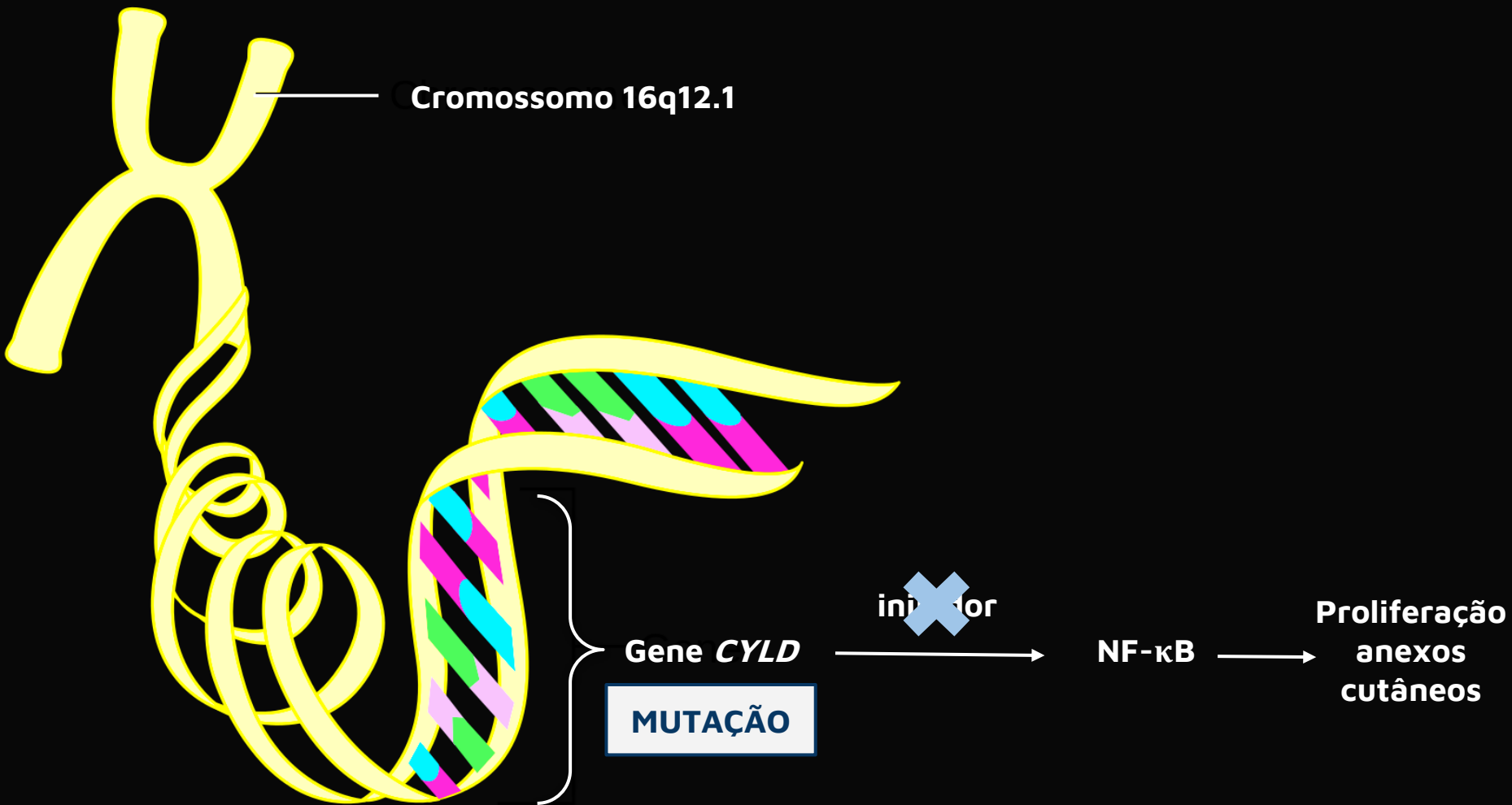
Pápula normocrômica na
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