An Unusual Pink Papule with a Halo

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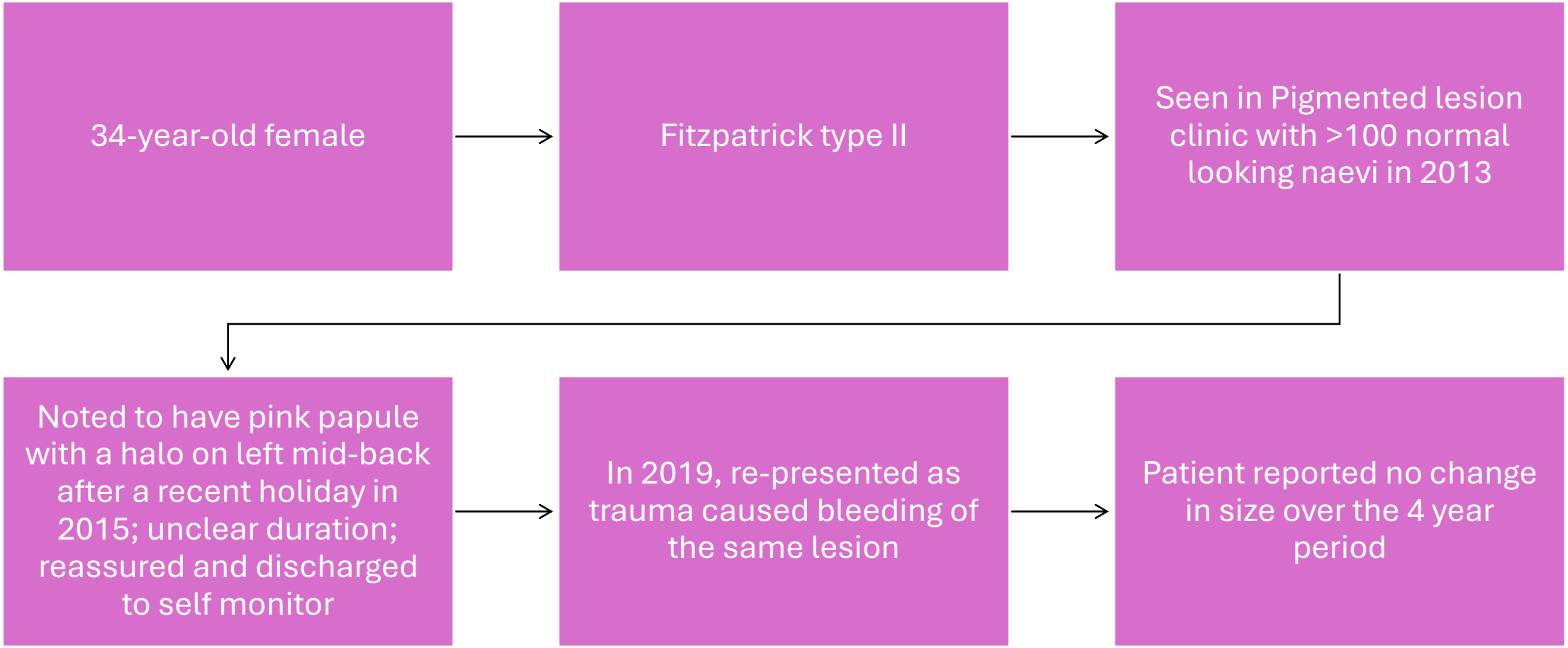
