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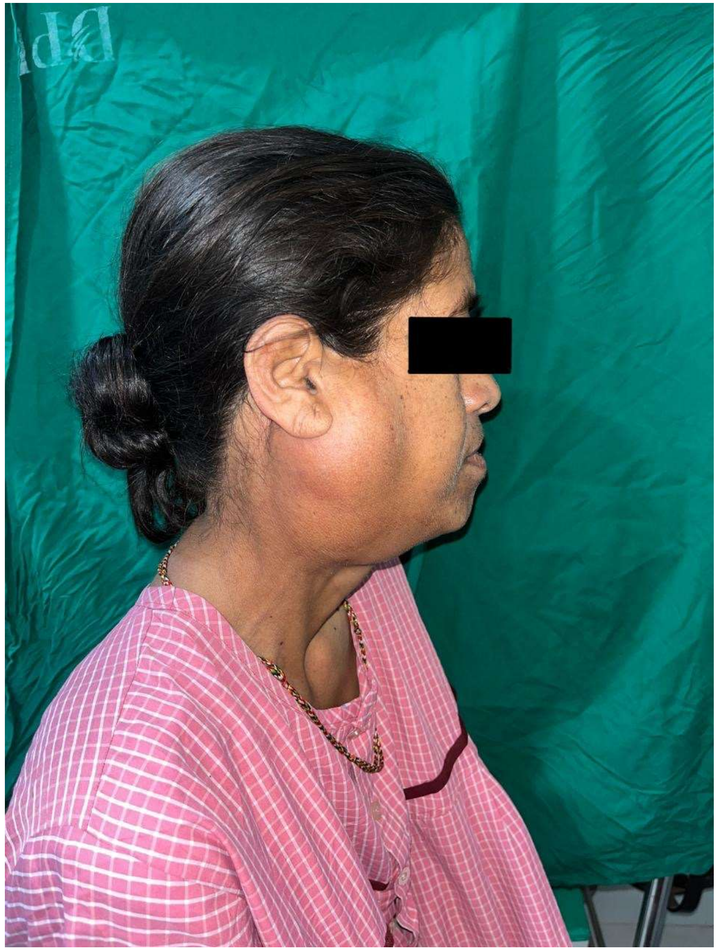
(DEEMED UNIVERSITY)

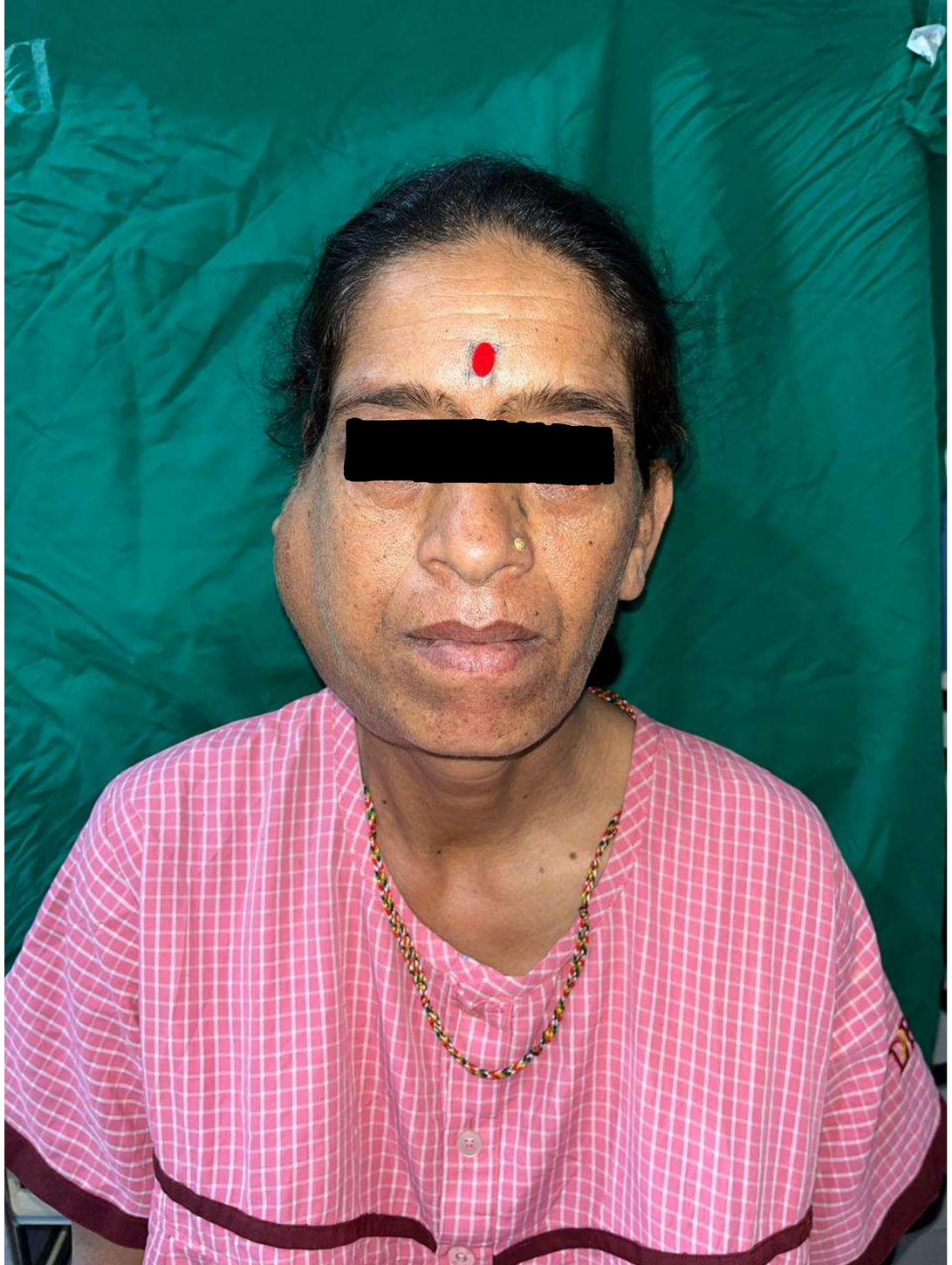
Parotid swelling with a twist: A diagnostic surprise



46/F
Homemaker
Khed, Maharashtra

History - Presented with swelling on the right side of the face x 8 years
- initially of 1x1cm and gradually progressed to the size of 8x5cm.

- C/o difficulty in swallowing of solid foods.
- No c/o pain
- No h/o trauma
- No c/o breathlessness
- No c/o facial weakness, change in voice, weight loss , loosening of teeth.
- No c/o excessive salivation
- No c/o difficulty in opening of jaw

