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Case Report

Epithelial myoepithelial carcinoma in an ectopic salivary gland: A diagnostic dilemma

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ABSTRACT

The ectopic Salivary Gland is the presence of salivary tissue in unusual locations which do not normally host it. Moreover, neoplasms arising from these ectopic tissues are very rare, however, tumourigenic changes in these inclusions have been observed in a few cases.

We present a clinically unsuspected case of an ectopic salivary gland tissue present on the mid-neck, which was diagnosed as Small lymphocytic leukaemia outside Jawaharlal Nehru Medical College, Aligarh. On further evaluation, a definitive diagnosis of epithelial myoepithelial carcinoma arising in ectopic salivary gland tissue within the cervical lymph node was made. Therefore, these tumours need to be included in the differential diagnosis of neck masses, for a prompt diagnosis and adequate treatment.

1. Case Report

We present a case of a thirty-four-year-old male who presented with a single right mid-cervical nodular swelling (Figure 1). He underwent an excision biopsy and was diagnosed as SLL outside Jawaharlal Nehru Medical College(JNMC), Aligarh Muslim University, Aligarh. One month later he came for an Immunohistochemical evaluation for subtyping of lymphoma. On microscopic evaluation, diffuse infiltration of lymphoid was seen throughout the biopsy (Figure 2). However, on careful examination, a vague lobular architecture with sheets of polygonal cells having clear to eosinophilic cytoplasm and small round nuclei, separated by thin fibrous septa was seen.(Figures 3 and 4). No lymph node architecture was identified. There was no atypia. Mitosis was 1-2/HPF only. No necrosis was seen. Infiltration/invasiveness could not be commented upon as surrounding tissue/well-defined capsule was not seen. Since we received a single block, and the biopsy was highly fragmented, it was very difficult to rule in/out the diagnosis of lymphoma. A differential diagnosis of SLL, ectopic parathyroid adenomas, cellular pleomorphic adenoma, low grade epithelial myoepithelial neoplasm was made based on histopathology. Special staining for Periodic Acid Schiff (PAS) stain was done and came out to be negative. On applying Immunohistochemical Markers, CD45 was negative and showed positivity in only reactive lymphoid cells (Figure 5) Pancytokeratin showed positivity in epithelial cells (Figure 6) S100 and SMA highlighted the myoepithelial cells (Figures 7 and 8). Ki 67 showed 1% positivity (Figure 9). Serum calcium, calcitonin, thyroid function tests were within normal limits.

2. Discussion

Ectopic salivary gland tissue shows a normal salivary gland tissue architecture but has an abnormal location, also known as aberrant accessory, ectopic, heterotopic or salivary gland choristoma.[1] Occurrence of the ectopic salivary gland in the neck is very rare though other locations like temporal bone, lymph nodes, and mandible,

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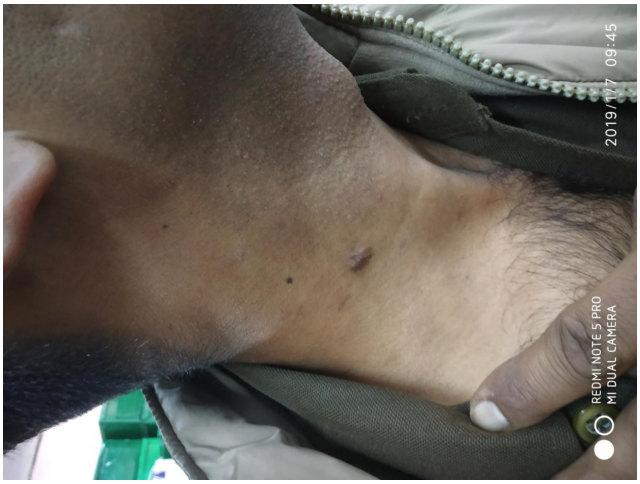