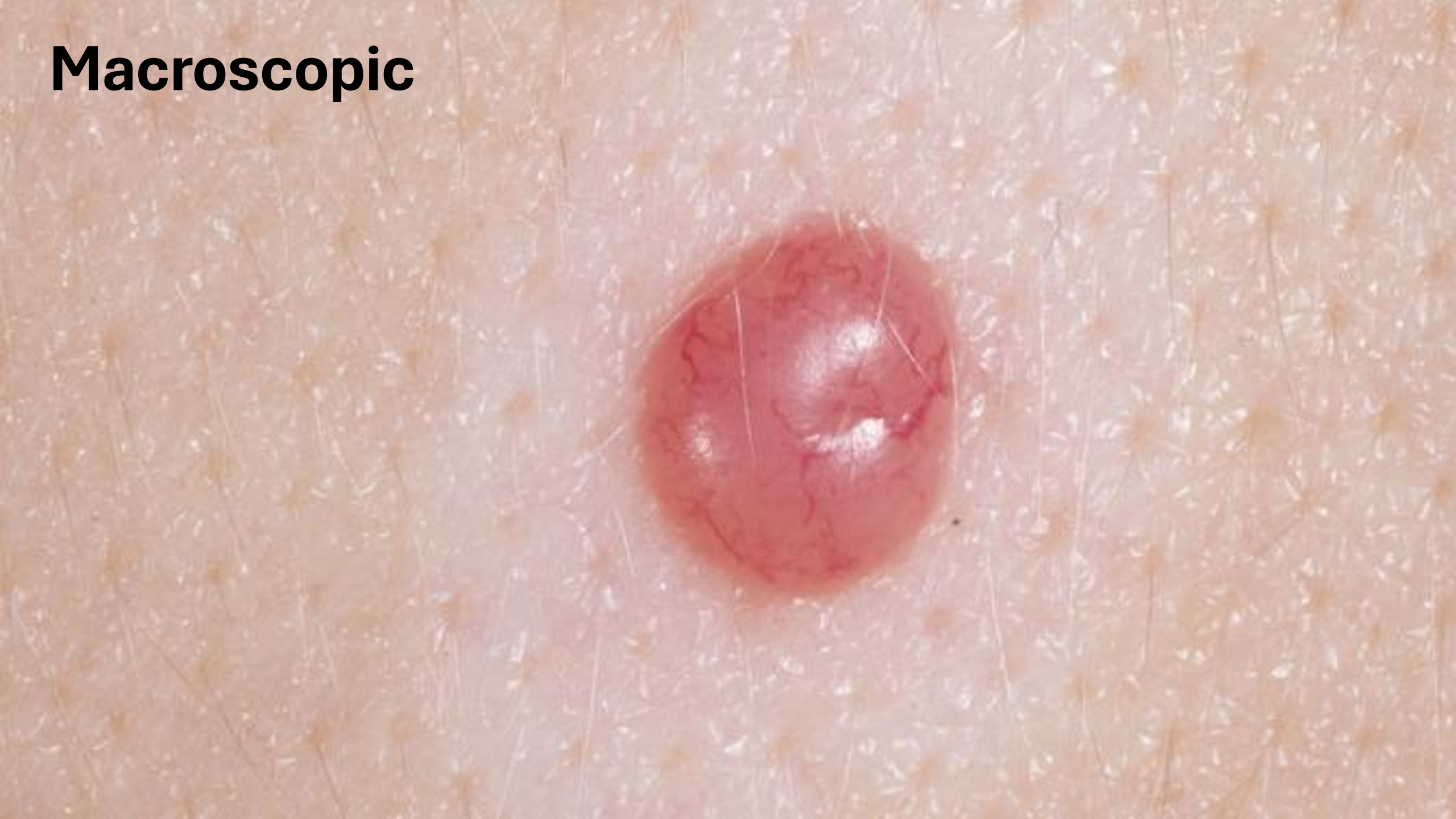
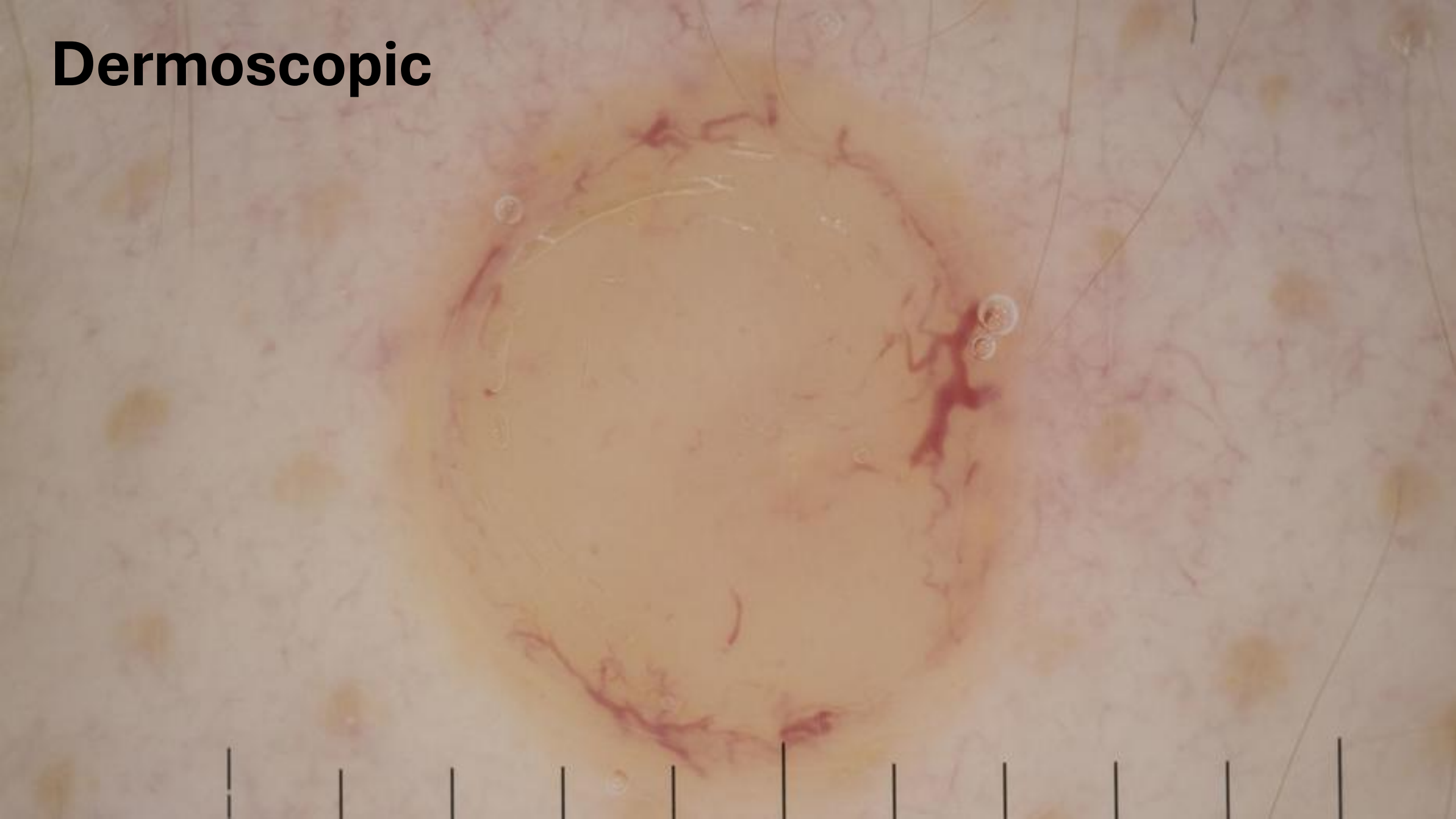
History


