Case of the Month

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Dr. VIDYA MENON
Senior Resident
Department of Pathology, RGCIRC
New Delhi

Case

- A 64-year-old male presented with complaints of left nasal cavity blockage, increased watering from the left eye and proliferative growth over the left upper alveolus since 3 months

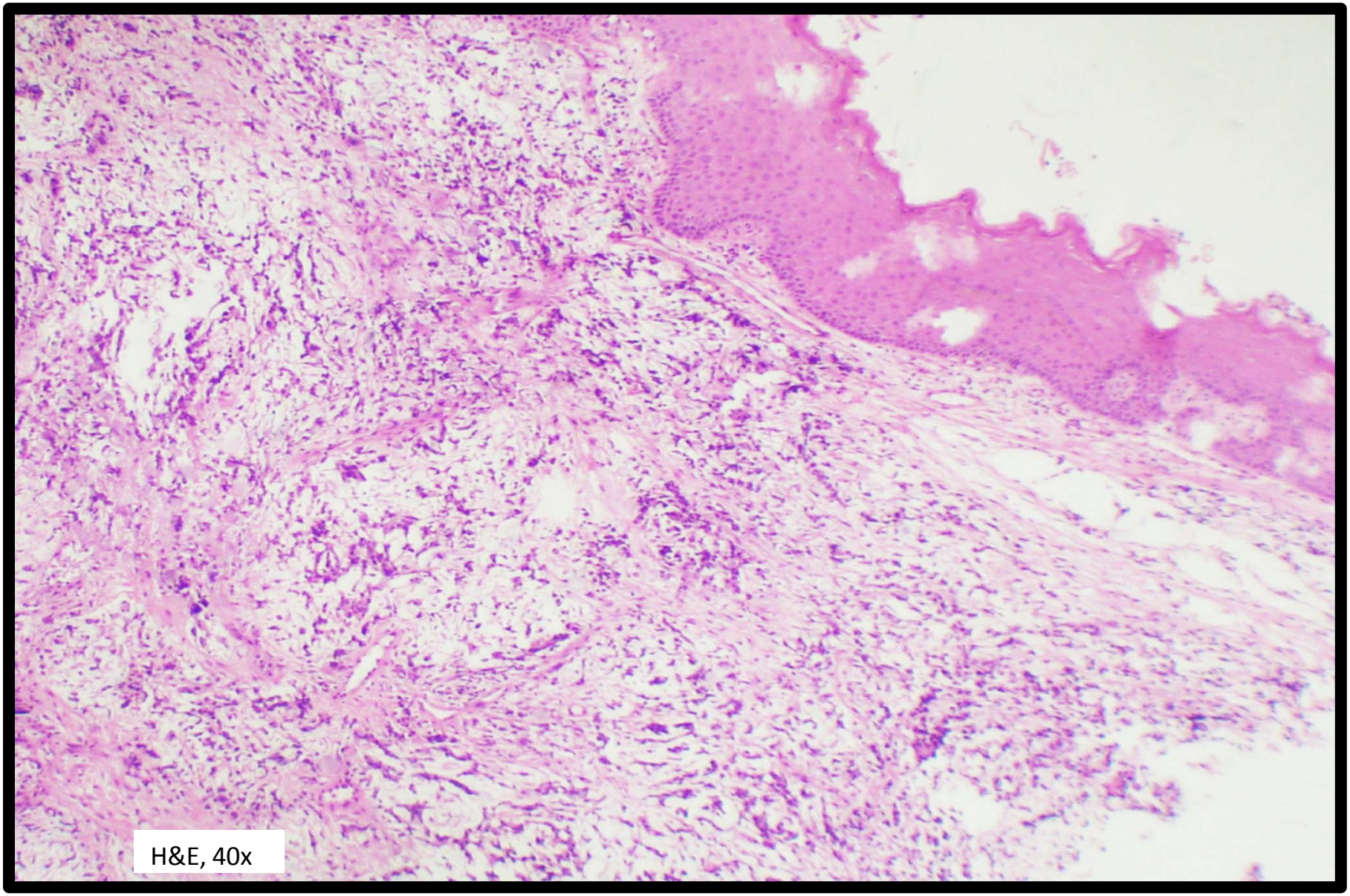
- On examination, a proliferative growth was noted around 6 x 4 cm involving the left upper alveolus extending 1.5 cm from the central incisor region upto the retromolar trigone posteriorly, 0.5 cm from midline over the palate upto the upper gingivobuccal sulcus laterally