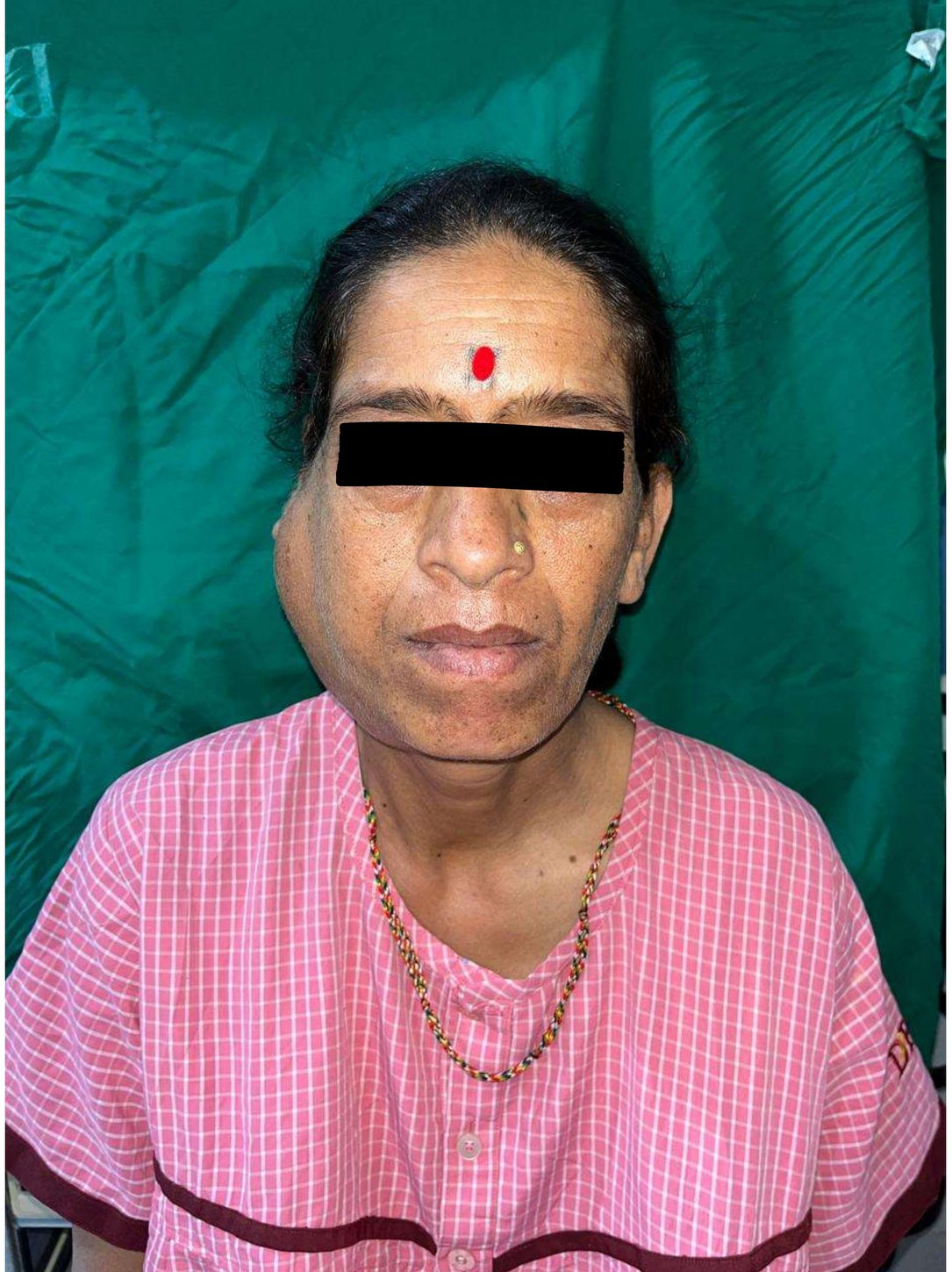


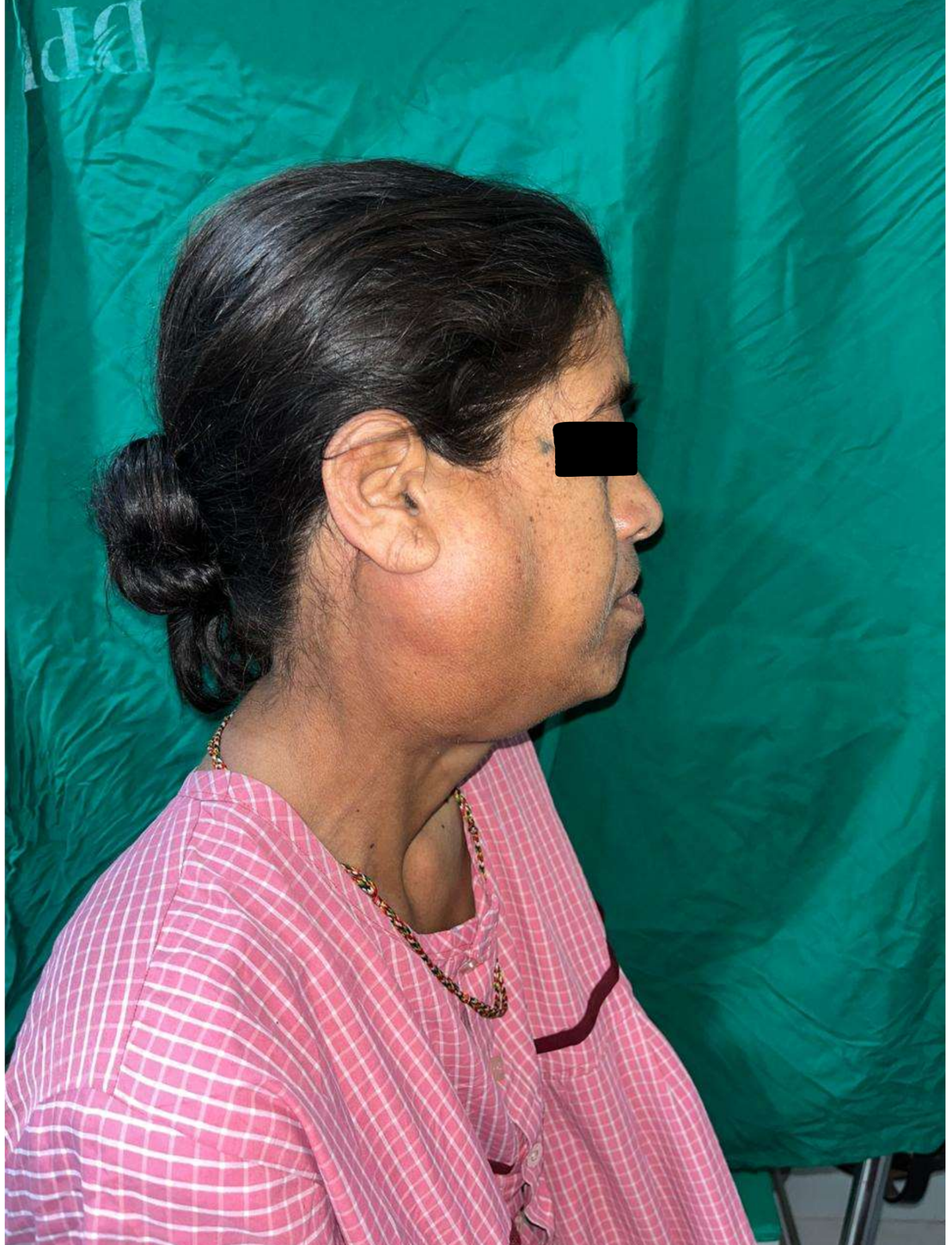
- No history of radiation exposure or exposure to harmful chemicals or drugs[1]
- No comorbidities.
- No similar complaints in family