Fig. 1: Clinical picture ofthe lesion after the excision

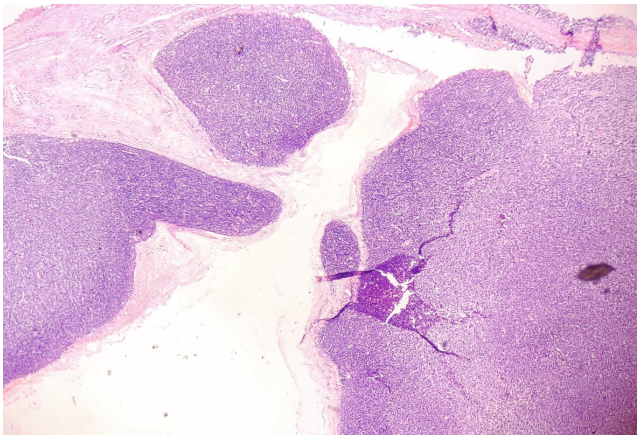

Fig. 2: Lobules and sheets of round to polygonal blue cells separated by fibrous stroma (H and E X4)

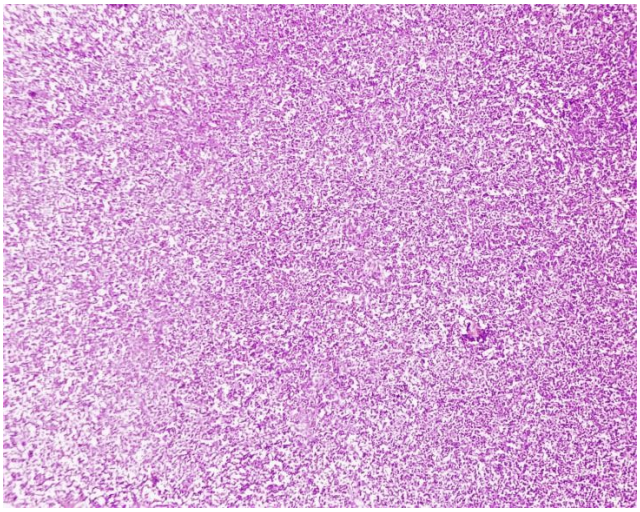

Fig. 3: Clusters of polygonal cells with scant cytoplasm separated by thin fibrous septa (H and E X40)

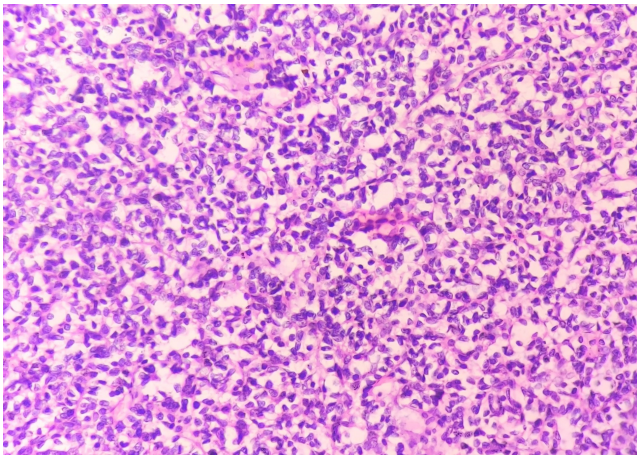

Fig. 4: Clusters of polygonal cells with scant cytoplasm separated by thin fibrous septa (H and E X40)

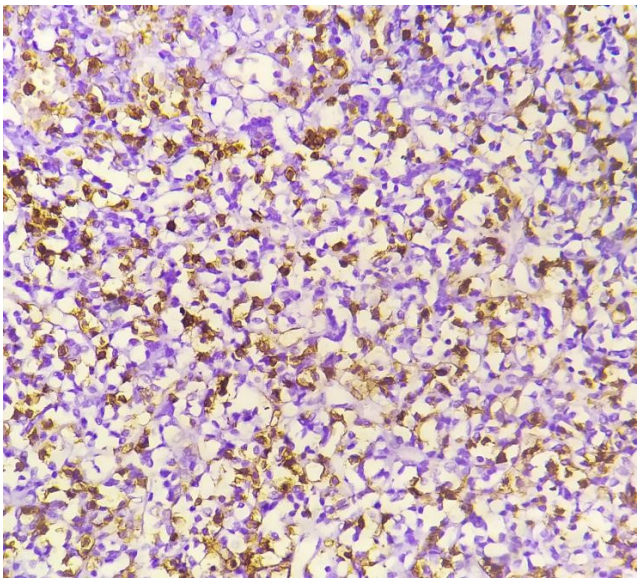

Fig. 5: CD45 positivity seen in the lymphoid cells (X40)