Case

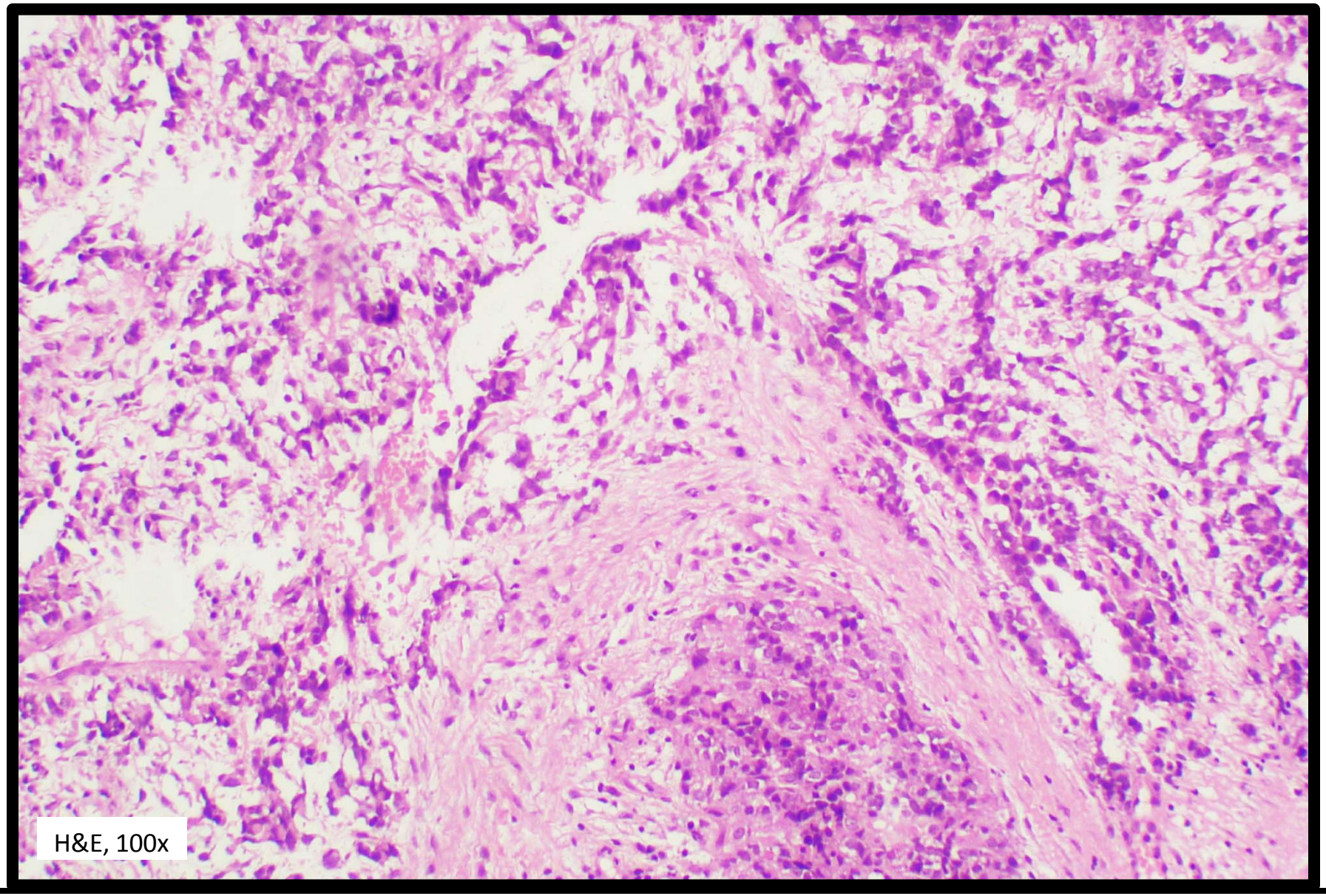
- Ptosis and proptosis were present in the left eye
  A diffuse bulge was present in left cheek