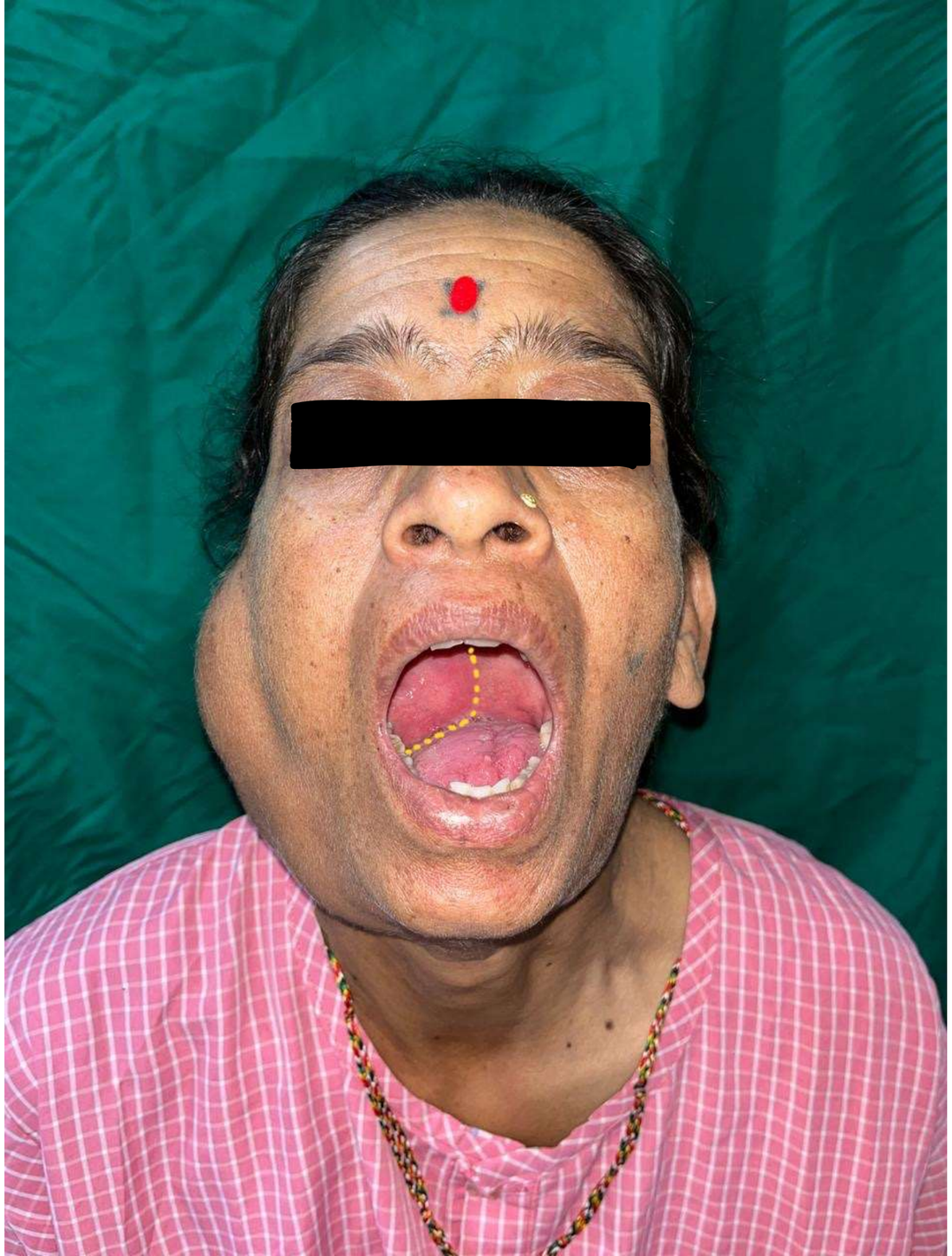
General physical examination

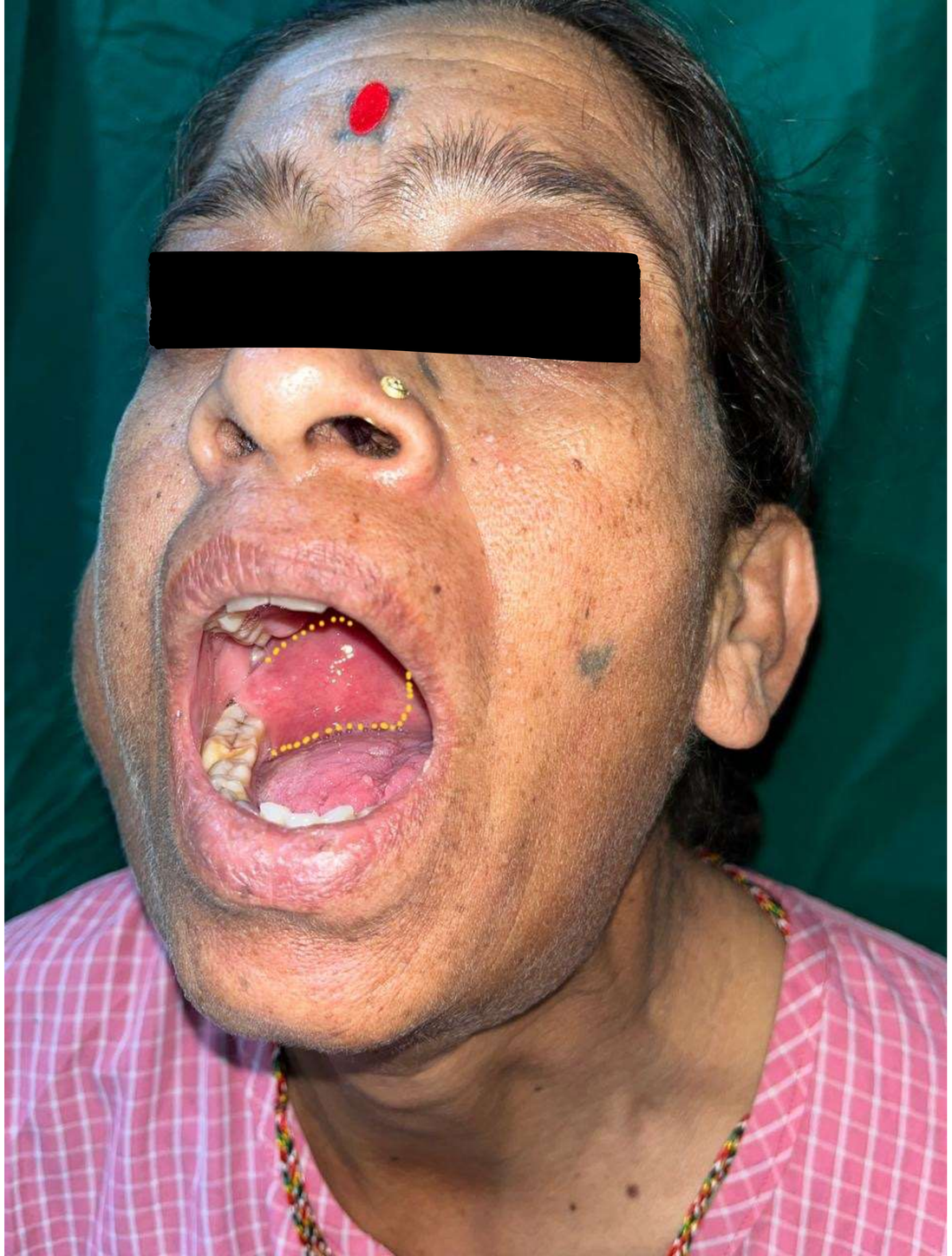
Patient was well built and nourished

1. **Sabiston textbook of surgery , 21st edition, chapter 34, page no 789.**









Systemic examination

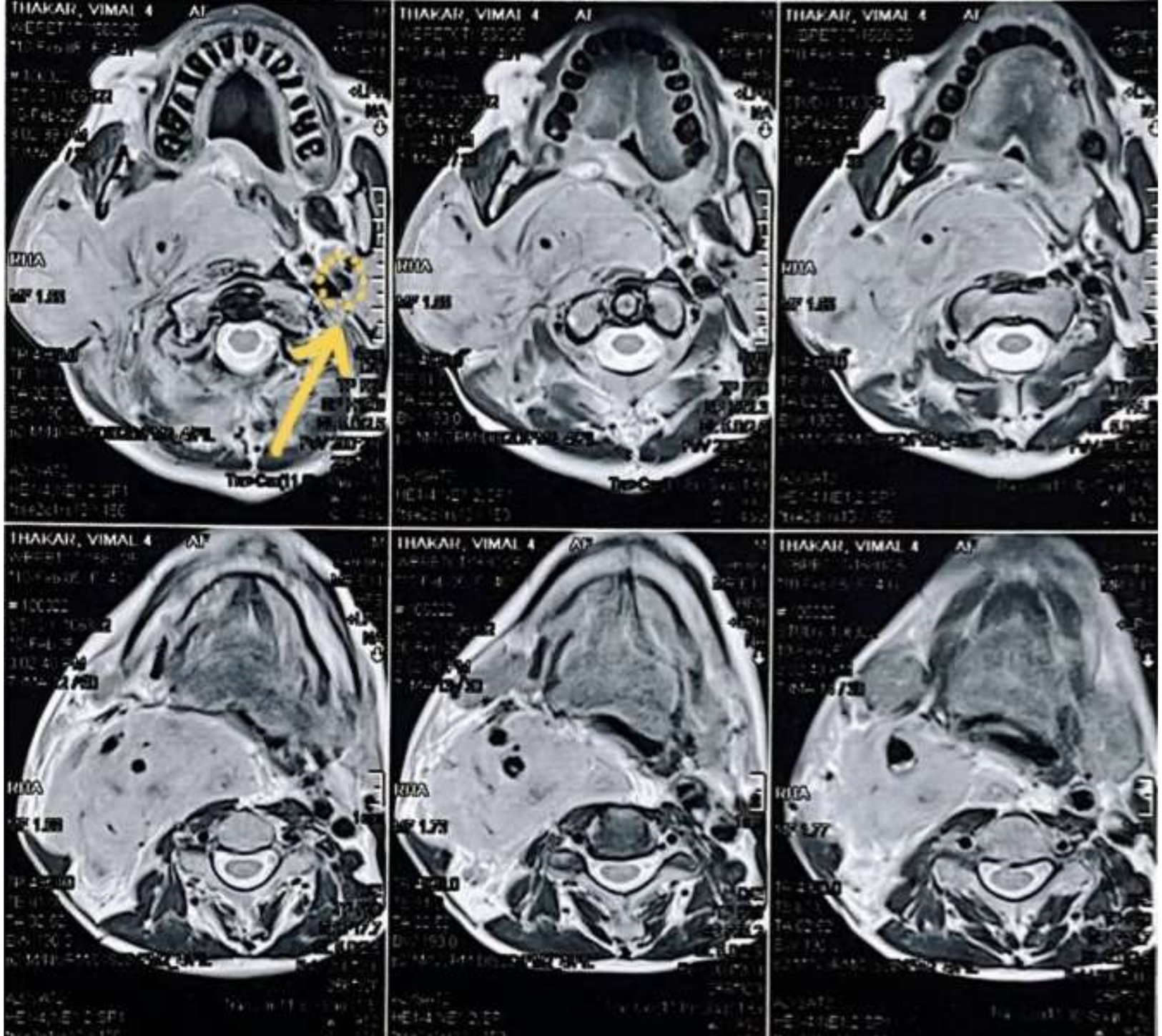
CVS- S1, S2 normal , no murmur
RS - normal vesicular breath sounds
PA - soft , non tender , no organomegaly
CNS - conscious and oriented
Breast - Bilateral breasts were normal , no evidence of lump.

Provisional diagnosis

- Parotid swelling under evaluation.

MRI report

- Large well defined heterogeneous intensely enhancing mass lesion involving the right parotid, carotid space
- Extending medially into the right prevertebral and right parapharyngeal space
- Superiorly into the right masticator space with extent and relations as described. - ? carotid body tumour (Shamblin Type III - histology suggested.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FNAC report

IMPRESSION:

Right parotid swelling, FNA

• Neoplasm- Benign (Category IV A)

Comments:

Findings are suggestive of monomorphic adenoma / myoepithelioma
Differential diagnosis includes cellular pleomorphic adenoma.

