## Nasal Dorsum Mass case and review

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#### م 11:36pm

ER doc: we have a 65 yo female, DM, HTN, c/o nasal dorsum mass since 11 months with MRI report "hemangioma"... please come and assess now!!



#### Hx

- Started as small pimple
- Progressive increase in size
- No aggreviating factors
- No reliveing factors or use of medications
- No associated symtoms
- No sun exposure
- No biopsy

- -ve FHx
- Non smoker or substance use.
- -ve surgical hx or trauma
- Unremarkable PHx

### PE:

- Looks well
- 2\*3 cm smooth rounded firm pedunculated mass over nasal dorsum, non tender, no hotness, or fluctations.
- There is pulsations
- Does not change with valsalva
- Scope: patent NC bil, no extension to NC, clear NP, OP, HP, mobile VCs
- Neck: no LAP
- Skin: no other skin lesions
- Eyes: intact LR and EOM
- CNs grossly intact
- No smilar lesion elsewhere



## Next?

### Imaging





• MRI: midline cutaneous subcutaneous pedunculated nasal mass with underlying bone defect represent primary skin lesion or **haemangioma** 

Labs: WNL

# How about biopsy?

### DDx?

- Hemangioma
- Primary skin lesions (BCC, SCC.. Others)
- Post traumatic deformitis
- Dermoids
- Gliomas
- Encephaloceles
- Extension from inside nose (esthisionneuroblastoma, lymphoma, rhabdomyosarcoma..)
- Infection: leprosy, rhinoscleroma, rhinophyma

# How would you manage this pt?



# She underwent excision of lesion with staged paramedian forehead flap







#### Pathology report (final)

- Diagnosis: <u>Merkle cell carcinoma</u>
- **Tumour size**: 2.8 cm in maximum dimensions at dorsum of the nose
- Margins: carcinoma is <u>present</u> at surgical resection margins
- **Tumour extent**: epidermis dermis and skeletal muscle fibres
- Lymphovascular invasion: present

# What would you do next?

#### Survillance

- **CT CAP**: bil level IIB metastatic LNs. -ve CT chest, abdoment and pelvis.
- **PET CT:** hypermetabolic locoregional upper cervical lymphadenopathy in keeping with the locoregional metastasis. Rest of body –ve.

# Knowing this, whats you plan for next surgery?

### 2<sup>nd</sup> stage surgery

- Realease of PFF pedicle
- Extension of excsion margines with frozen
- All came back –ve



#### Tumer board

- Case presented in TB
- Final Staging: pT2, pN1, cM0, III
- Final plan: Bilateral MRND (I-IV) + for post operative radiotherapy

#### Follow up

- Surgical sites healed well
- Started RTx last week

**Topic Review** 





#### Editorial Merkel Cell Carcinoma: An Update and Review

2023

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#### Introduction

- MCC is a rare, aggressive neuroendocrine skin cancer with high mortality rate (~30%).
- Described by Toker in 1972.
- Predominantly affects older individuals and has a high risk of metastasis.
- Since the 1990s, the number of diagnosed cases has increased by 5% to 10% per year.

#### Merkel Cell Carcinoma (MCC)

MCC is a rare skin cancer that usually develops on sun-exposed skin.

It is most common in men with fair skin over the age of 50.

MCC usually develops as a painless, rapidly growing bluish to red bump.

#### Pathophysiology

- Originates from **Merkel cells**, which are touch receptors in the skin.
- Detection of the Merkel cell polyomavirus (MCPyV) in approximately 80% of cases.
- Genetic mutations and **UV radiationinduced** damage also play a significant role.



#### **Risk Factors**

- Significant UV exposure and having a light skin phenotype.
- Immunocompromised states
- History of other cancers or chronic inflammatory conditions affecting the skin.



#### **Clinical Presentation**

- Painless, firm, red to violaceous nodule on sun-exposed skin.
- Commonly appears on the head, neck, and extremities.
- Often mistaken for other skin malignancies such as melanoma or lymphoma.
- "AEIOU": Asymptomatic/lack of tenderness, Expanding rapidly, Immune deficiency, Older than 50 years, and UV exposure.



## Diagnostic Approaches

- Biopsy revealing characteristic small, round, blue cells. "<u>most</u> <u>important</u>"
- Imaging: CT, MRI, PET/CT scans for detecting metastasis and surgical planning.
- MCPyV serology (-ve = poor prognosis).



**Fig 1**. MCC stained by A) hemotoxylin and eosin showing mononuclear cells with scanty population and B) CK 20 showing a characteristic perinuclear pattern. (Feng et al. Science 2008 319: 1096-100)

\*\*CK20 is a diagnostic marker with high sensitivity and specificity

#### Treatment

- Surgical removal with wide local excision (1-2m) is the first-line treatment.
- Adjuvant radiation therapy to reduce recurrence risk.
- Recent approvals in immunotherapies (e.g., checkpoint inhibitors) anti-PD-1/PDL1 inhibitors such as Avelumab, Pembrolizumab, and Nivolumab. *"all still in phase 2 trials"*



#### **Prognosis and Survival Rates**

- Prognosis depends on stage at diagnosis, with a 5-year survival rate varying widely from 51% to 64%.
- High recurrence rate ~40%; regular follow-ups are critical.
- Importance of patient education on self-monitoring for new lesions or changes.