- Subsequently, a biopsy from the palatal extension of the mass was performed

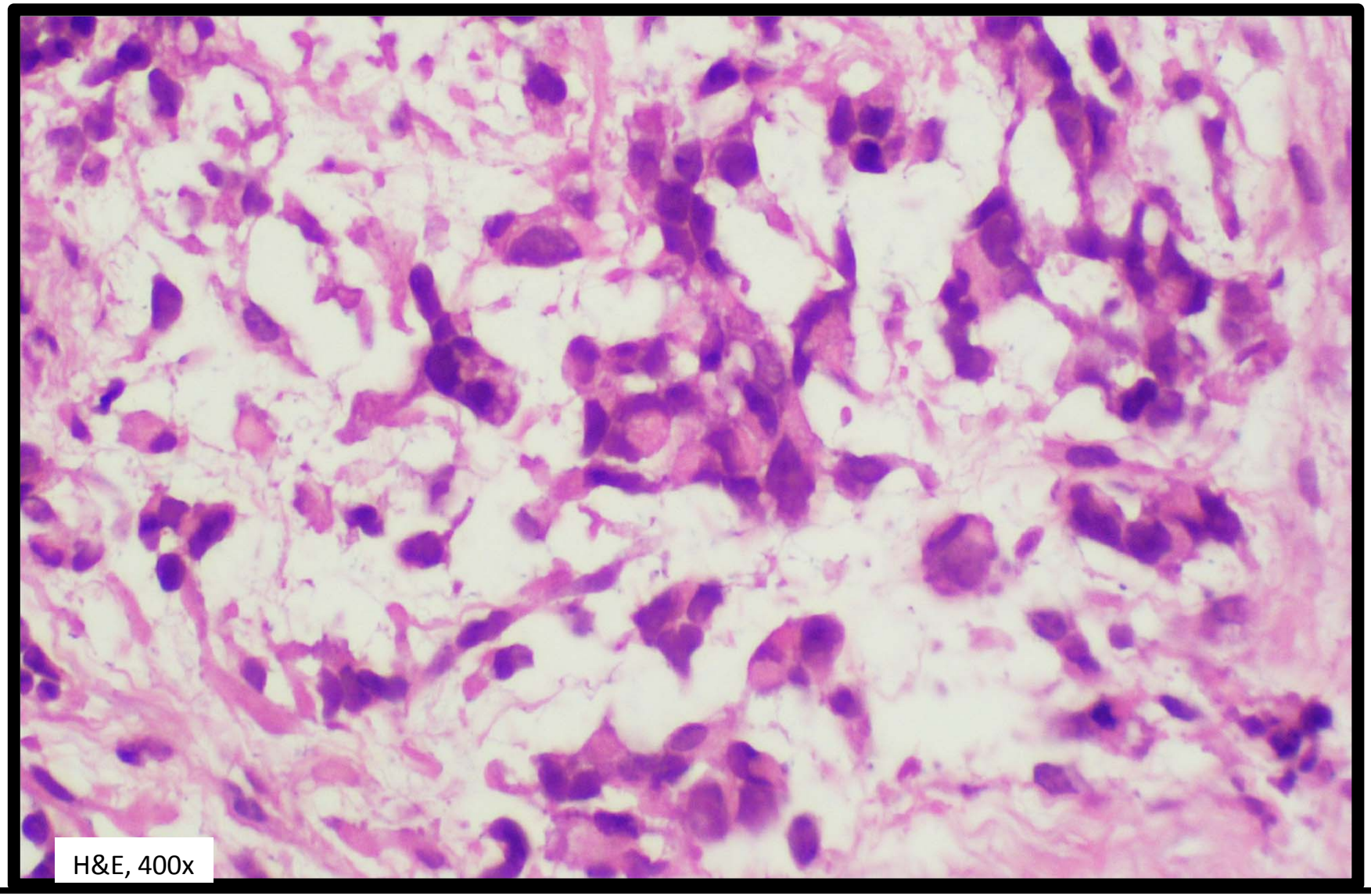
Microscopic Findings



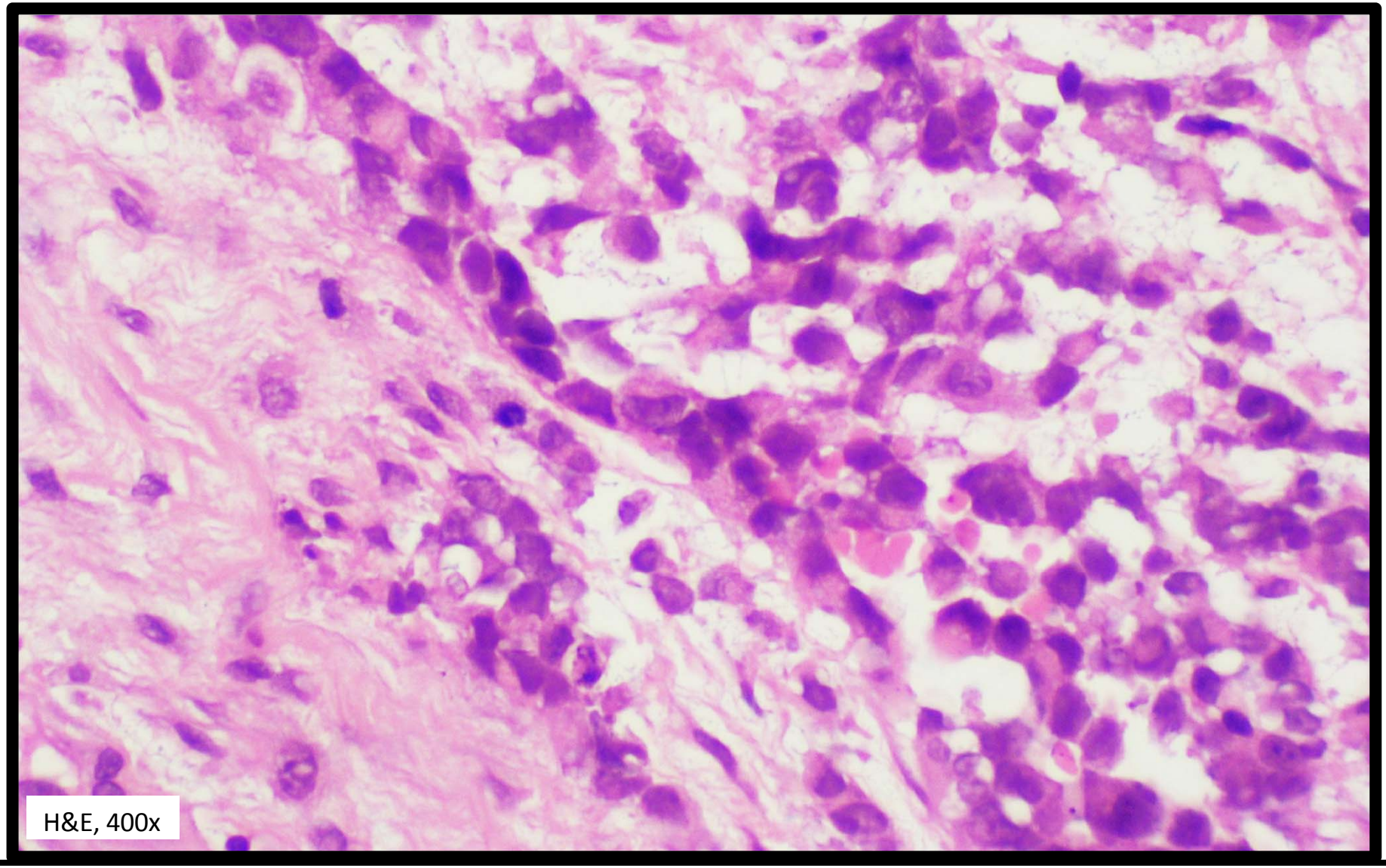
Biopsy showed partially ulcerated squamous mucosa with underlying stroma extensively infiltrated by a poorly differentiated malignancy. The stroma showed significant myxoid change