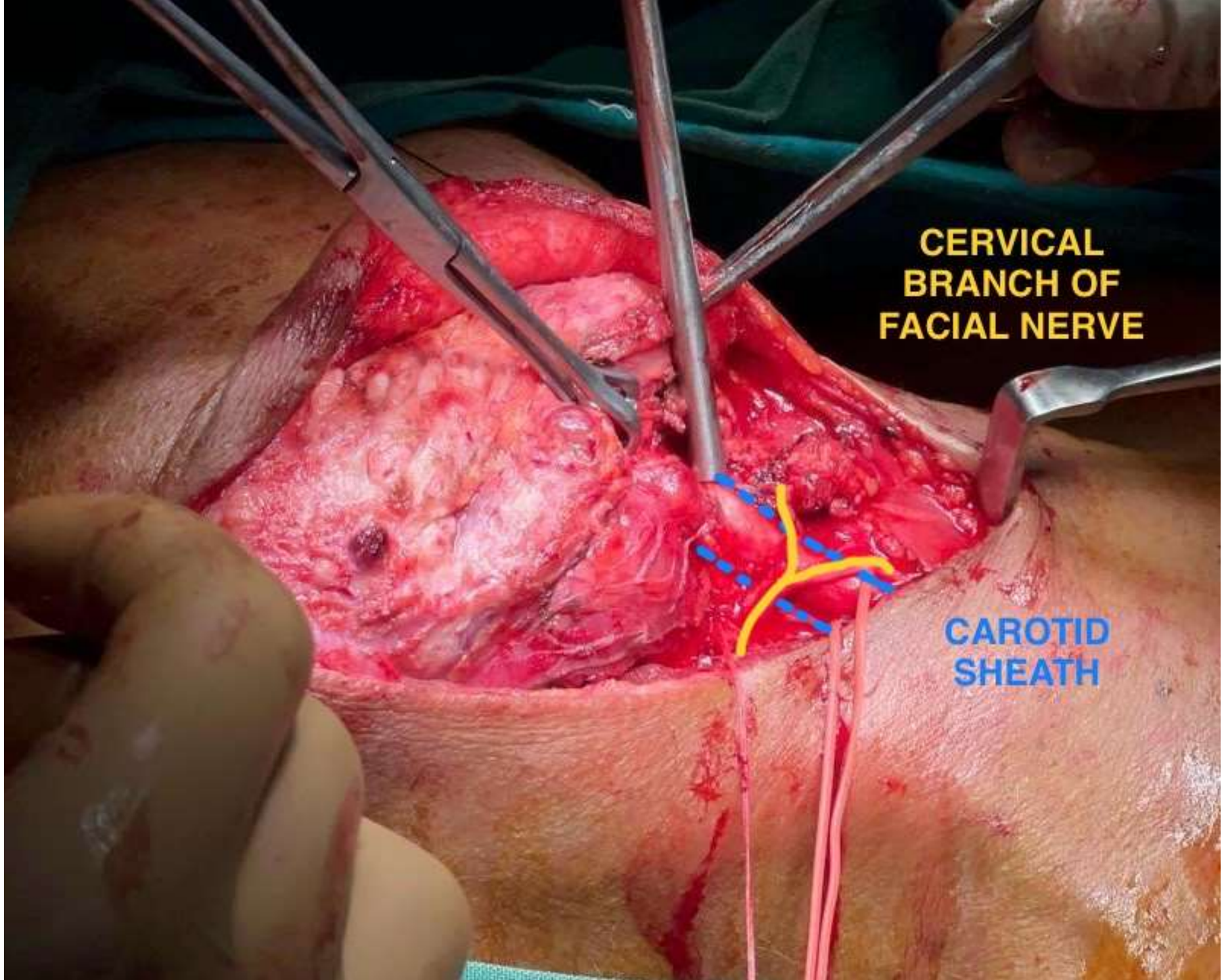
Risk of malignancy- < 5%

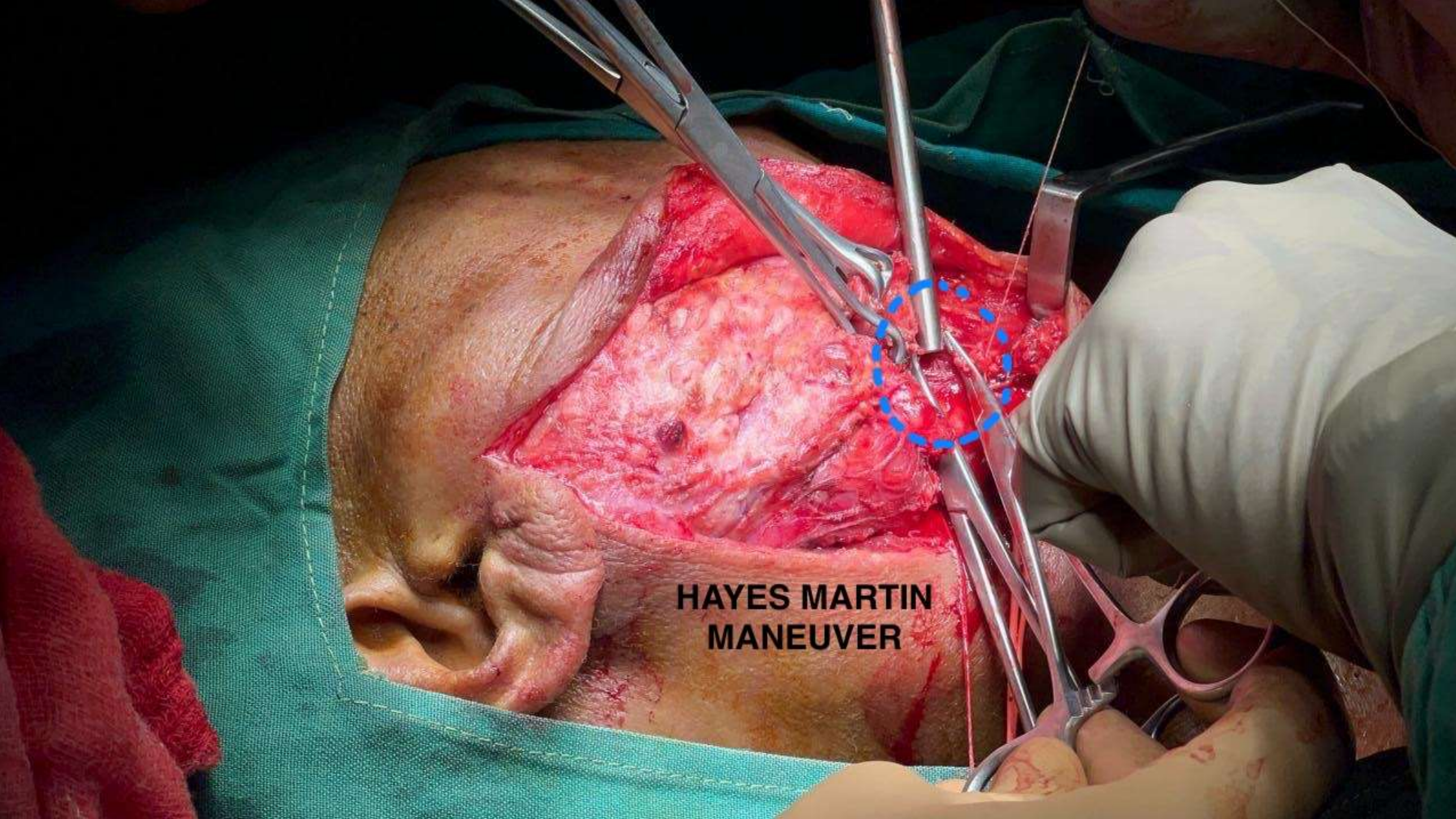
Reporting system used: The Milan System for Reporting Salivary Gland Cytopathology

Table 1: Milan System for Reporting Salivary Gland Cytopathology

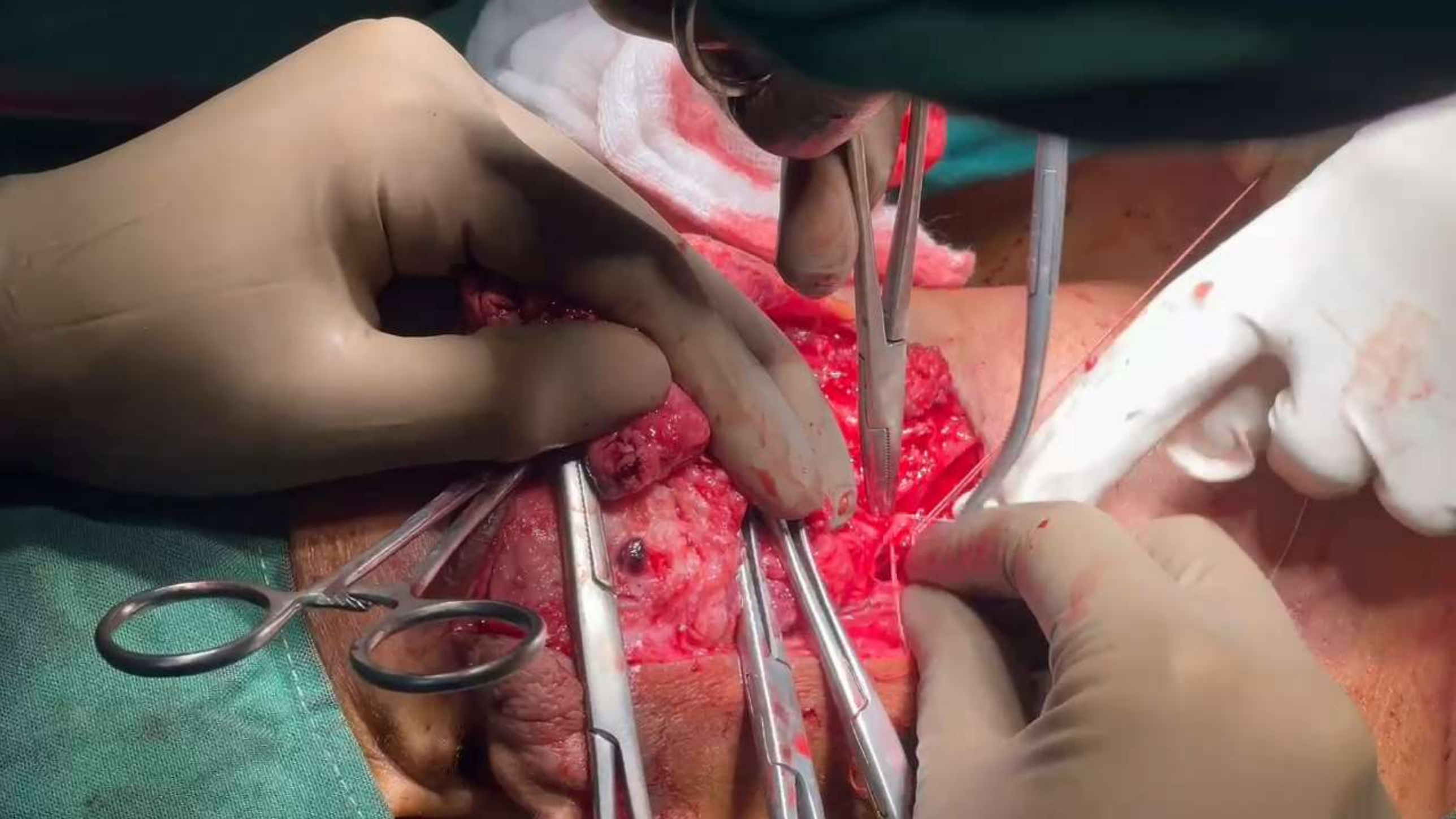
Diagnostic Categories		Risk of Malignancy %	Management
I:	Nondiagnostic	25	Clinical and Radiographic Follow-Up / Repeat FNA / Ancillary studies
II:	Non-neoplastic	10	Clinical and Radiographic Follow-Up / Ancillary Studies
III:	AUS	20	Repeat FNA / Ancillary Studies / Surgery
IVa:	Neoplasm: Benign	<5	Surgery / Ancillary Studies / Clinical Follow-Up
IVb:	Neoplasm: SUMP	35	Surgery / Ancillary Studies
V:	Suspicious for Malignancy	60	Repeat FNA / Ancillary Studies / Surgery
VI:	Malignant	>90	Surgery / Ancillary Studies

