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ANSWER: ANGIOSARCOMA OF EXTERNAL EAR CANAL.

Biopsy of the EAC mass performed under local anaesthesia revealed squamous epithelium, ulcerative malignant cells with anastomosing vessels (Figure 2) suggestive of angiosarcoma with positive CD31 and Ki67 of 80%. CT Brain till abdomen showed no signs of metastasis. Patient was referred to the Oncology unit for chemoradiotherapy as she refused surgery.

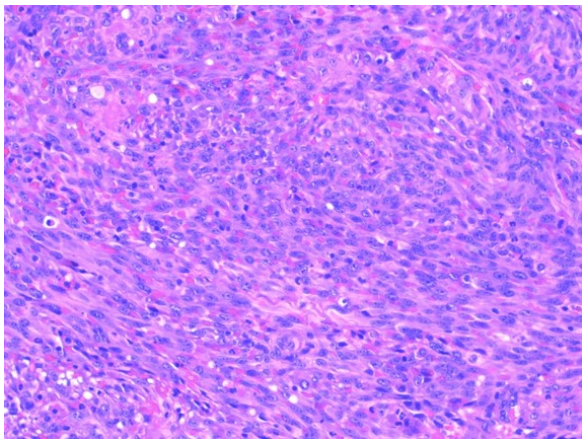

Figure 2: Infiltration of malignant cells with high mitotic activity (40x magnification; H&E).

Angiosarcoma is an aggressive, rare vascular malignancy of endothelial cells and represents 2% of all sarcomas[1] which exhibits high local recurrence rate with early metastatic potential Albeit 50% of angiosarcoma involves skin of head and neck,[2] this is the first case reporting on angiosarcoma involving EAC. Previous aural cases reported involved angiosarcoma of the external ear or pinna[3] and middle ear.[4] Accruing vague presentation, EAC angiosarcoma can be overlooked and diagnosed late. Chronic otorrhea has been deemed as the main risk factor of sarcoma and is the main presentation in our patient. Additionally, presentation of chronic otorrhea with otorrhagia resistant to treatment should alarm attending physicians of a more sinister condition notably malignancy warranting immediate biopsy and imaging.

Gold standard in diagnosis of EAC angiosarcoma is histopathological examination along with immunohistochemistry study. Computed tomography aids in evaluating the site of involvement along with extension of tumour in addition to presence of metastasis. Staging classification for EAC angiosarcoma follows University of Pittsburgh Staging System for Squamous Cell Carcinoma of the Temporal Bone (Appendix 1) as it can guide both treatment as well as prognosis.[3]

Treatment of choice for EAC Angiosarcoma is wide surgical resections with canalplasty. Margin involvement is common in angiosarcoma following its multifocal nature.[1] Additionally, ensuing the high risk of local recurrence, adjuvant radiotherapy is (> Gy 50) is recommended. To date, no solid evidence is present on role of neoadjuvant or adjuvant chemotherapy after surgery or radiotherapy. Cytotoxic chemotherapy has been reported with promising results for various angiosarcomas albeit no standard regime has been reported.

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