



# August Case of the Month

- A 30-year-old female presents to her follow-up visit with Dermatology with a chief complaint of an **itchy painful lesion** on her right **frontal scalp**.
  - Exam revealed a 6 cm x 2cm pink, firm, nodular plaque with several scattered terminal hairs
- PMH:
  - Hx of **tuberous sclerosis complex (TSC)**, renal transplant, and cardiovascular disease
- FH:
  - Parkinsons disease (mother)
- Pertinent Medications:
  - patient is on multiple drugs for renal transplant and cardiovascular disease
- What is your diagnosis?

# Fibrous Cephalic Scalp Plaque: a Manifestation of Tuberous Sclerosis Complex

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# History of Present Illness

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PMH: Hx of **tuberous sclerosis complex (TSC)**, renal transplant, and cardiovascular disease

FH: Parkinsons disease (mother)

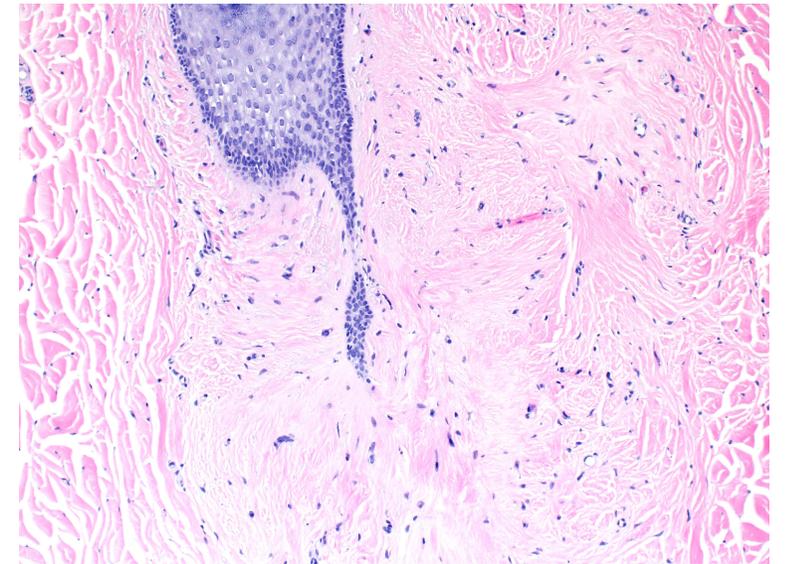
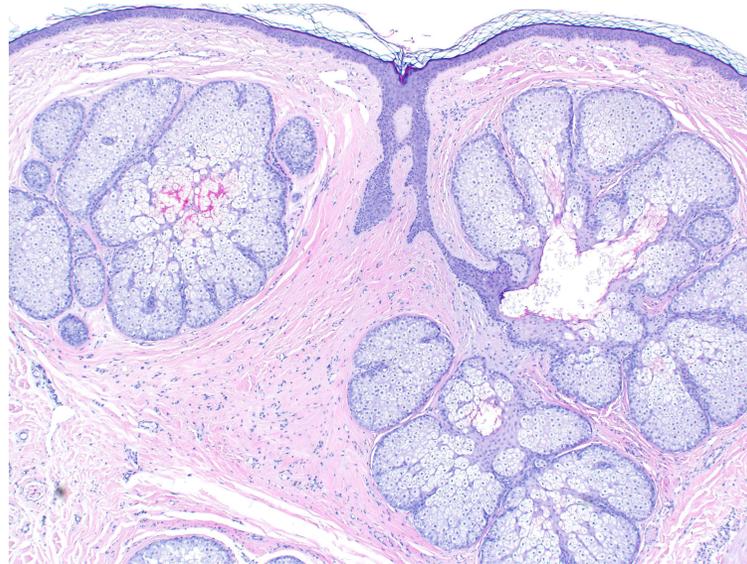
Allergies: ranitidine and penicillins

Pertinent Medications: patient is on multiple drugs for renal transplant and cardiovascular disease