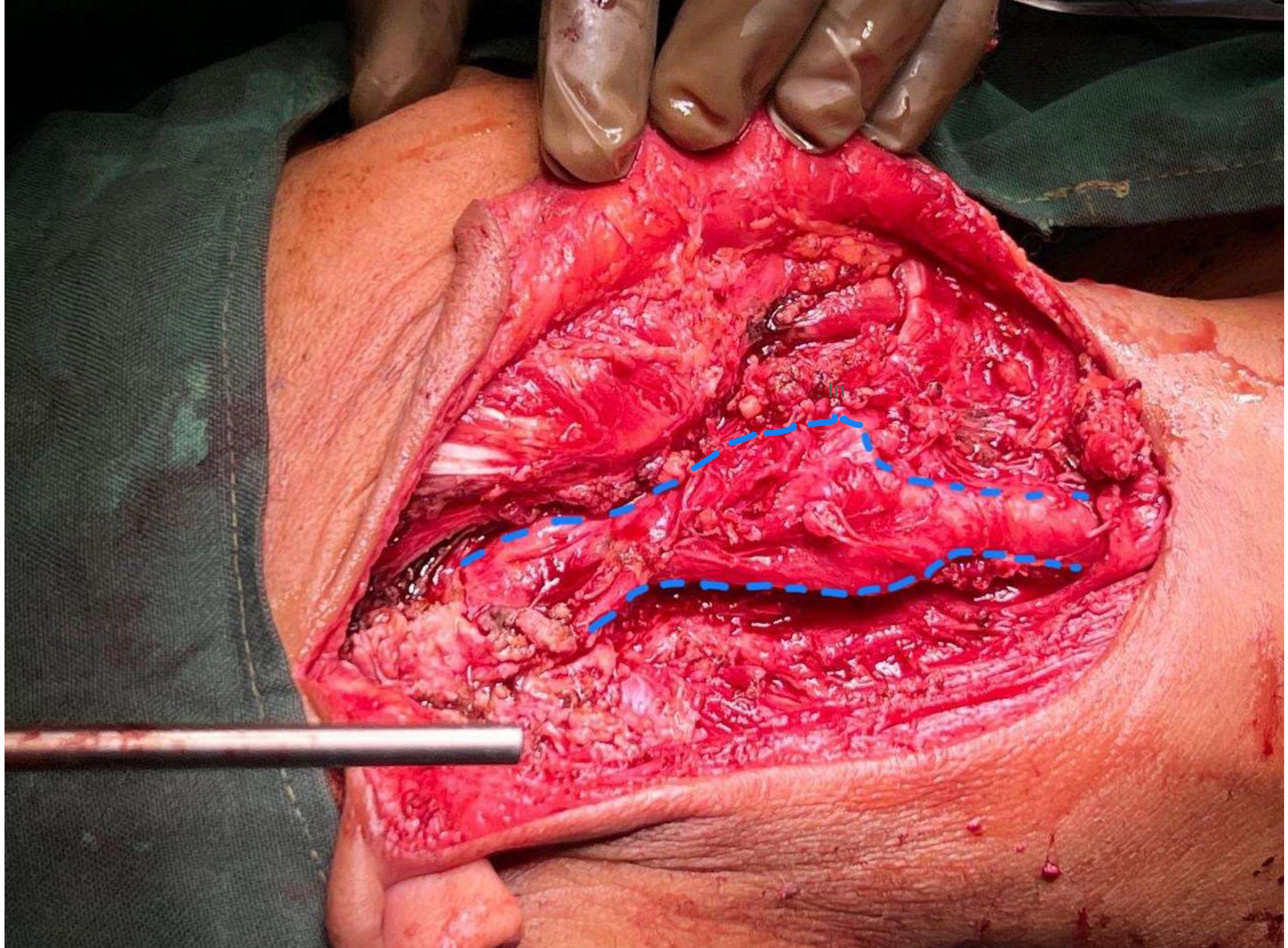
Abbreviations: AUS = atypica of undetermined significance, FNA = fine needle aspiration, SUMP = salivary gland neoplasm of uncertain malignant potential