34-year-old female
Fitzpatrick type II
Seen in Pigmented lesion clinic with >100 normal looking naevi in 2013
Noted to have pink papule with a halo on left mid-back after a recent holiday in 2015; unclear duration; reassured and discharged to self monitor
In 2019, re-presented as trauma caused bleeding of the same lesion
Patient reported no change in size over the 4 year period

Macroscopic



Dermoscopic

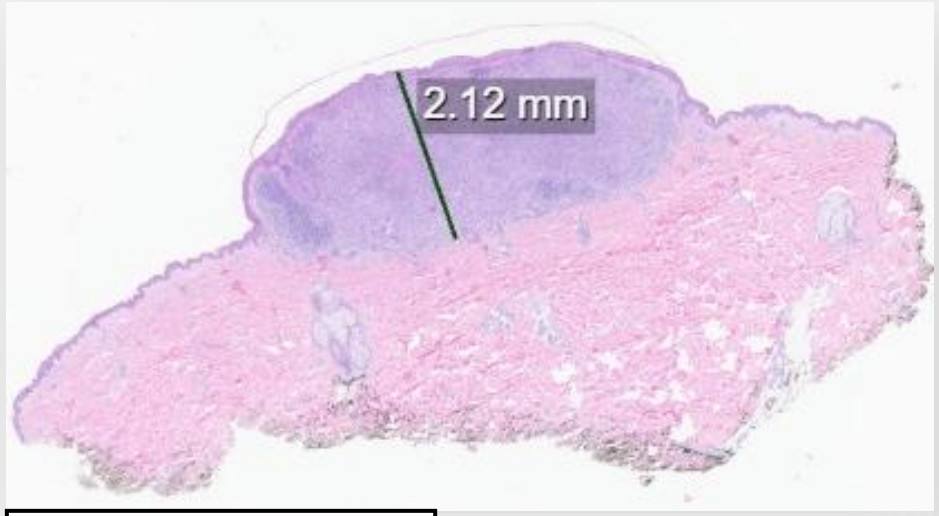


Differential Diagnosis

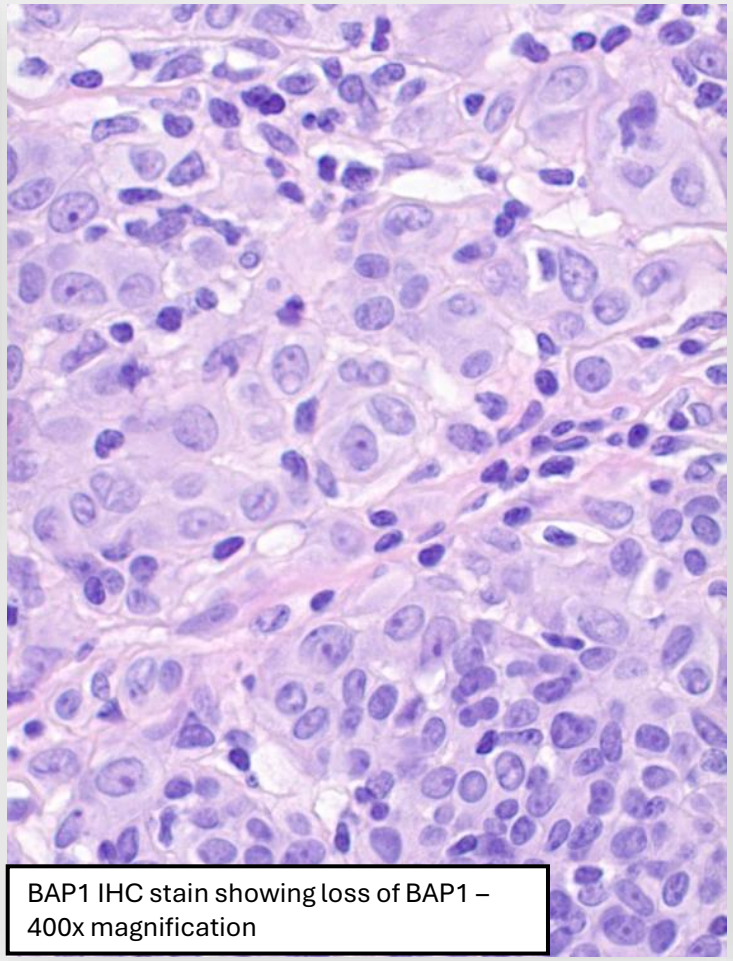
1. Intradermal Halo Naevus
2. Spitz naevus with Halo
3. Melanocytoma
4. Melanoma
5. Basal cell carcinoma

Follow-up and Management:

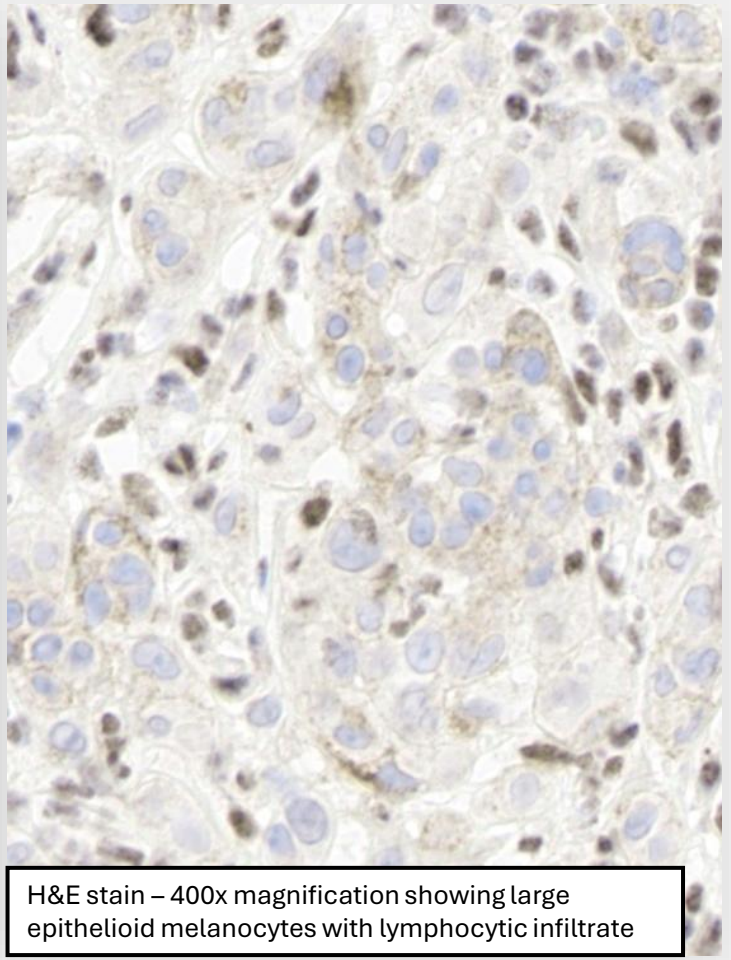
The lesion had slightly increased in size so it was excised with a 2mm clinical margin – removed with a histological margin of 2.4mm peripheral and 3.1mm deep



H&E stain – x4 magnification



BAP1 IHC stain showing loss of BAP1 – 400x magnification



H&E stain – 400x magnification showing large epithelioid melanocytes with lymphocytic infiltrate

Diagnosis:
BAP1- inactivated melanocytoma (BIM)

Further Management:

Melanoma MDT Plan

- 1cm Wide Local Excision - Clear
- Germline genetic testing – NEGATIVE for BAP1 tumour predisposition syndrome

Risk-stratification in 2023 (with further molecular testing based on experience at specialist centre)