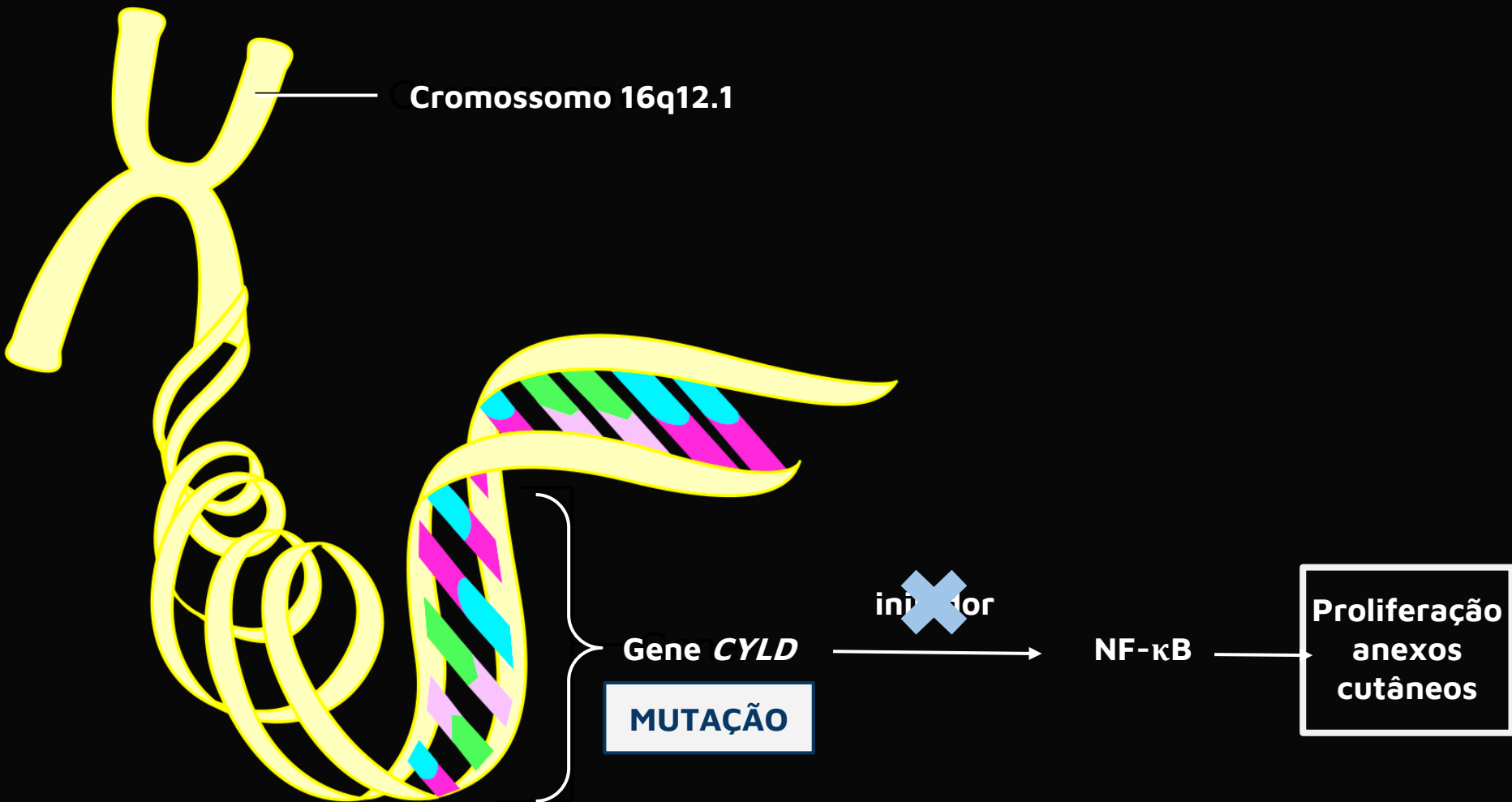
SÍNDROME DE BROOKE-SPIEGLER