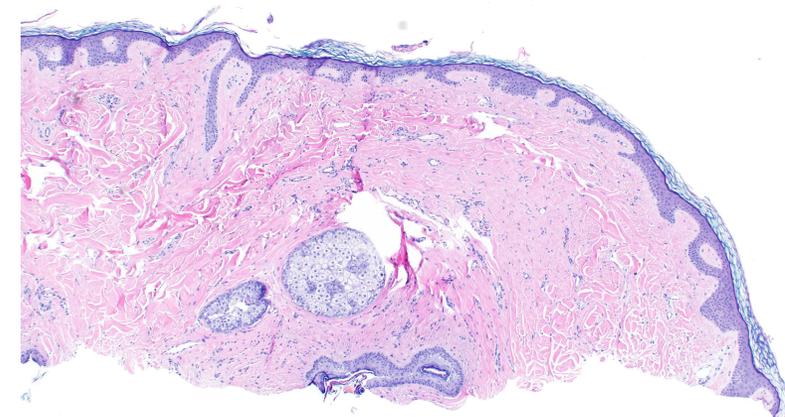
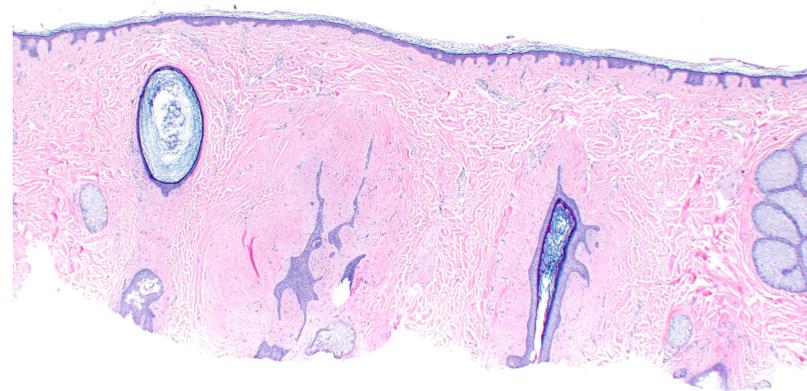
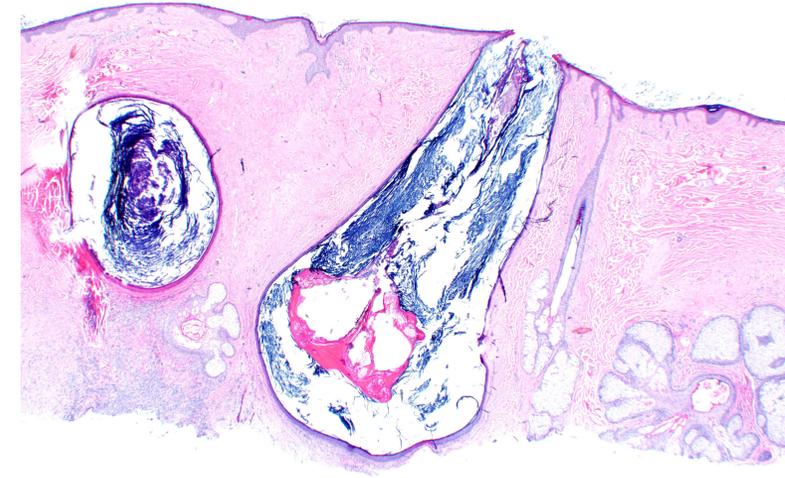
## Laboratory/Pathology results

Shave removal of her right frontal scalp cobblestone skin colored plaque was performed

- Dermatopathology revealed sections containing enlarged sebaceous lobules attached to the undersurface of the epidermis, as well as attached to patulous follicles.



- There are **thin columns of epithelial cells** arranged in a fenestrated pattern within a circumscribed rounded mass of fibrotic and hypervascular stroma, connected to an infundibulum.
- The lesions displayed dermal collagenosis, **increased sebaceous lobules**, and features of angiofibroma and fibrofolliculomas.



# Fibrous Cephalic Plaque(FCP)

- Other dermatologic manifestations of TSC include:
  - Facial angiofibromas
  - Hypomelanotic macules
  - Shagreen patches
  - Ungual fibromas
  - FCPs are **pathognomonic for TSC** <sup>1</sup>.
- With a goal of early TSC diagnosis, this case is particularly significant as it highlights a more uncommon presentation of FCPs.
  - FCPs commonly occur on the forehead but can also more rarely occur on the scalp, face, or neck <sup>2</sup>.
  - Familiarization with these presentations of FCPs, which commonly present from birth to early childhood, will help reduce delayed TSC diagnosis, which can be especially common in patients who lack seizures.
  - With prompt diagnoses, treatment and follow-up of other TSC manifestation can be better managed.
- Treatment/relevant follow-up
  - The FCP was shaved flat to scalp. Will monitor for regrowth and/or symptoms, otherwise no further intervention required.

# References

- 1. Teng JMC, Cowen EW, Wataya-Kaneda M, et al. Dermatologic and dental aspects of the 2012 International Tuberous Sclerosis Complex Consensus Statements. *JAMA Dermatol*. 2014;150(10):1095-1101. doi:10.1001/jamadermatol.2014.938
- 2. Oyerinde O, Buccine D, Treichel A, et al. Fibrous Cephalic Plaques in Tuberous Sclerosis Complex. *J Am Acad Dermatol*. 2018;78(4):717-724. doi:10.1016/j.jaad.2017.12.027

## Question 1.

Rapamycin is the topical treatment of choice for angiofibroma in TSC. What is the mechanism of action of this drug?

- A. Inhibition of mTOR pathway
- B. Inhibition of Calcineurin
- C. Inhibition of IMP dehydrogenase
- D. Binding of the Tac epitope on the IL-2 receptor  $\alpha$ -chain (CD25 and, effectively blocking the formation of the high-affinity IL-2 receptor

## Question 2.

According to the Recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference, **which of the following is NOT a major clinical criteria for TSC?**

A. Fibrous cephalic plaque

B. Ungual hamartomas ( $\geq 2$ )

C. Connective tissue nevus

D. Subependymal nodule

E. Hypomelanotic macules  $< 5\text{mm}$