Key Histopathological features

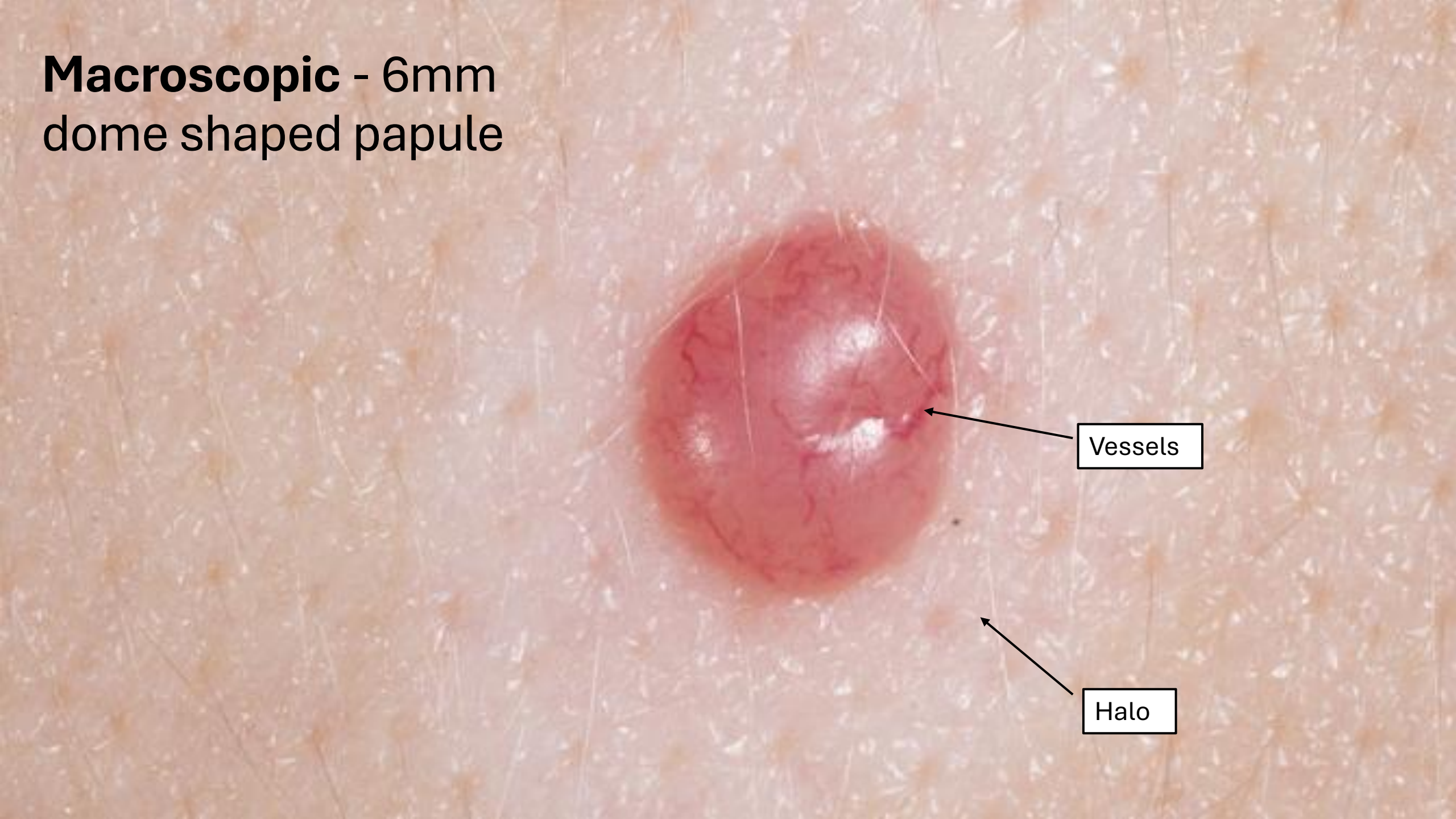
- Tumour thickness: 2.12mm
- Mitoses: 1 per mm^2
- Ki67: <5%
- No atypical junctional component
- PRAME: Negative
- P16: Retained

