RARE CASE OF MALIGNANT PROLIFERATING TRICHILEMMAL TUMOUR

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PRESENTATION OF CASE

- A 45 years old female patient came with c/o non-healing ulcer over left occipital region since a year.
- She gave history of being operated thrice for nodular growth over occipital region, which recurred after two surgeries and after the third surgery she developed an ulcer.
- The three surgeries took place in 2015- Satara, May 2016- Phaltan and in Dec 2016- Phaltan.
- CT brain was done during all these surgeries, which revealed no intracranial abnormality or involvement of cranial vault.
- HPE done after surgery in May 2016 was s/o Epidermoid tumour/ infected sebaceous cyst in the subgaleal region.

Presentation in Casualty

Preoperative Presentation

- Local Examination- An 8 x 11 x 1 cm ulcer was seen in left occipital region which was irregular in shape, covered with granulation tissue and slough.
- The ulcer edge was sloping and ulcer bed bled on touch.
- A single firm mobile non-tender cervical lymph node (Level 2) was palpable 3 x 2 x 1 cm on the left side.

DIFFERENTIAL DIAGNOSES

- Cutaneous squamous cell carcinoma.
- Sebaceous carcinoma.
- Clear cell hidradenocarcinoma.
- Cutaneous metastasis of RCC.
- Cylindroma.
- Dermoid cyst.
- Epidermal inclusion cyst.^[1]

CLINICAL DIAGNOSIS

Patient underwent routine lab investigations, four quadrant biopsy of occipital lesion and MRI brain after being admitted in our hospital in Dec 2017.

- HPE of biopsy was s/o malignant proliferating trichilemmal tumour.
- MRI brain- ill-defined heterogeneously enhancing lesion in left occipital region invading underlying bone and dura.

CT (Brain) s/o Breach in Bone

MRI (Brain) s/o Intracranial Extension

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DISCUSSION OF MANAGEMENT

Preoperative workup was done and patient was prepared for surgery and decision was taken to operate the patient by oncosurgeon, neurosurgeon and plastic surgeon-

- Firstly, oncosurgeon did WLE of occipital lesion with 2cm margin and left level 2, 3, 4, 5 neck lymph node dissection.
- Then neurosurgeon removed the involved underlying bone of approx. 6 x 5 cm and dura 2 x 1 cm.
- Then plastic surgeon harvested tensor fascia lata graft and pedicled trapezius flap 15 x 10 x 1 cm.
- The dural defect was covered with tensor fascia lata graft.
- The occipital defect was covered with trapezius flap with transverse cervicalis artery pedicle after creating a tunnel below the skin and rotating it.
- The defect which developed on the donor site of trapezius flap was covered with STSG from right thigh.
- Vascularity of trapezius flap was checked.
- Three drains were kept in the neck region, trapezius flap region and occipital region above dura.

Intraoperative finding- involved Dura removed- Brain Tissue seen Bulging from Below

Postoperatively-Neck Lymph Node Dissection

Wide Local Excision-Trapezius Flap

- 4 specimens were sent for HPE-neck dissection, WLE tumour, occipital bone, dura.
- Patient was given antibiotics for 7 days and check dressing of skin graft was done on POD 5 - 100% graft take was there.
- All three drains were removed on POD 10.
- Patient complained of headache and fever on POD 15, USG revealed loculated collection of approx. 8 x 1.6 cm- IV antibiotics and acetazolamide tablets were given for the same for 5 days as advised by neurosurgeon.
- Follow-up- Patient was followed up every month. Patient had no e/o recurrence.
- Suture site was well.
- Patient had not started radiotherapy.
- Patient was counselled for radiotherapy and sent to higher centre for the same.

Follow-Up Picture after 5 Months

PATHOLOGICAL DISCUSSION

Histopathologically, WLE specimen s/o- Malignant proliferating trichilemmal tumour- all margins free of tumour.

- 3 out of 19 neck lymph nodes dissected showed tumour deposits.
- Bone and dura showed tumour deposits.

Abrupt Keratinisation

Cords and Nests Appearance

- IHC-CD 34-negative, Calretinin-positive, p53-70% positive, Ki67-70% positive.

Calretinin **P53**

- Calretinin was done to differentiate it from squamous cell carcinoma, which is the most common differential. Calretinin is never positive in SCC.

FINAL DIAGNOSIS

Proliferating Trichilemmal Tumour (PTT)

It is a tumour originating from the isthmus of the outer root sheath of a hair follicle-

- It is also commonly called a proliferating trichilemmal cyst or less commonly proliferating follicular-cystic neoplasm. It was first described by Wilson-Jones.^[2]
- The usual clinical presentation of PTT is that of a long standing, subcutaneous, cystic nodule that slowly progresses to a large nodular mass.
- Ye et al^[3] proposed three groups of PTT-
 - Group 1- Benign lesions that are non-recurrent display well-circumscribed margins, modest nuclear atypia and no pathological mitoses-
 - Group 2- Low-grade malignant lesions (locally aggressive) that may recur, exhibit irregular and locally invasive margins and involvement of deep dermis and subcutis.
 - Group 3- High-grade malignant tumours with a tendency towards recurrence and lymph node metastasis, invasive pattern of growth, prominent nuclear pleomorphism, atypical mitosis, necrosis and possible vascular and neural invasion.
- Malignant transformation in such a tumour is very rare, evidenced by regional or distant metastasis,^[4] unusual finding (only 39 cases have been documented in the English language literature till date).^[5]

- It is affecting the scalp of elderly women.^[6] It is a large, solitary, multilobulated lesion that may arise within a trichilemmal cyst (Pilar cyst). It yields lobulated and variably exophytic masses that occasionally ulcerate.^[7]
- Malignant transformation occurs occasionally, which can be manifested by sudden rapid growth. Histologically, malignant PTTs show severe nuclear atypia, marked cellular pleomorphism with atypical mitoses, dyskeratotic cells and infiltrating margins.^[8,9]
- Malignant transformation in such a tumour was introduced by Saida et al,^[10] who suggested three stages in the oncological development- adenomatous stage, epitheliomatous stage and carcinomatous stage.
- There are two growth patterns seen in malignant proliferating trichilemmal tumour-circumscribed nodular and diffuse spindle cell type.
- It is usually confused with squamous cell carcinoma.
- It is more common in females and in older individuals 50-75 years old.
- It is a chemo-resistant tumour.

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