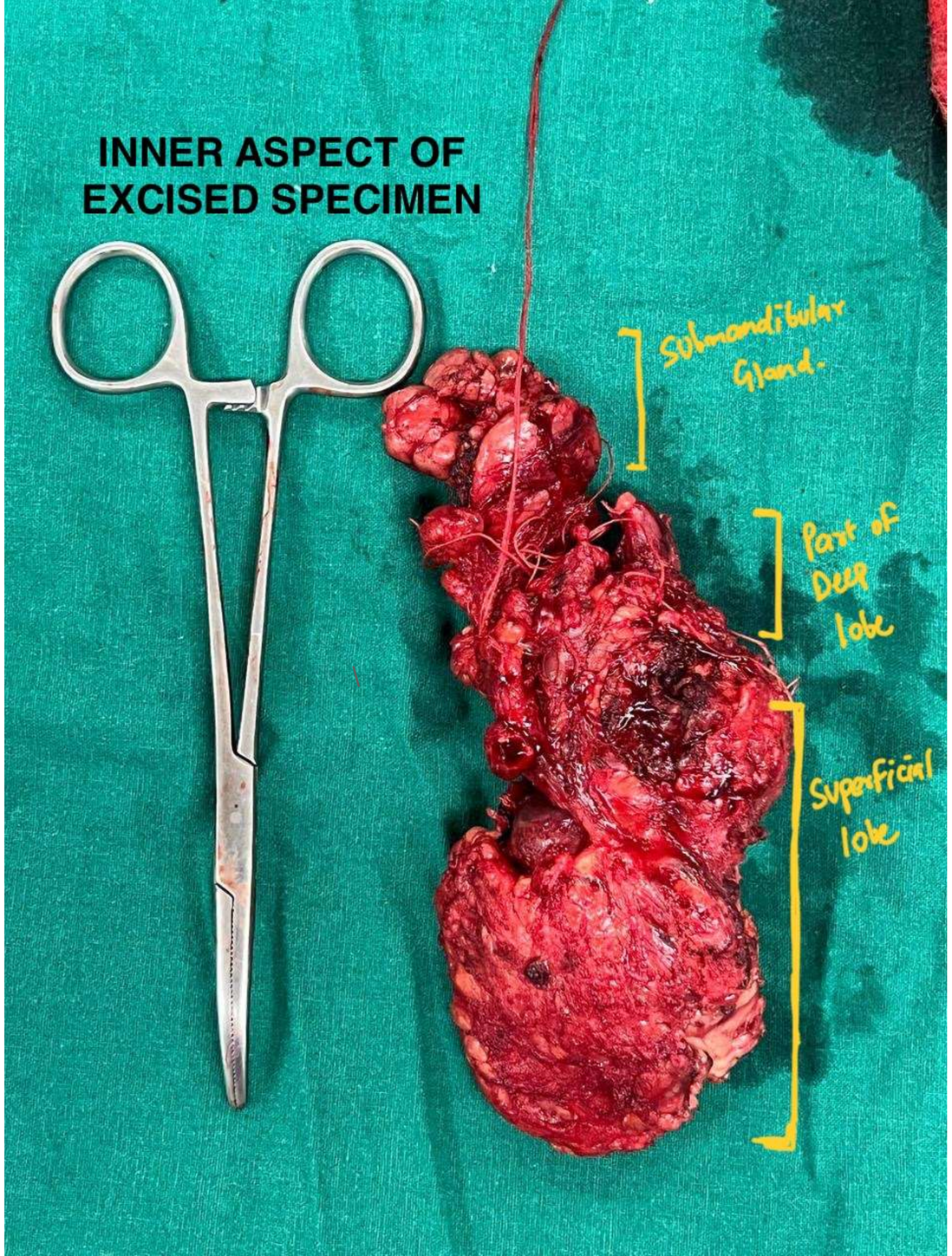

CERVICAL
BRANCH OF
FACIAL NERVE
CAROTID
SHEATH

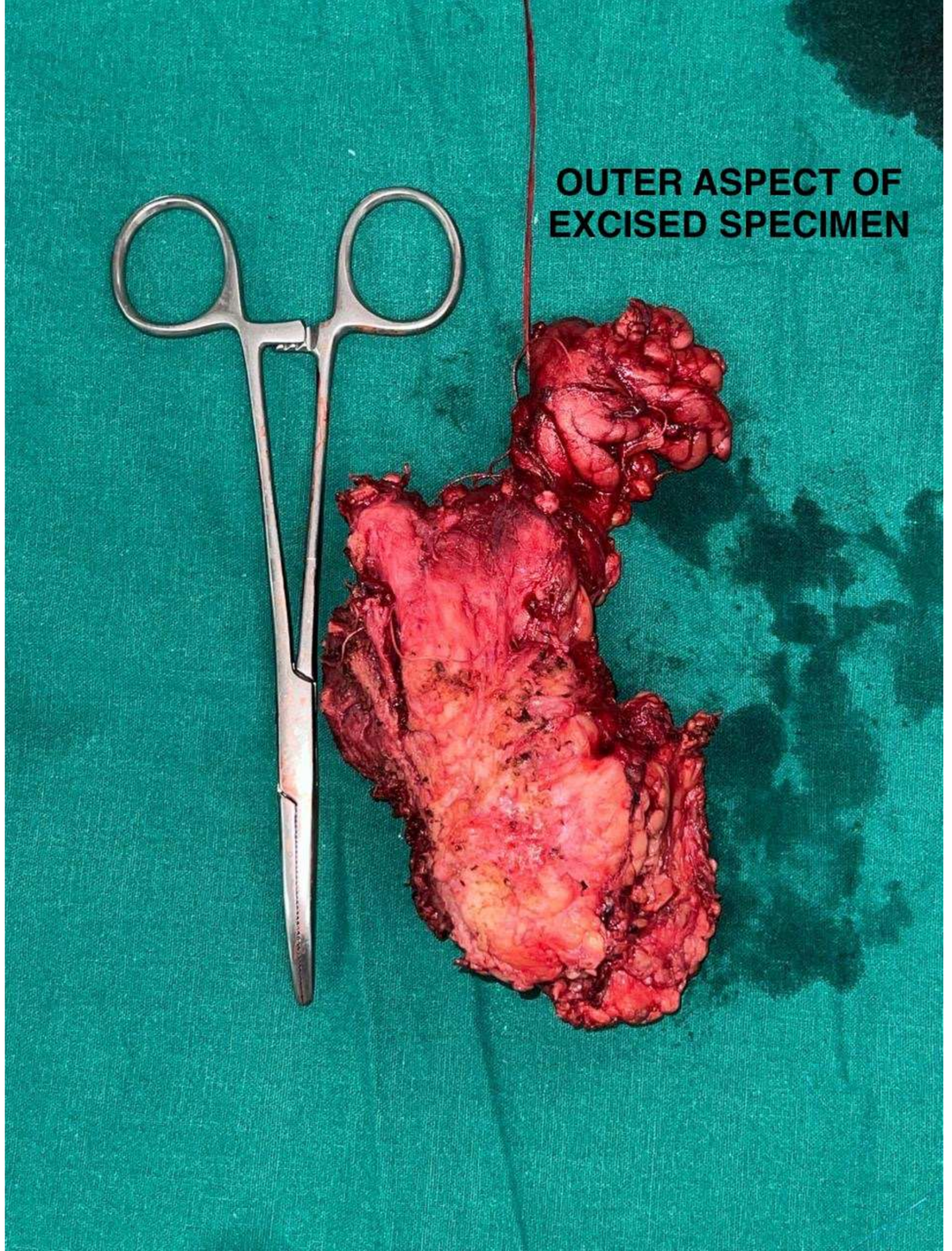

HAYES MARTIN
MANEUVER

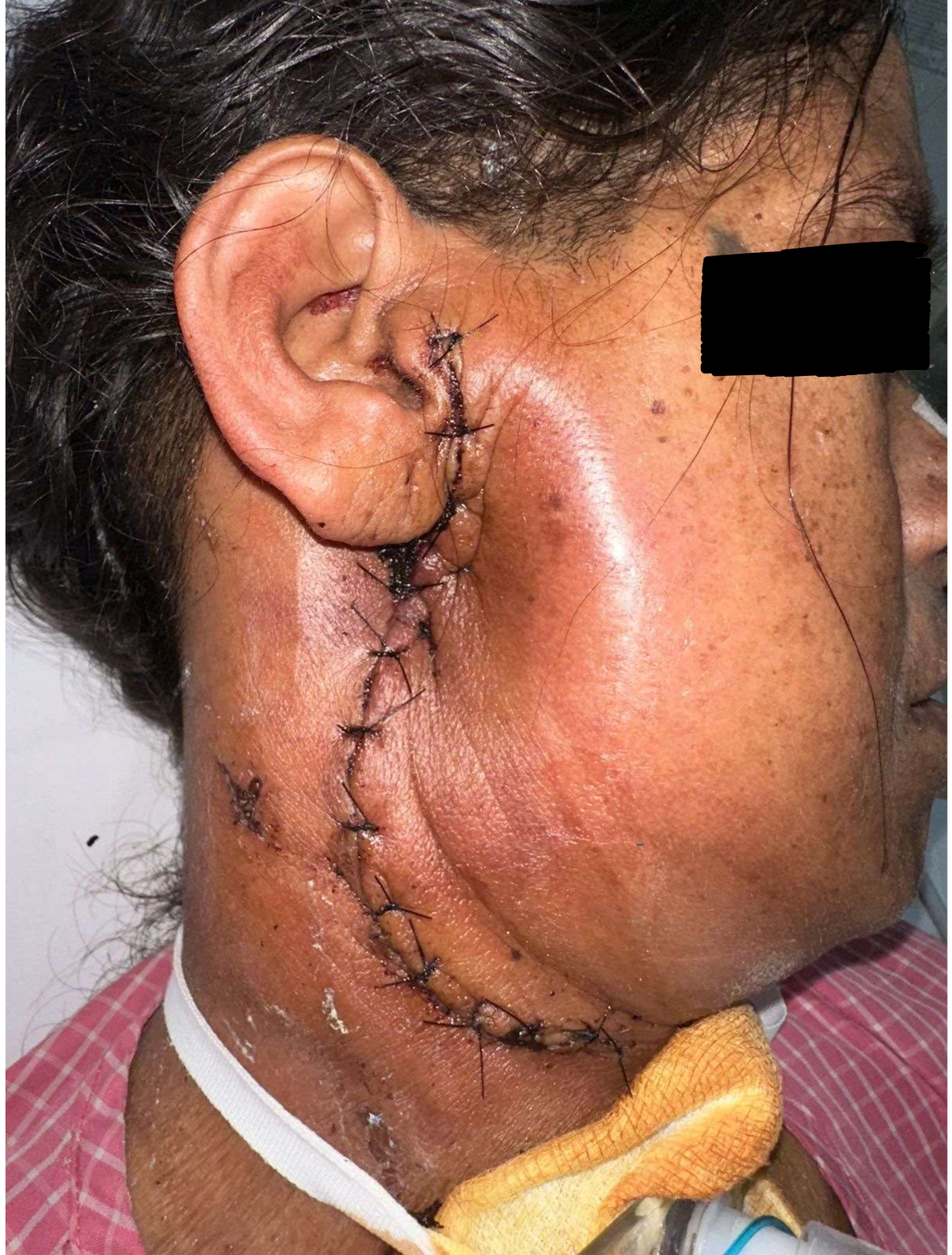

SUBMANDIBULAR
GLAND EXCISION



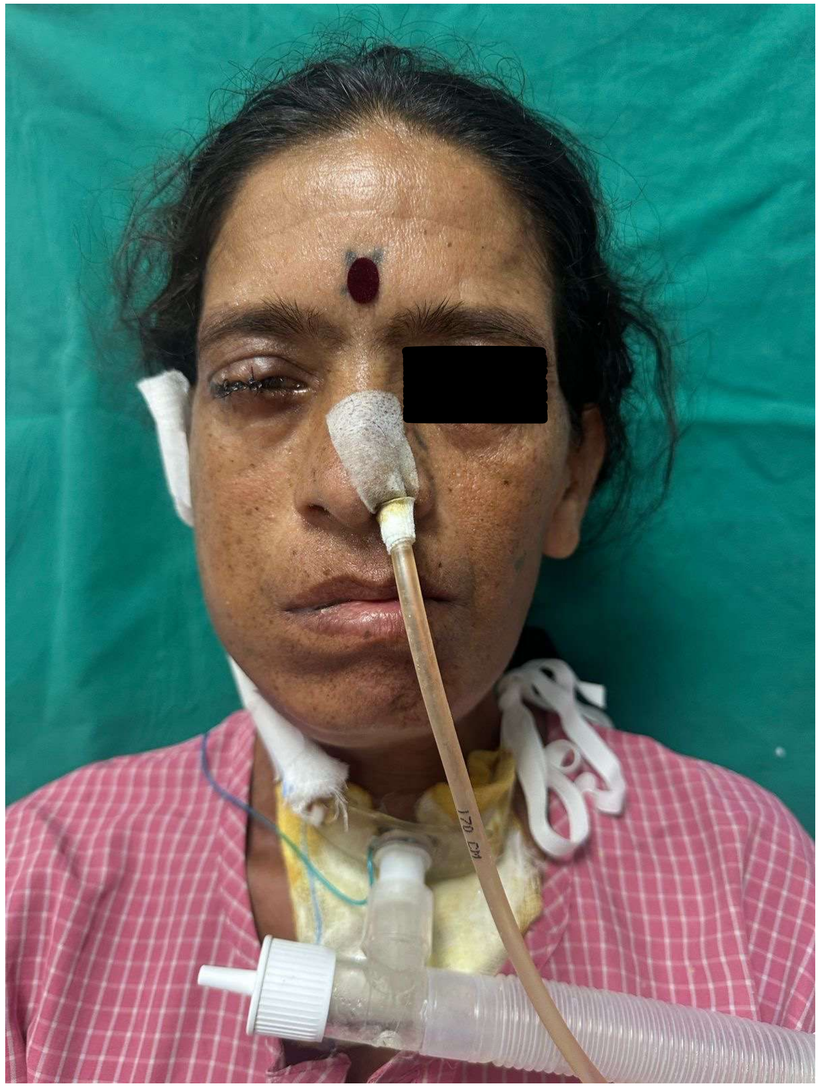


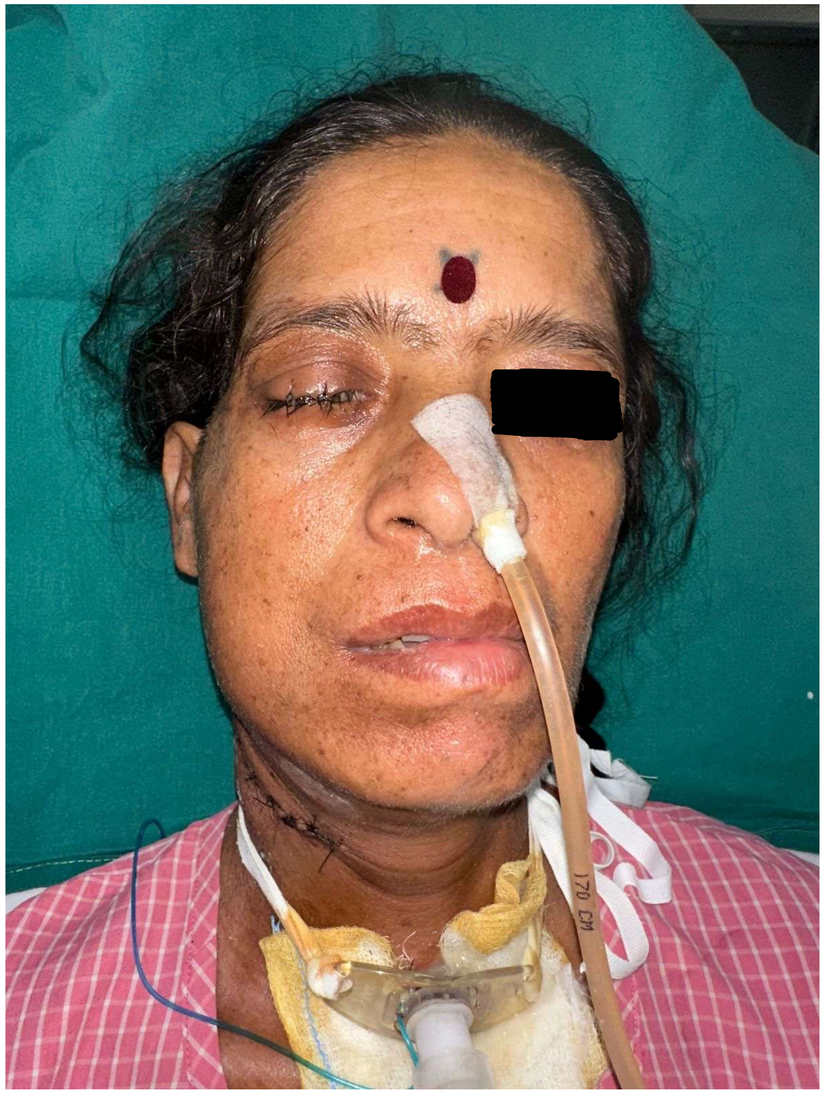

INNER ASPECT OF
EXCISED SPECIMEN
Submandibular Gland.
Part of Deep lobe
Superficial lobe


OUTER ASPECT OF
EXCISED SPECIMEN



POD-5





POD- 8

Histopathology of the specimen

Diagnosis -

Histological and IHC findings favour Extracranial Meningothelial Meningioma of parotid gland, WHO Grade I

IHC -

EMA - Diffuse strong positivity.