Key Molecular features

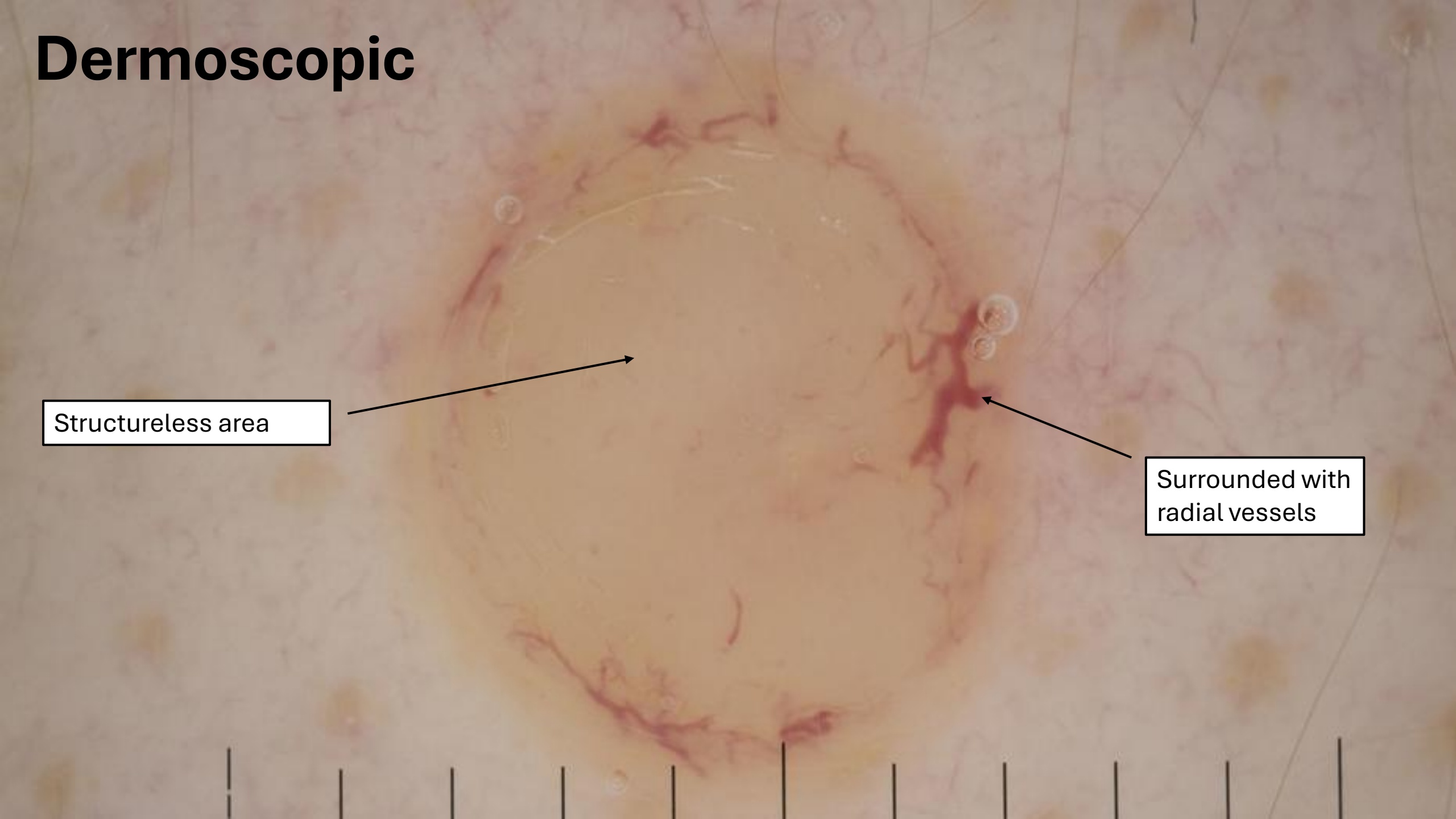
- BRAF driver mutation detected
- NRAS Negative
- TERTp negative
- Copy Number negative