Rara, autossômica dominante, mutação gene *CYLD*









SÍNDROME DE BROOKE-SPIEGLER

Rara, autossômica dominante, mutação gene *CYLD*

10-20 anos

Surgimento de neoplasias de anexos cutâneos

CILINDROMAS, TRICOEPITELIOMAS E ESPIROADENOMAS

SÍNDROME DE BROOKE-SPIEGLER

Surgimento de neoplasias de anexos cutâneos

CILINDROMAS, TRICOEPITELIOMAS E ESPIROADENOMAS

5-10% casos transformação maligna:

- Carcinoma Basocelular (CBC), cilindrocarcinoma, espiroadenocarcinoma

Rara associação com outras neoplasias:

- Adenoma e adenocarcinoma de parótida
- Cilindromatose pulmonar

CLÍNICA

Pápulas, nódulos, tumorações normocrômicas, eritematosas ou amareladas

Tamanho em média de 0,5 a 3cm, podendo ser maiores

Dezenas a centenas

Localização típica: cabeça e pescoço

CLÍNICA



Neema S et al.. Brooke-Spiegler syndrome. Indian J Dermatol Venereol Leprol 2023.



Trufant J et al Brooke-Spiegler syndrome. Dermatol Online J. 2012.

DIAGNÓSTICO

CILINDROMA

- Pápulas ou nódulos eritematosos
- Couro cabeludo

TRICOEPI TELIOMA

- Pápulas ou nódulos normocrômicos ou amarelados
- Centrofacial

ESPIROADENOMA

- Pápulas ou nódulos normocrômicos ou eritematosos
- Tronco, extremidades

DIAGNÓSTICO

CARACTERÍSTICAS CLÍNICAS SIMILARES E INESPECÍFICAS

CILINDROMA

- Pápulas ou nódulos eritematosos
- Telangiectasias
- Couro cabeludo

TRICOEPITELIOMA

- Pápulas ou nódulos normocrômicos ou amarelados
- Centrofacial

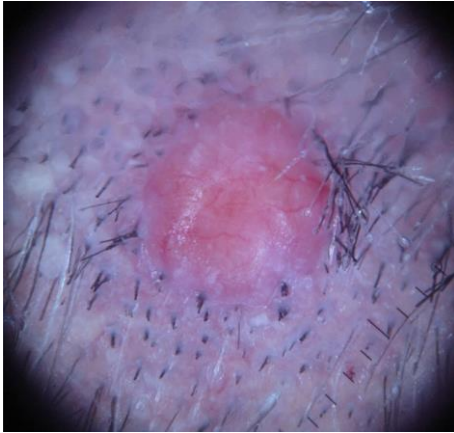
ESPIROADENOMA

- Pápulas ou nódulos normocrômicos ou eritematosos
- Tronco, extremidades

Histopatológico necessário na diferenciação

DERMATOSCOPIA

CILINDROMA



- Vasos arboriformes

TRICOEPI TELIOMA



- Vasos arboriformes
- Glóbulos amarelados

ESPIROADENOMA



- Vasos arboriformes
- Glóbulos azulados

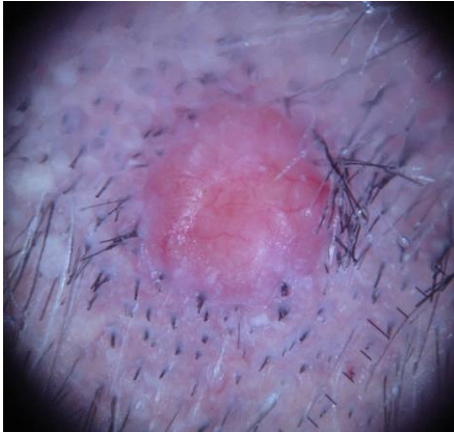
-Cohen YK et al. Dermatoscopic pattern of a cylindroma. Dermatol Pract Concept. 2014;

-Navarrete-Dechent C, Bajaj S, Marghoob AA, González S, Muñoz D. Multiple familial trichoepithelioma: confirmation via dermoscopy. Dermatol Pract Concept. 2016;

-Tschandl P. Dermatoscopic pattern of a spiradenoma. Dermatol Pract Concept. 2012

DERMATOSCOPIA

CILINDROMA



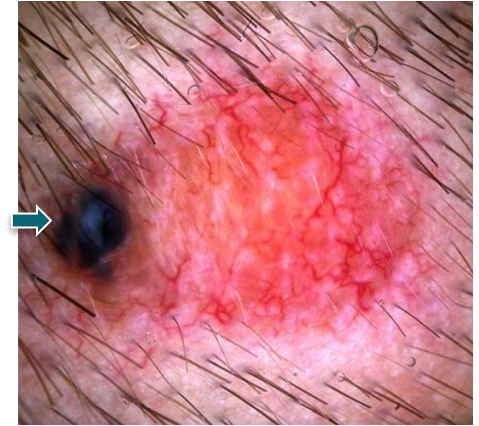
- Vasos arboriformes

TRICOEPI TELIOMA



- Vasos arboriformes
- Glóbulos amarelados

ESPIROADENOMA



- Vasos arboriformes
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TRATAMENTO

EXCISÃO

ELETROCIRURGIA

CRIOCIRURGIA

LASERTERAPIA

**ACONSELHAMENTO
GENÉTICO**

**ACOMPANHAMENTO
CLÍNICO-DERMATOLÓGICO:**

- Associação com outras neoplasias
- Transformação maligna

MOTIVO DA APRESENTAÇÃO

Demonstrar caso raro e
exuberante da Síndrome
de Brooke-Spiegler

BIBLIOGRAFIA:

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Obrigada!