Vimentin - Diffuse strong positivity

S100 - Positive

PR - Diffuse strong nuclear positivity.

ki67 - upto 2%

p63 - Weak nuclear positivity (<2%)

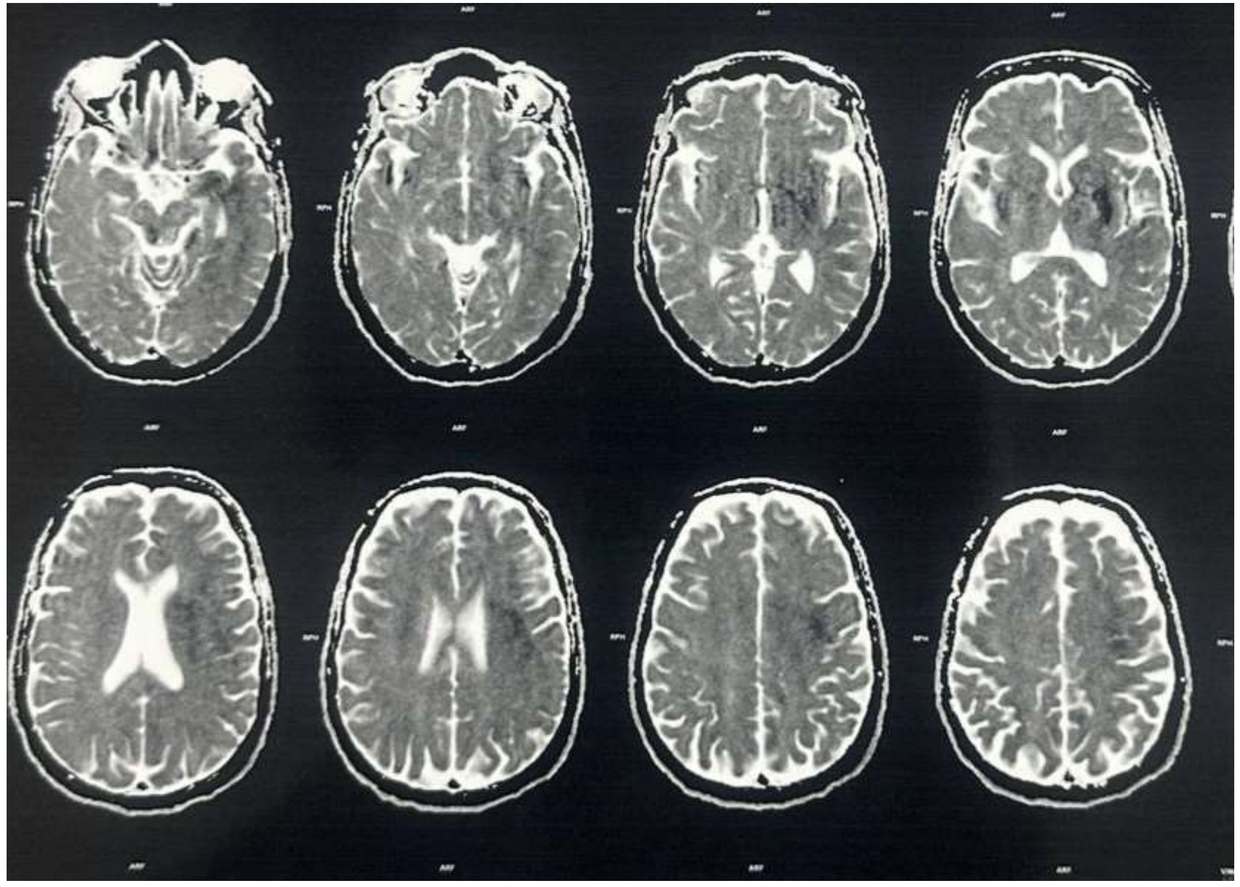
ck7 - Negative

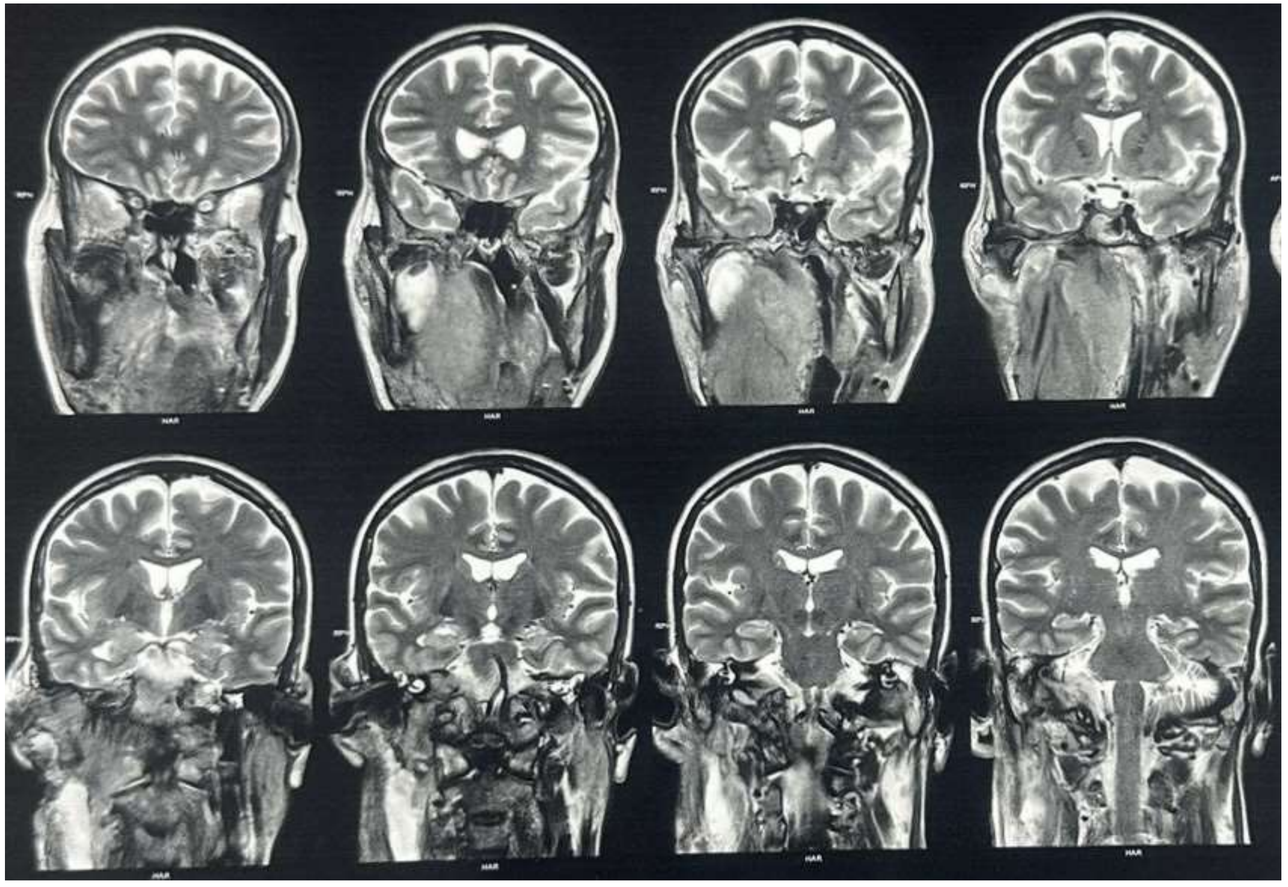
SMA - Negative

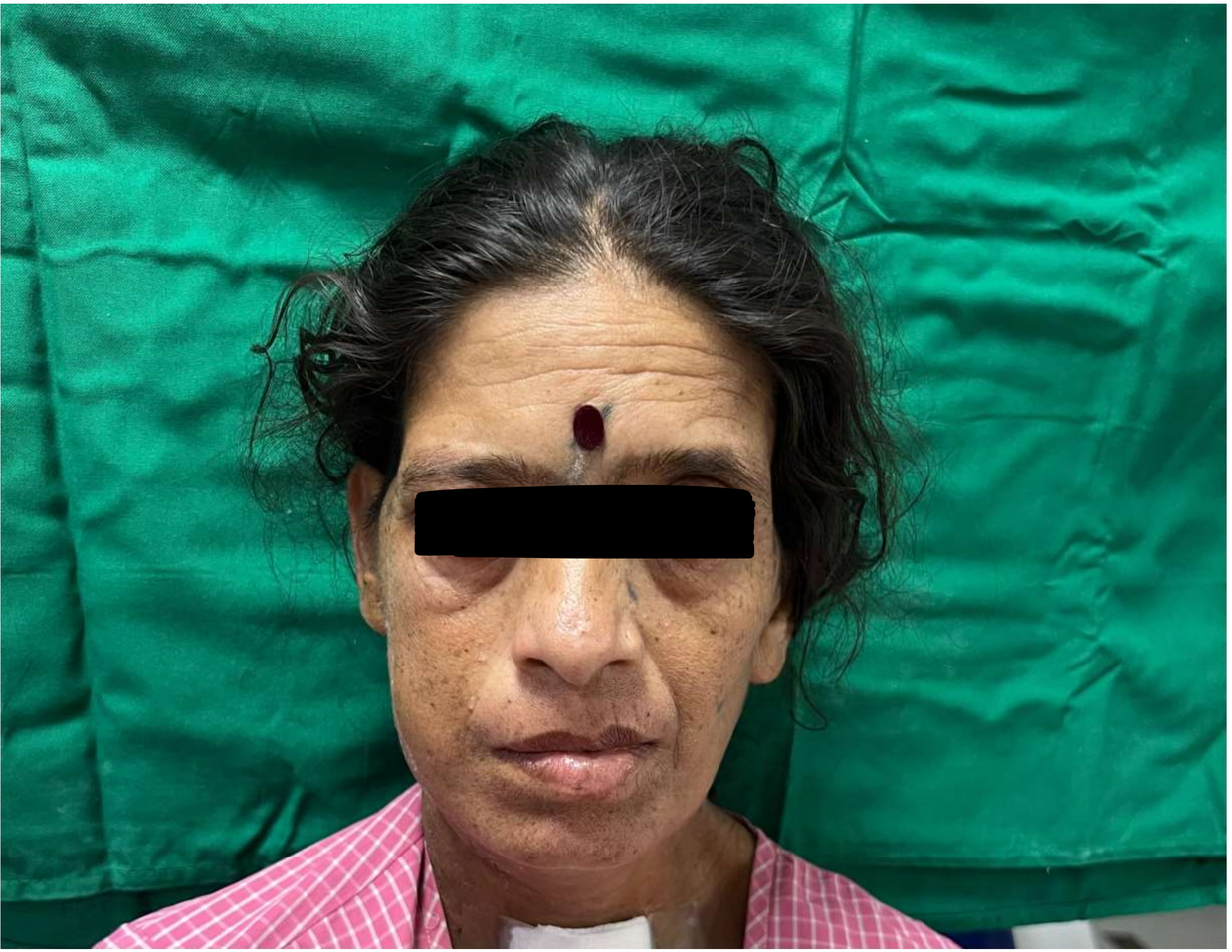
Calponin - Diffuse positive.

