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An unusual composite tumour: syringoma and angiofibroma in a clinically diagnosed tuberous sclerosis complex.

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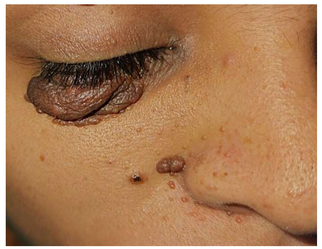
INTRODUCTION

- Syringomas are benign neoplasms that originate from the cutaneous adnexa i.e. intraepidermal segment of a sweat duct.
- Cutaneous angiofibroma is a benign skin tumor characterized by the presence of fibrovascular tissue. These are associated with certain genetic conditions, such as tuberous sclerosis complex, where multiple angiofibromas may be observed.
- This case reports the unusual histopathological finding of a composite tumor incorporating both typical(angiofibroma) and atypical(syringoma) components in a clinically diagnosed tuberous sclerosis complex.

CLINICAL DETAILS

A 21-year-old female with a history of seizures and ash-leaf macules presented with a large (5 x 4 cm) hyperpigmented plaque over the right infraorbital area and multiple skin colored to hyperpigmented papules over nose area since birth.

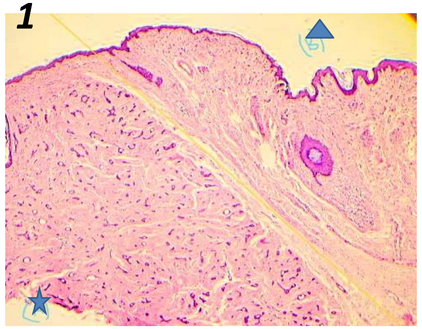


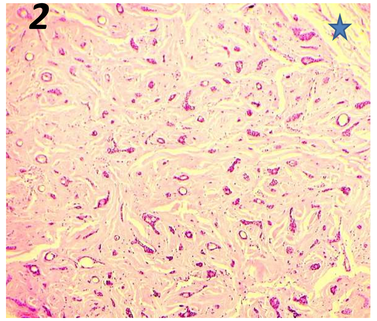
DISCUSSION

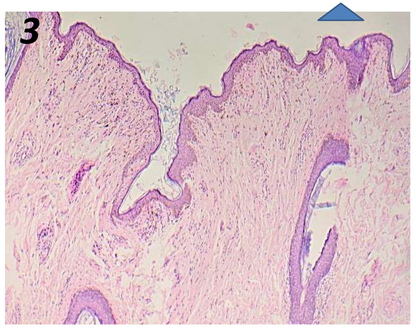
- The pathophysiology of syringomas is unknown, but some speculate hormonal influence to be a major cause , whereas others cite an inflammatory trigger in response to autoimmune conditions, trauma from waxing, radiation, or picking, and heat stimuli.
- In our Case is it reactive hyperplasia seconadary to altered dermis or sporadic in nature needs further studies.
- This case represents a novel phenotypic variation of TSC, suggesting that the cutaneous spectrum of the disease may involve more complex architectural intermingling of adnexal structures than previously recognized.

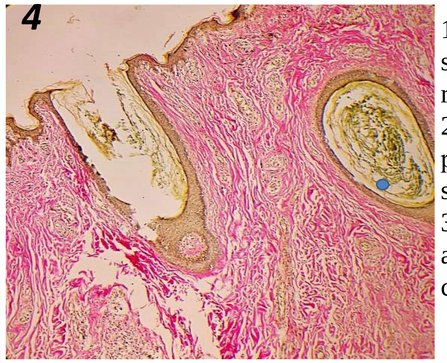
INVESTIGATIONS

Patient undervent investigations to rule out the involvemrent of organs due to TSC. Where MRI brain showed subependymal nodules and cortical tubers. Remaining screening inveastigationsn are withnin normal limits. Patient undervent excision for above mentioned complaint.









1(Low power) Histopathology image showing the composite tumor showing the two part i.e. left side(star side) shows the syringoma and right side(triangle side) angiofibroma.
2(High power) shows multiple small aggregates of epithelial cells producing “comma” & “tadpole” configurations embedded in sclerotic stroma of upper dermis.
3 and 4[EVG(Verhoeff-Van Gieson) stain] shows the vertically arranged thick collagen bundles, blue dot shows "onion-skin" or concentric arrangement of collagen is surrounding the hair follicles.

CONCLUSION

This case highlights the rare finding of a composite syringoma and angiofibroma within a single lesion in a clinically diagnosed TSC. This necessitates histopathological confirmation for all ambiguous cutaneous lesions in TSC patients.