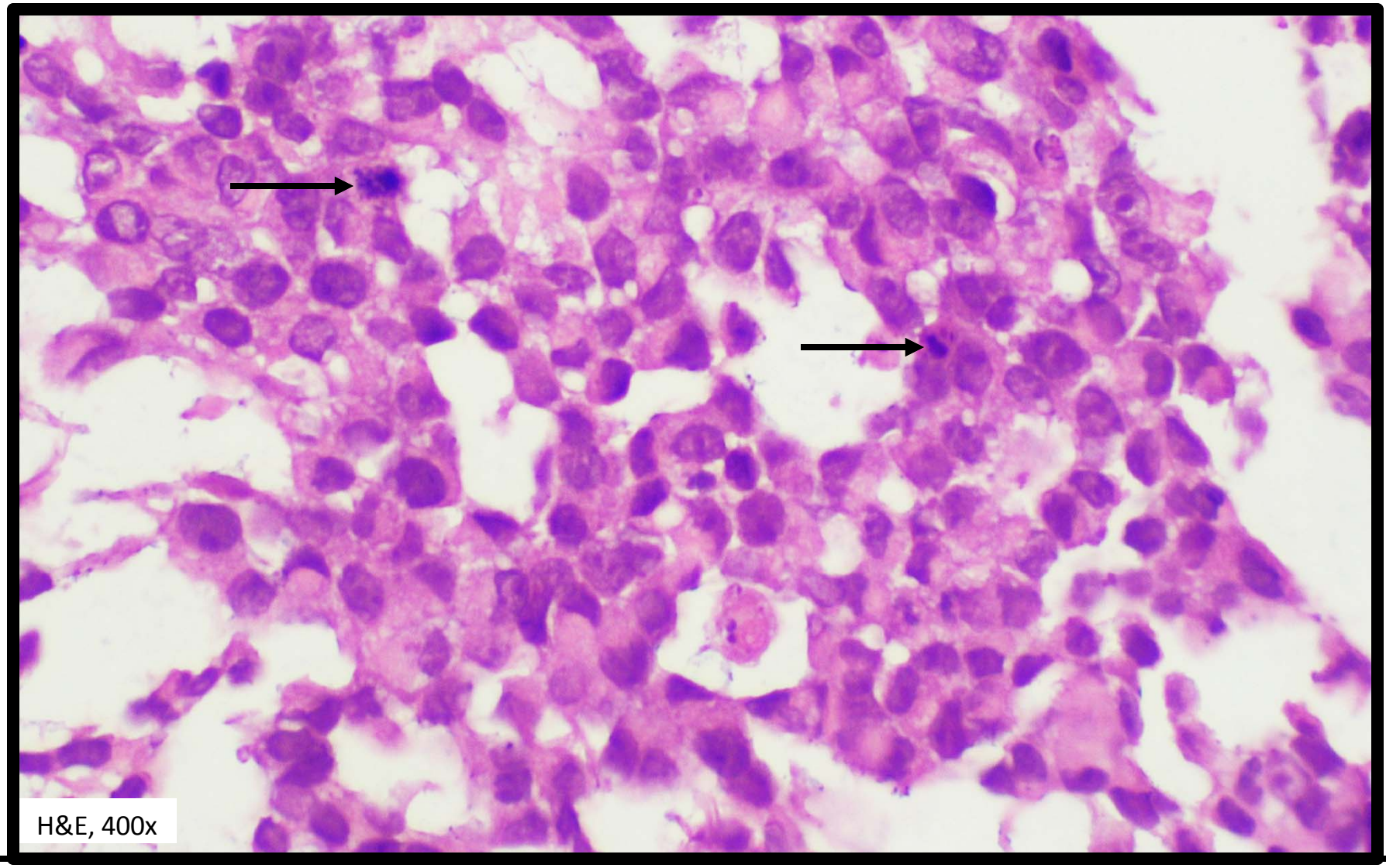
The neoplasm showed a biphasic morphological pattern comprising of polygonal cells and spindle shaped cells



An area showing predominantly polygonal cells with few spindle shaped cells



The tumor cells exhibit marked nuclear atypia .

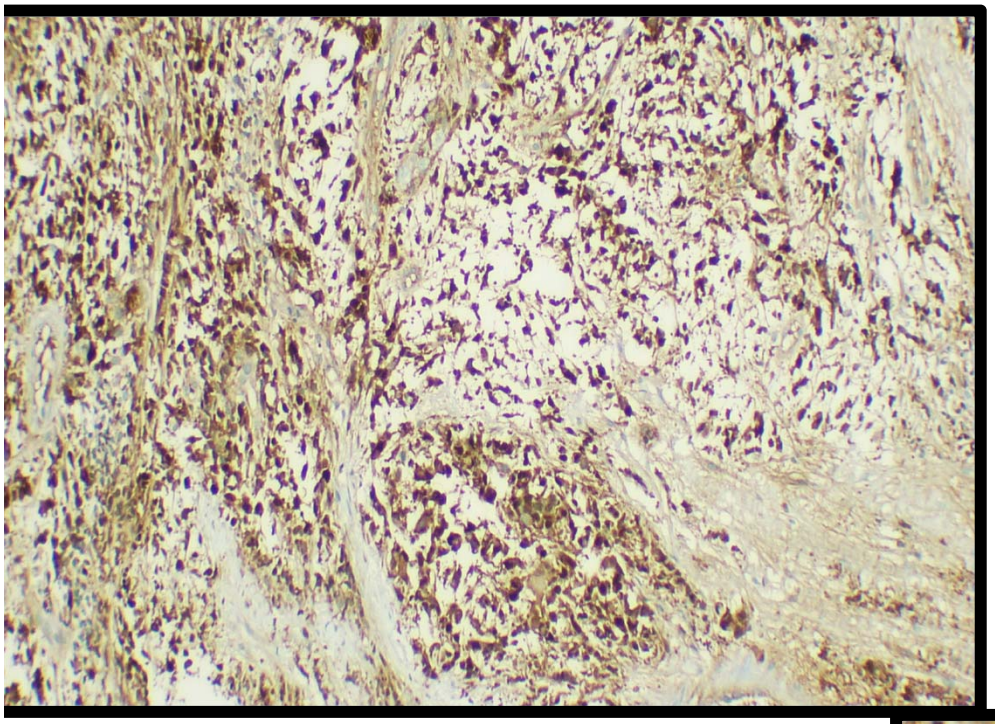


The tumor cells exhibit significant mitotic activity (arrows)