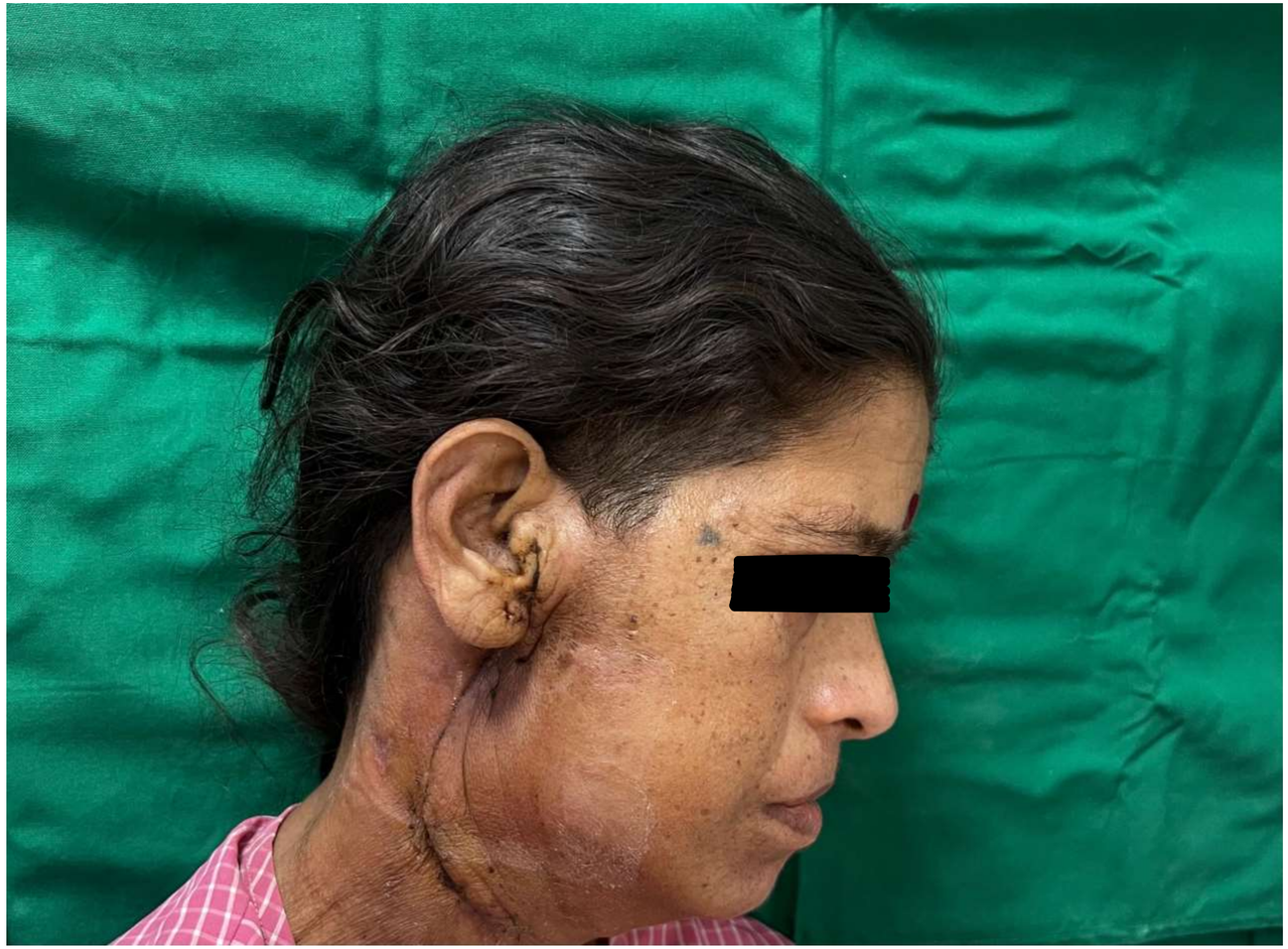
MRI brain

Brain was normal , no evidence of intracranial extension









POD- 21

Discussion

- Meningioma is a tumour arising from arachnoidal cells and, in the majority of cases, its behaviour is benign. It accounts for 24-30% of all intracranial tumours**[2]**
- In India meningeoma ranges from 11.6% to 21% of all brain tumors,
- 0.9% and 2% for extracranial meningiomas. **34 case reports have been published with meningeoma in sites such as hard palate , maxilla, cheek , occipital region and parotid , where as meningiothelial meningioma is reported only in 7 case reports[3], which makes our case extremely rare**

2. DN, Ohgaki H, Wiestler OD, Cavanee WK, editors. WHO Classification of Tumours of the Central Nervous System. Lyon: International Agency for Research on Cancer; 2007. pp. 164–172.

3.Umana, GE, Scalia G, Vats A, Pompili G, Barone F, Passanisi M, et al. Primary extracranial meningiomas of the head and neck. Life 2021;11:942.

- The pathophysiology of extracranial meningioma’s has been associated with defects of cell migration from the neural crest. However, several possible mechanisms have been proposed: origin from arachnoid cells of nerve sheaths protruding from the skull foramina, ectopic arachnoid granulations related to traumatism, intracranial hypertension (which could cause the movement of groups of arachnoid cells),oral surgery or finally, a possible origin from undifferentiated mesenchymal cells[4-5].

  According to Hoye classification, this case is (Type C - Ectopic with no connection to a foramen of a cranial nerve or to intracranial structures).

4. Serry, P.; Rombaux, P.; Ledeghen, S.; Collet, S.; Eloy, P.; Hamoir, M.; Bertrand, B. Extracranial sinonasal tract meningioma: A case report. Acta Oto-Rhino-Laryngol. Belg. 2004, 58, 151–155.

5. Tokgoz N, Oner YA, Kaymaz M et-al. Primary intraosseous meningioma: CT and MRI appearance. AJNR Am J Neuroradiol. 2005;26 (8): 2053-6

2016 WHO classification system for grading meningiomas (histopathological)

Grade 1

- Meningothelial
- Fibrous
- Transitional
- Psammomatous
- Angiomatous
- Microcystic
- Secretory
- Lymphoplasmacyte-rich
- Metaplastic

Grade 2

- Atypical
- Chordoid
- Clear cell

- 4–19 mitoses per 10 high-power fields (HPF)
- Brain invasion
- At least 3 of :
  - *high cellularity*
  - *high nuclear-to-cytoplasmic ratio*
  - *sheeting*
  - *prominent nucleoli*
  - *spontaneous necrosis*

Grade 3

- Anaplastic (malignant)
- ~~Papillary~~ (removed in 2021 CNS5)
- ~~Rhabdoid~~(removed in 2021 CNS5)

- ≥ 20 mitoses per 10 high-power fields (HPF)
- Overtly malignant cytology:
  - *carcinomatous*
  - *Sarcomatous*
  - *melanomatous*

2021 WHO classification (addition of genetic markers)

- *TERT* promoter mutation
- Homozygous deletion of *CDKN2A and/or CDKN2B*

**As per the new classification, papillary and rhabdoid meningiomas can be grade 1, 2 or 3 and should not be graded based on histology alone*

- Immunohistochemistry is used to differentiate between various types of tumors, such as neurofibroma, Schwannoma, soft tissue perineuroma, paragangliomas, and solitary fibrous tumor.**[6]**

- Extracranial meningiomas, expressing EMA and vimentin, may suggest aggressive behavior but have no histopathological correlation with malignancy. Low Ki-67 indicates differentiation from benign and malignant neoplasms.**[7]**

6. Boulagnon-Rombi C, Fleury C, Fichel C, Lefour S, Marchal Bressenot A, Gauchotte G. Immunohistochemical approach to the differential diagnosis of meningiomas and their mimics. J Neuropathol Exp Neurol 2017;76:289-98

7. Fraioli, M.F.; Marciani, M.G.; Umana, G.E.; Fraioli, B. Anterior Microsurgical Approach to Ventral Lower Cervical Spine Meningiomas: Indications, Surgical Technique and Long Term Outcome

- Extracranial meningioma have a favorable clinical outcome
- Radical removal is the gold standard to prevent residual tumor growth. Histological investigation is crucial for diagnosis, and frozen sections confirm benignity. **In our case impression on frozen section was low grade infiltrating tumor.**
- Inoperable or recurrent tumors can be treated with fractionated radiotherapy, targeting somatostatin receptors and systemic pharmacotherapy have shown modest results, but no standard of care exists. **[8]**

8. Goldbrunner R, Stavrinou P, Jenkinson MD, Sahm F, Mawrin C, Weber DC, Preusser M, Minniti G, Lund-Johansen M, Lefranc F, Houdart E, Sallabanda K, Le Rhun E, Nieuwenhuizen D, Tabatabai G, Soffietti R, Weller M. EANO guideline on the diagnosis and management of meningiomas. Neuro Oncol. 2021 Nov 2;23(11):1821-1834. doi: 10.1093/neuonc/noab150. PMID: 34181733; PMCID: PMC8563316

Challenges faced

- Diagnosis

- Intraoperatively
  - Main trunk of facial nerve could not be idenified.
  - Infiltration of tumor to surrounding structure
  - Frozen section

- Uncommon presentation in the final Histopathology.

Thank you