



The Curious Case of Melting Tumours: Organoid Nevus Syndrome

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Introduction

- Nevus comedonicus, when extensive, can be associated with other cutaneous developmental anomalies, including trichilemmal cysts
- Proliferating trichilemmal tumour are usually de-novo, rarely can arise in long standing trichilemmal cysts
- Surgical excision is described as treatment of choice: difficult in areas of high vascularity/ extensive involvement

Discussion

- Associations of extensive nevus comedonicus include
 - Trichilemmal cysts : multiple
 - Skeletal abnormalities
 - Central nervous system involvement
 } Rare
- Underlying Fibroblast growth factor receptor – 2 mutation: explains associations
- Proliferating trichilemmal cyst (aka proliferating trichilemmal tumour) : usually de-novo
 - Association with nevus comedonicus has not been reported previously
 - Surgical excision is treatment of choice
 - Not enough literature on efficacy of radiotherapy : we saw excellent response

Conclusion

- Proliferating trichilemmal tumour, in background of nevus comedonicus, may arise in young age
- In our case, response with radiotherapy (given for 5 weeks) was surprisingly good and sustained over 2-year follow up

References

- Yadav P, Mendiratta V, Rana S, Chander R. Nevus comedonicus syndrome. Indian J Dermatol. 2015;60(4):421.
- Sharma R, Sharma S, Verma P, Yadav P. Proliferating trichilemmal tumor of scalp: Benign or malignant, A dilemma. J Cutan Aesthet Surg. 2012;5(3):213.
- Arora R, Nagarkar N, Prabha N, Hussain N. Nevus comedonicus associated with epidermoid cyst. Indian J Paediatr Dermatology. 2016;17(4):277.

Case Report

Clinical

A 29 year old woman presented with large lobulated tumour over scalp along with loss of hair since last 10 years

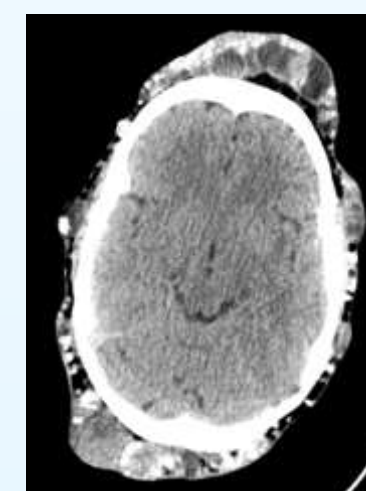


Pitted papules over centrofacial region

Lobulated tumour about 15 x 17 cm
History of **pain and bleeding on minor injury**
History of **recurrence after excision**



Pitted papules in Blaschkoid distribution, bilateral
Nevus comedonicus

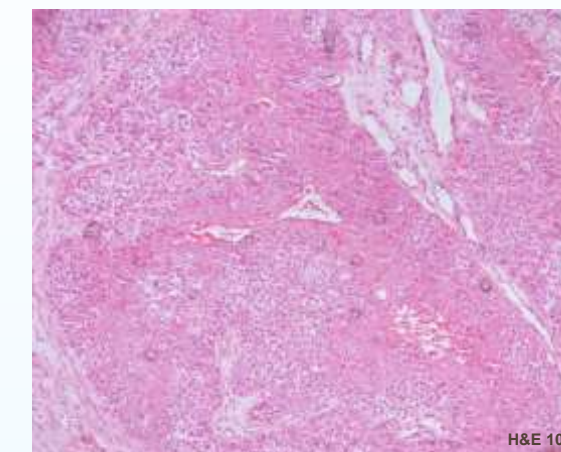


NCCT Head
Multiple nodular solid-cystic subgaleal masses with foci of coarse chunky calcifications

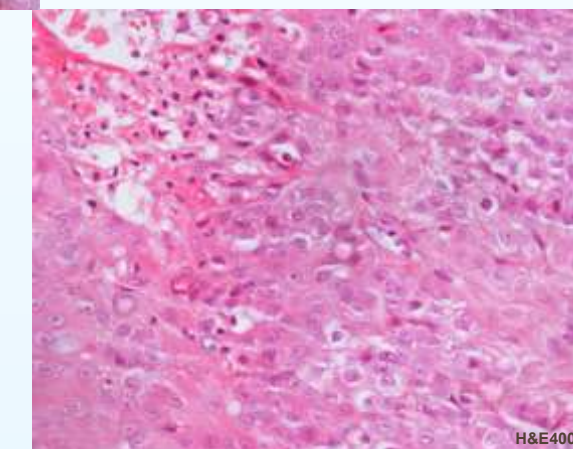
Final diagnosis:
Nevus comedonicus syndrome with trichilemmal cyst and proliferating trichilemmal tumour

Histopathology

Characteristic features of proliferating trichilemmal tumour



Multiple sharply-demarcated lobules of squamous epithelium, with palisading of peripheral layer seen
Abrupt (trichilemmal) keratinization: amorphous eosinophilic keratin noted in the centre of lobules
No cellular atypia noted



Central amorphous keratin,
Individual cell keratinization seen

Treatment

High vascularity of scalp tumour: curative excision could not be planned

Localised Radiotherapy
50Gy/ 25 fractions/ 5 weeks by VMAT technique
Complete replacement by scar tissue in 9 months post-RT

Other trichilemmal cysts:
excision and primary closure