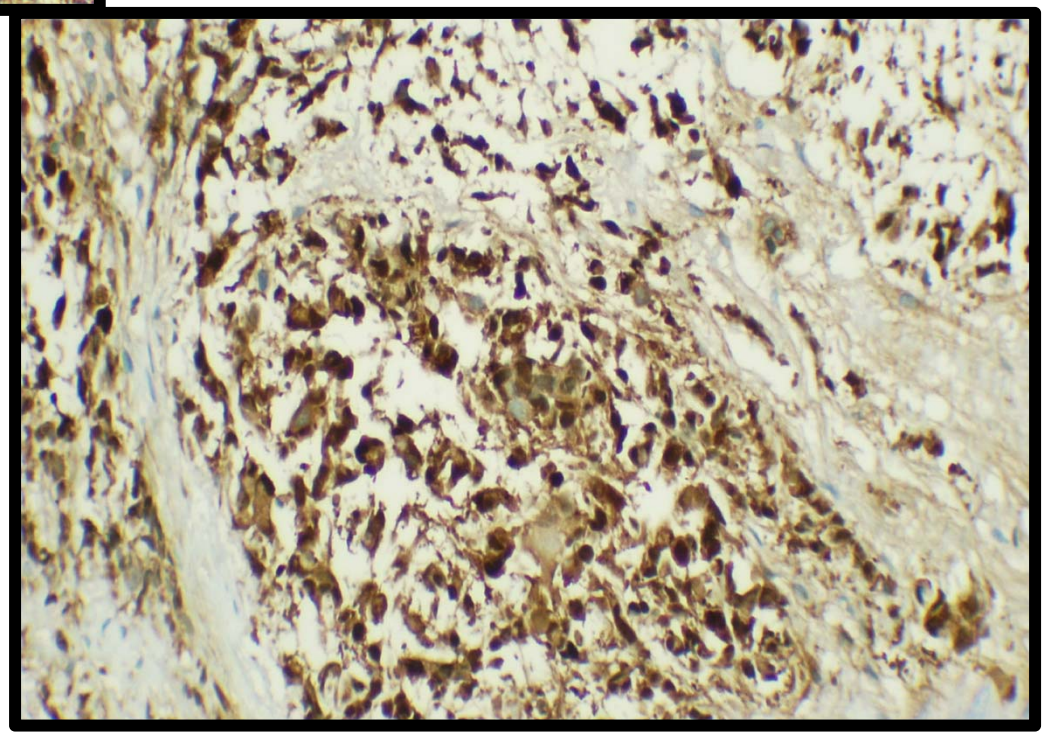
Findings:

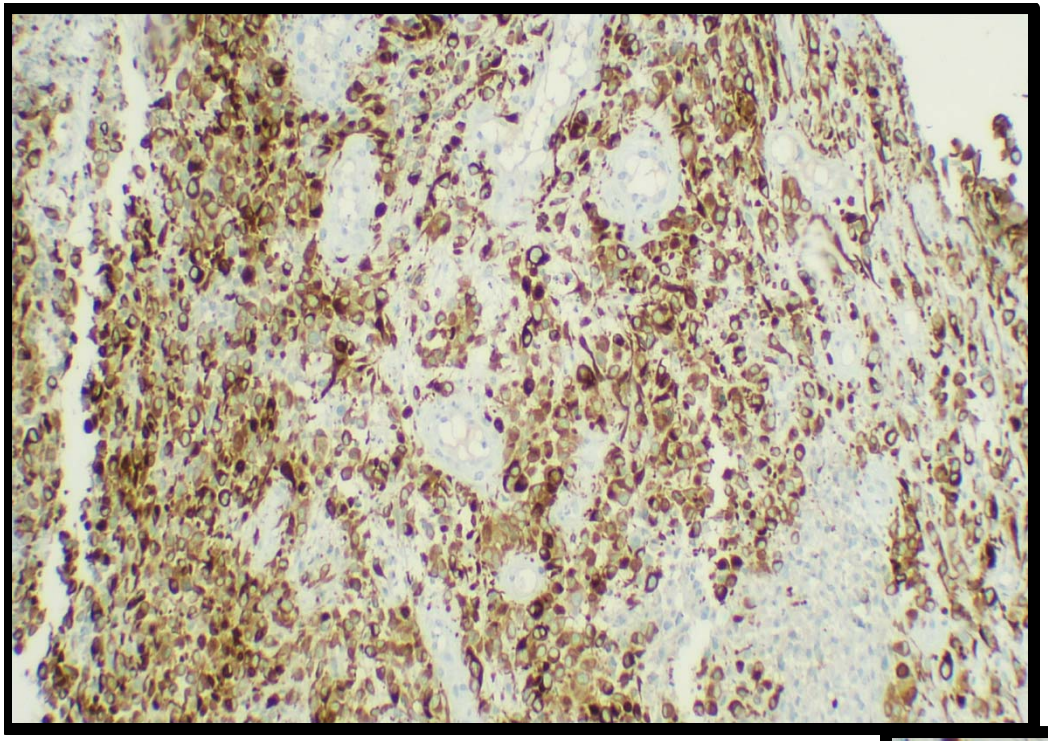
Based on these findings, a diagnosis of a high grade malignancy was made