thyroid, and parathyroid tissues in the upper and lower neck have been reported but is still rare.[2] Heterotopias have been described in lymph nodes, external auditory canal, hypophysis, mandible, mastoid bone, middle ear, tongue, sternoclavicular joint, thyroid and parathyroid glands.[2] The embryogenesis of the ectopic salivary gland is not clear and is related to the anatomic site. The location of the ectopic salivary gland in the lower neck, suggests a relationship to the branchial apparatus, particularly the pre cervical sinus.[3] In the sixth week of gestation, growth of the 1st and 2nd branchial arches exceeds and encloses the 3rd and 4th arch in an ectodermal pouch forming a sinus which normally gets obliterated by the end of 2nd month, failure of which leaves cystic which are said to give rise to salivary tissue.[3] Development of neoplasm in an ectopic salivary

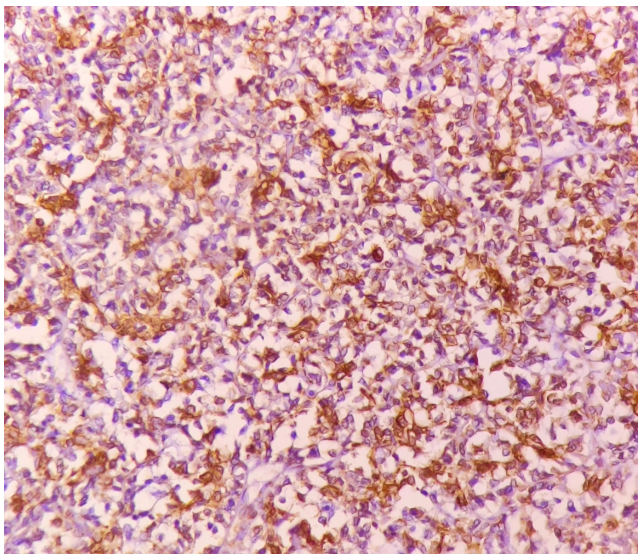

Fig. 6: Pan-CK positivity in epithelial lining (X40)

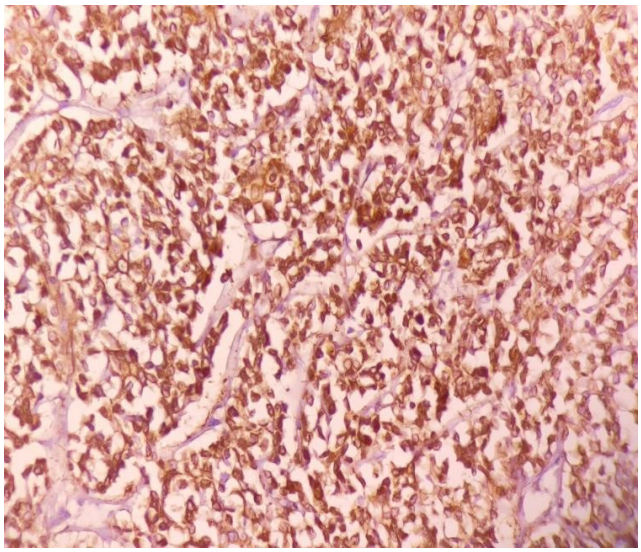

Fig. 7: S100 positivity inmyoepithelial cells (X40)

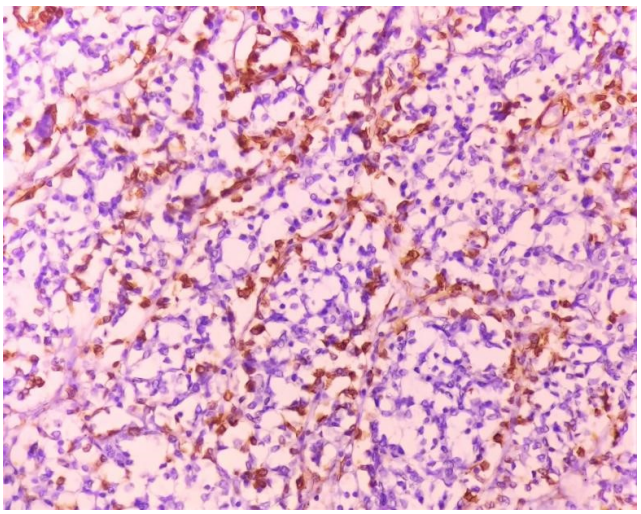

Fig. 8: SMA positivity inmyoepithelial cells (X40)