Conclusion:
Low-risk BAP1-inactivated melanocytoma

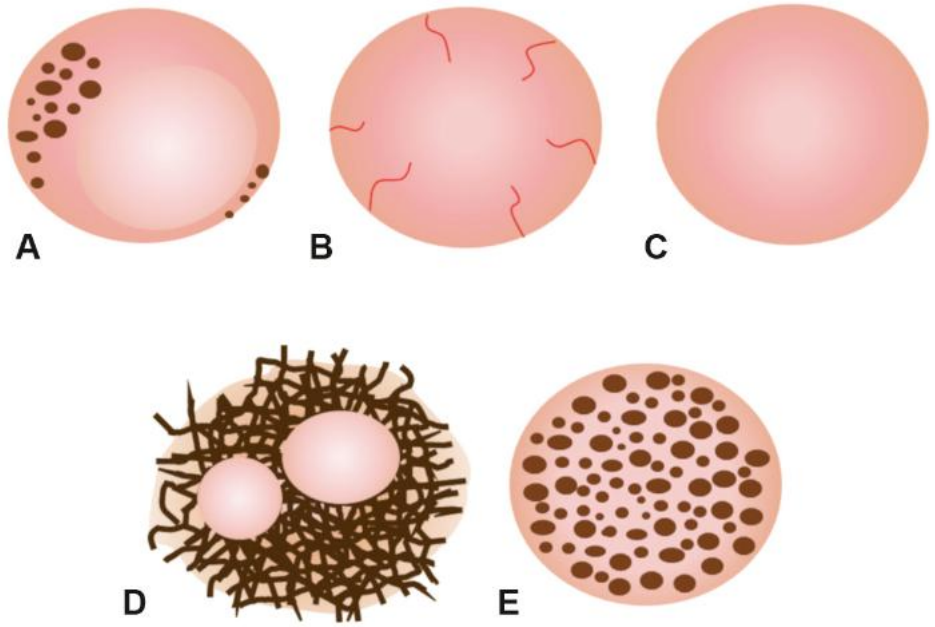
Macroscopic - 6mm
dome shaped papule



Dermoscopic



Brief summary of diagnosis and associations



Schematic showing the five identifiable patterns of BIMs as categorised by Yelomas, 2019. (2)
A, Structureless pink/tan with atypical eccentric clods.
B, Structureless pink with radial vessels
C, Structureless pink/tan
D, Network with raised structureless areas
E, Globular.

Background

- Melanocytomas are intermediate melanocytic tumours distinct from both naevi and melanoma in histology and genetics *(1)*.
- BIMs were first reported by *Wiesner et al. (3),* typically appearing as dome-shaped pink to brown papules.
- Halo phenomenon in BIMs is not reported in the literature, however there are limited clinical papers describing these lesions.
- BIMs fall within the low cumulative solar damage melanoma pathway, typically driven initially by a **$BRAF^{V600E}$ mutation**. Progression involves **biallelic loss** of the BAP1 tumour suppressor gene *(4)*.
- While most cases are sporadic, **~12% involve germline BAP1 mutations** combined with somatic second-hit loss, indicating BAP1 tumour predisposition syndrome, which confers increased risk for several malignancies e.g., melanomas *(5).*

Dermoscopic patterns

- *Yélamos et al.* identified **five dermoscopic patterns** of BIMs in patients with germline mutations (2). Our case exhibited the ‘B’ pattern. More work is required to describe the dermoscopic patterns of these lesions.

Other learning points:

Purpose of Risk-Stratification	Specialist MDT referral allows for risk stratification of BIMs into low-risk, high-risk and melanoma. This should be done by considering the key histological and molecular features we have described. The lesion in our case would be considered **low risk** as there are no atypical features. Therefore, a WLE would not be performed.
Value of Early Discussion	Early recognition and MDT review enables timely decisions regarding excision, follow-up, or genetic referral.
New germline testing criteria	As only one BIM is present, our patient **does not meet <u>current</u> NHS criteria** for germline BAP1 testing (**≥2 core BAP1 related tumours, which would include 2 BIMs) – updated in 2024** with our patient undergoing germline testing in 2021.
Ongoing Vigilance	We present this case to highlight the interesting dermoscopic patterns seen in BIMs and to help clinicians recognise these lesions.

References

1. Elder DE, Massi D, Scolyer RA, Willemze R (Editors). *WHO Classification of Skin Tumours*. 4th edition. International Agency for Research on Cancer (IARC), Lyon, France; 2018.
(World Health Organization Classification of Tumours, Volume 11).

2. Yélamos O, Cuevas R, Gutiérrez C, Dusza SW, Marchetti MA, Chen L, et al. Dermoscopy of BAP1-inactivated melanocytic tumors: a morphologic study of 32 cases. *J Am Acad Dermatol*. 2019;81(5):1096–1104.

3. Wiesner T, et al. A distinct subset of atypical Spitz tumours is characterized by BRAF mutation and loss of BAP1 expression. *Am J Surg Pathol*. 2012;36:818–30

4. Shain AH, et al. The Genetic Evolution of Melanoma from Precursor Lesions. *N eng J med* 2015;373:1926-1936

5. Carbone M, et al. BAP1 cancer syndrome: malignant mesothelioma, uveal and cutaneous melanoma, and MBAITs. *J Transl Med*. 2012;10:179

BSDS Consent Form

- Signed BSDS consent form attached