Immunohistochemistry

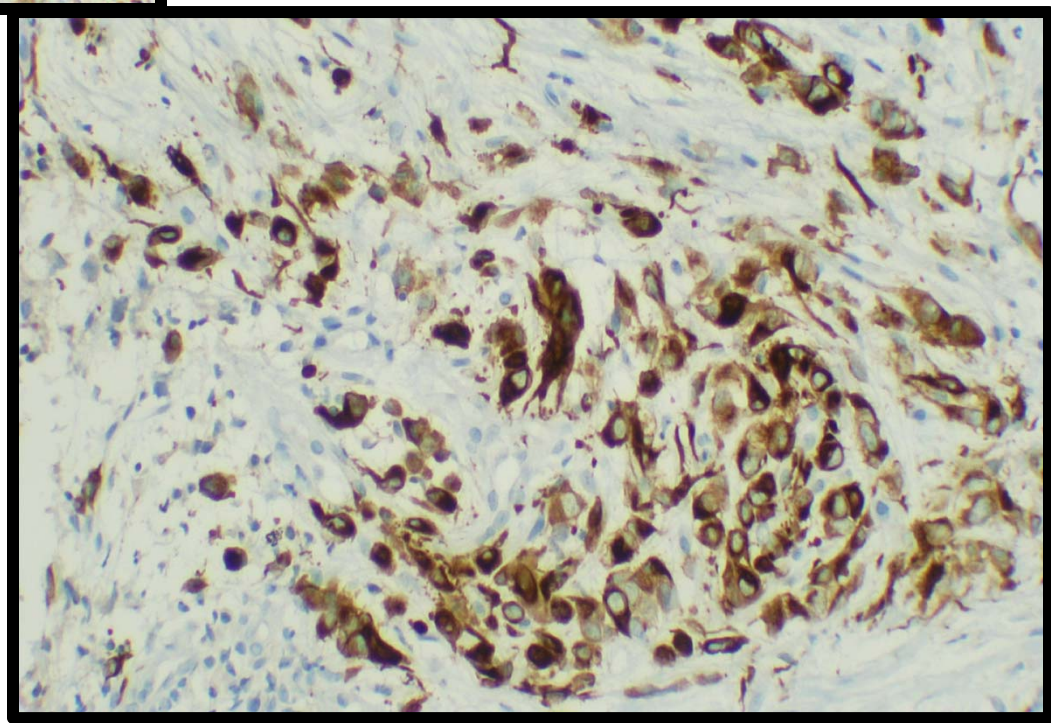


S100: Diffusely and strongly positive

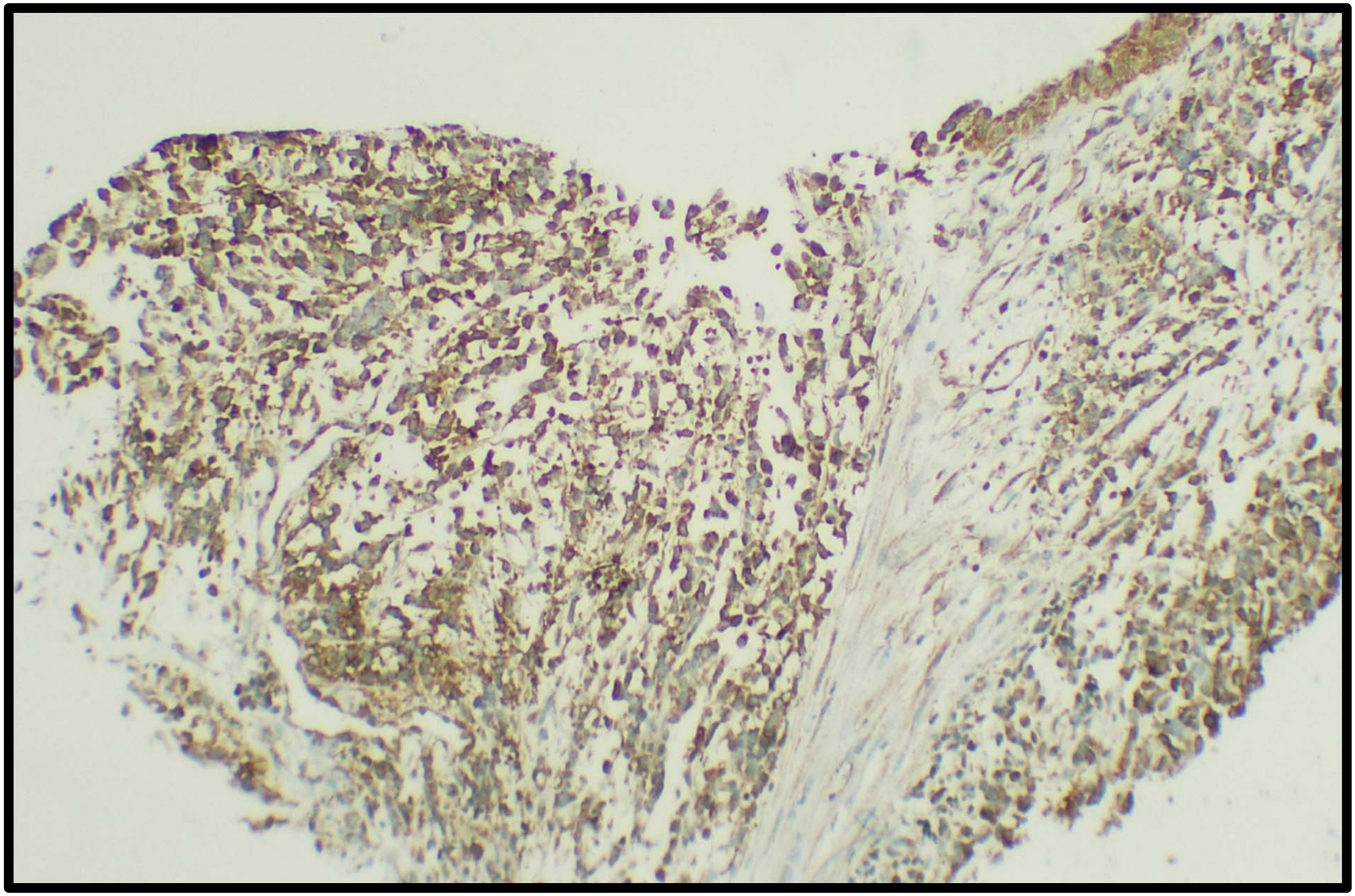