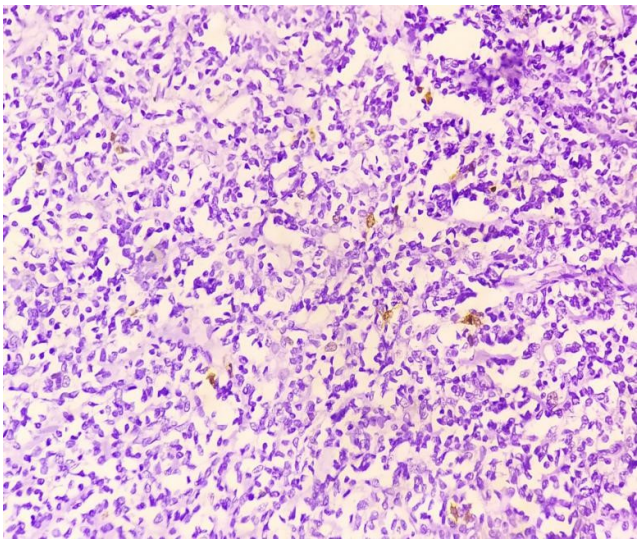

Fig. 9: Ki-67 positivity -1%(X40)

gland is very rare, and when present is usually located in the upper region of the neck with Warthins tumour as the most common benign neoplasm followed by pleomorphic Adenoma arsing in the ectopic salivary gland.[4]

Epithelial-myoepithelial carcinoma (EMC) is a rare glandular epithelial neoplasm of the salivary gland, with an incidence of less than 1 per cent of all the salivary gland tumours.[5] EMC is a rare tumour first which was first described in 1972 by Donath, et al., and till now only around 320 cases have been reported so far worldwide.[6] Parotid gland is the major site in most cases (62%), but can also arise from the nasal cavity, sinuses, palate, submandibular gland, lesser salivary gland, larynx and trachea.[7] This tumour is more common in females with an average incidence in the sixth decade of life. Overall EMC originating from an ectopic salivary gland is very rare with limited literature till now.[8] Diagnosis of EMC is based on histopathology. Microscopic findings include a bilayered arrangement of small luminal cells with eosinophilic cytoplasm and outer myoepithelial cells with clear cytoplasm. These tumours may show a classic, epithelial dominant, Myoepithelial dominant pattern depending on the proportion of epithelial and myoepithelial cells present.[9] Small lymphocytic lymphoma (SLL) can arise in a similar location as this case and such ectopic can be mistaken for the lymph node. On microscopy, diffuse proliferation effacement of mature lymphocytes with a round nucleus and scant cytoplasm can be confused with myoepithelial cells.[10] Pleomorphic adenoma also shows

the variable proportion of epithelial and myoepithelial cells arranged in various pattern In which epithelial (ductal) component forms the inner layer of cysts and tubules while myoepithelial cells from the outer layer are scattered within the myxoid stroma.

Thus, the ectopic appearance of salivary tissue in an unusual location can pose a diagnostic dilemma for both the clinician and pathologist. The nature of the lesion needs to be ascertained, whether malignant or benign and should be promptly treated. Moreover, a thorough investigation should be undertaken to confirm whether it is a primary tumour in the ectopic site or metastasis from a primary tumour of the normal salivary glands.

3. Conclusion

The neck is a common site for multiple swellings with numerous differential diagnoses but, rarely a tumour of ectopic salivary gland origin may be seen in the neck. In the presence of a slow-growing unilateral mass in the upper neck, we should be careful with its diagnosis and treatment, which often present an incidence of unexpected pathology. Early diagnosis offers the possibility of more complete excision with adequate care being taken not to disrupt the tumour to prevent the local and distant spread of neoplastic cells. Long-term follow-up is essential to exclude malignancy, even if the tumour appears to be clinically benign and removed completely.

4. Source of Funding

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5. Conflict of Interest

The authors declare no conflict of interest.

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