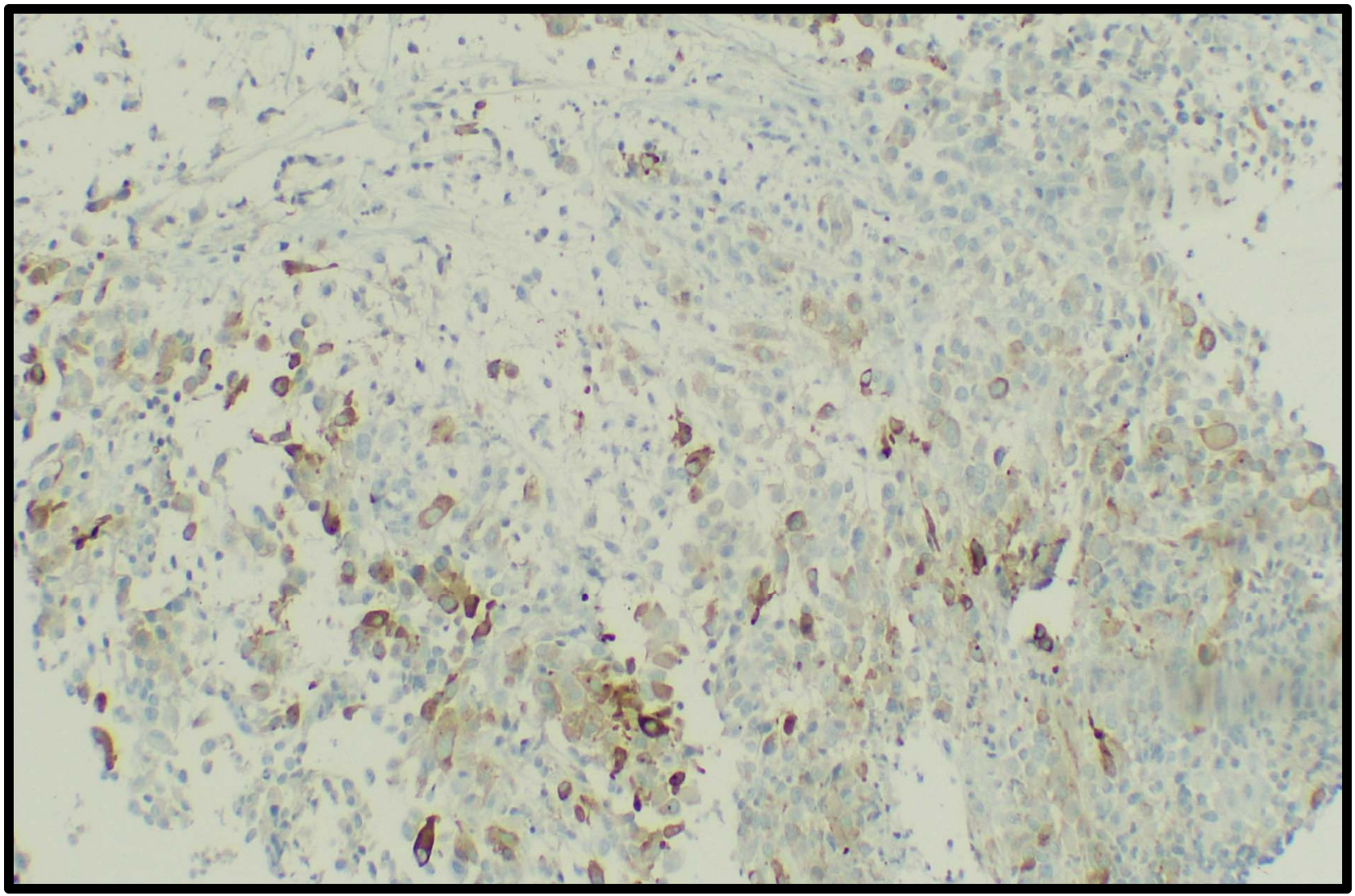
Heterogenously positive for CK (immunoexpression evident in the areas showing rhabdoid differentiation)

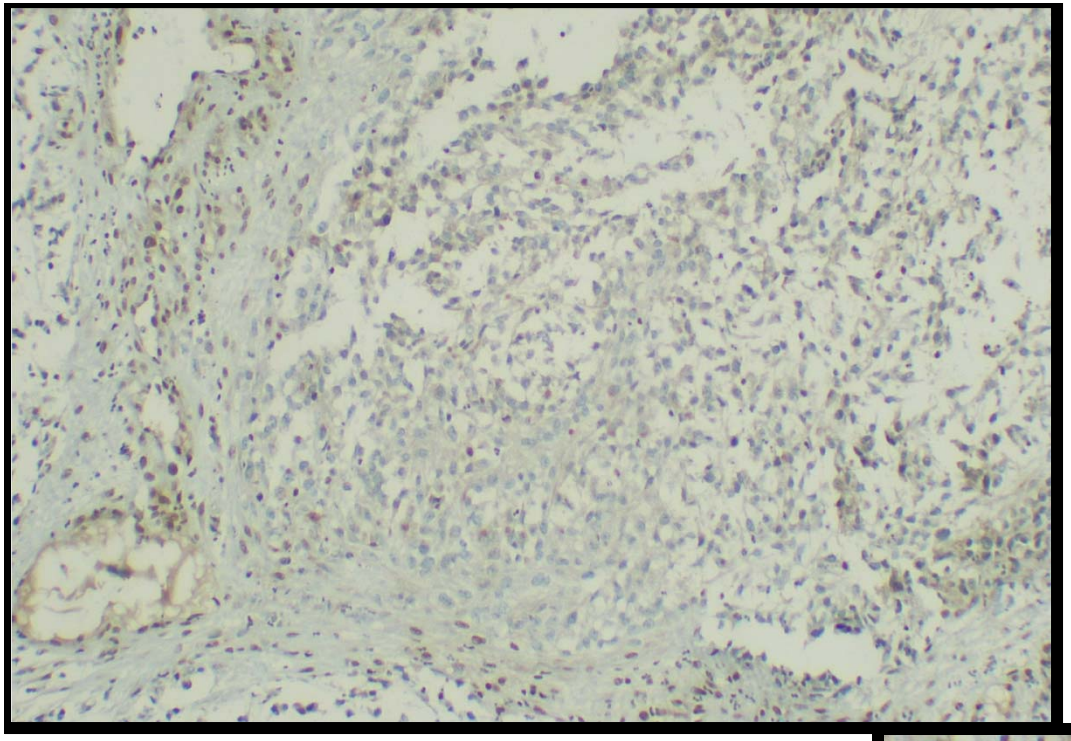


SMA: Positive

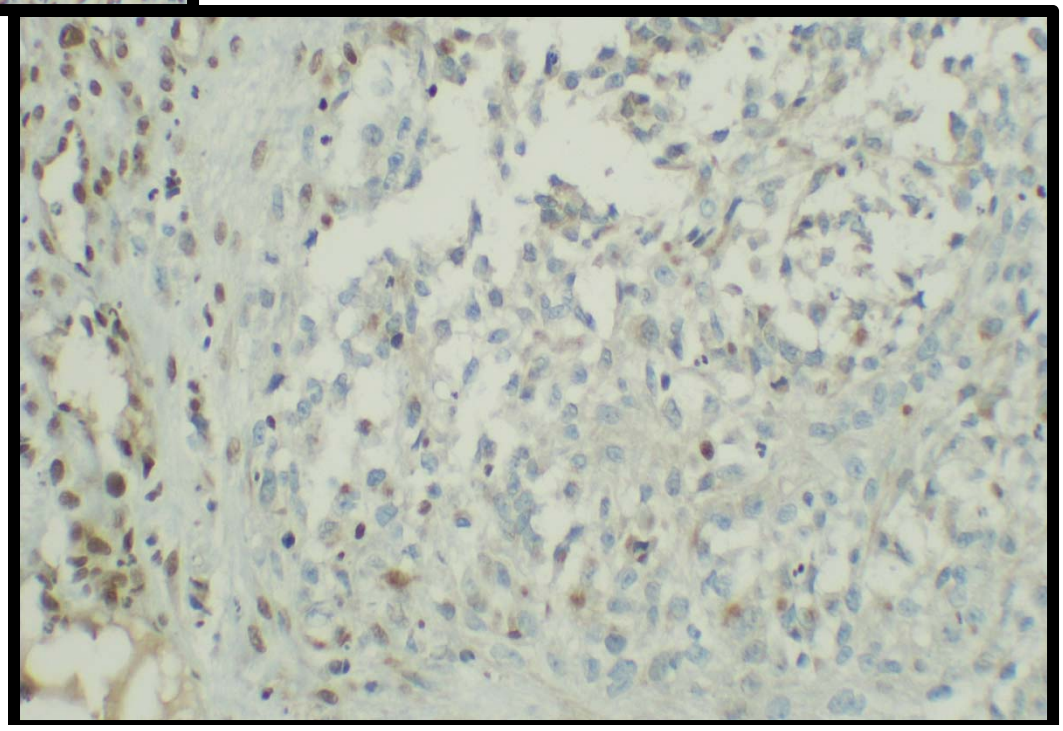


Calponin: Positive





INI1 expression: lost in tumor cells



IHC Findings

The tumor cells were negative for p40, CD34, SOX10 and MelanA

Summary of IHC findings:

Positive

- S 100
- CK
- SMA
- Calponin

Negative

- INI1
- P40
- CD34
- SOX-10
- MELAN-A

The Final Diagnosis

High grade myoepithelial carcinoma with rhabdoid differentiation

Discussion (1/5)

- Myoepithelial neoplasms represent a clinically, behaviourally and histologically varied group

- These, along with mixed tumors (which contain ductal or glandular elements) have been well characterized in the salivary glands but are increasingly documented in a range of other sites, including those in which myoepithelial cells are normally found in relation to various ductal structures (such as the skin, lung, breast and larynx)

- These have also been documented where myoepithelial cells are usually absent, such as superficial and deep soft tissues and bone

Discussion (2/5)

- Myoepithelial tumors, either benign or malignant, account for about 1% of all primary salivary gland tumors
- Approximately 70% and 20% of them arise in the parotid and submandibular glands, respectively, whereas 10% in the minor salivary glands
- Approximately 10% of all myoepithelial tumors are myoepithelial carcinomas
- Notably, myoepithelial carcinomas arising in unusual head and neck locations have been reported, such as nasal cavity , nasopharynx and maxillary sinus

Discussion (3/5)

- Their unifying feature is the morphologic and immunohistochemical evidence of myoepithelial differentiation, but this encompasses a large spectrum of different histologic appearances and varying immunoprofiles

- Myoepithelial tumors express S100 protein, cytokeratins and EMA with varying combinations of SMA, calponin, CD10, p63 and glial fibrillary acidic protein

- The immunophenotype varies markedly between cases, but a minimal requirement might be expression of S100 protein and an epithelial marker

Discussion (4/5)

- There are two myoepithelial carcinomas with rhabdoid features described in literature affecting the breast and vulva and one soft tissue myoepithelial carcinoma of the neck with rhabdoid morphology, which showed loss of nuclear SMARCB1 (INI-1) immunoexpression and EWSR1 rearrangement by fluorescence ISH

- Noteworthy, EWSR1-rearranged myoepithelial carcinomas of salivary gland origin can also be observed

Discussion (5/5)

- The cytological features in myoepithelial tumors includes epithelioid, spindle, hyaline (plasmacytoid), clear and mixed cell types with myoepithelioma occasionally presenting basaloid and oncocytic cytoplasmic features

- A mucinous variant of myoepithelial tumors has also been recognized

Conclusion

- In summary, although rare, myoepithelial carcinoma with rhabdoid features should be considered in the differential diagnosis when assessing malignant neoplasms in unusual head and neck locations

THANK YOU