Diagnostic Challenges in a Rare Submandibular Epidermoid Cyst Transforming into Squamous Cell Carcinoma - A Case Report

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Abstract

Rationale: Submandibular gland epidermoid cyst is extremely rare and malignant transformation into squamous cell carcinoma (SCC) is even rarer. No such case report is published. **Patient Concerns:** A 49-year-old female reported with a large, slowly growing left-sided neck swelling present for two years. Her primary concern was large swelling. **Diagnosis:** Clinical finding, cytology and imaging were suggestive of 'Epidermoid Cyst of Submandibular Gland'. Intraoperative frozen section and post-operative definitive histopathological examination (HPE) revealed malignant transformation into SCC. **Treatment:** In toto surgical excision was performed. Once the frozen section HPE confirmed transformation into SCC, clearance of 4–6 mm margin and modified supraomohyoid neck dissection was completed. **Outcome:** No sign of recurrence on 1-year postoperative follow-up. **Take-Away Lesson:** This case is unique of its kind without any previous literature report. In case of large and rapidly growing submandibular gland lesion, possibility of malignant transformation should not be overruled.

Keywords: Epidermoid cyst, malignant transformation, squamous cell carcinoma, submandibular gland

Introduction

Epidermoid cysts are a common benign pathological entity encountered usually on the skin. They are derived from ectopically situated ectodermal tissue. It is unusual to appear in major salivary glands and more so in the submandibular gland.[1-3] Malignant transformation of epidermoid cyst into squamous cell carcinoma (SCC) is extremely rare and reported to be between 0.011% and 0.045%.[3,4] No such incidence report in submandibular salivary gland could be traced in existing literature. Diagnosis is a clinical challenge and could be misinterpreted as salivary gland neoplasm, submandibular dermoids, branchial cyst, external laryngocoele etc.[1] The ensuing dilemma may lead to inappropriate and incomplete treatment. Multiple diagnostic investigations are often required to establish an appropriate treatment plan.[5] There is a gap in existing literature as no such case report could be traced on electronic search describing incidence of large epidermoid cyst in submandibular salivary gland transforming into SCC. In this case report, one such rarest case is reported.

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Case Report

A 49-year-old female without any comorbidity reported to our outpatient department with chief complaint of slowly growing large swelling with discomfort in the left side of the neck just below the lower jaw for two years. There was no associated tenderness, history of discharge, difficulty in swallowing or similar lesion anywhere in the body. On examination, it was nontender, soft to firm, diffuse lesion not fixed to surrounding tissue and with normal overlying skin. Clinically, the lesion measured 10 cm × 8 cm × 6 cm in size extending posteriorly to the sternocleidomastoid (SCM) muscle, anteriorly crossing the midline, superiorly overlapping the lower border of mandible

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and inferiorly at the level of cricoids [Figure 1a and b]. Regional lymph nodes could not be palpated. It was fluctuant at the most prominent point of the bulge only and the whole lesion was sagging inferiorly due to gravitational effect. Intraoral findings of dentition, tongue, floor of the mouth, lingual vestibule, salivary flow and salivary duct opening were unyielding. Bimanual palpation and illumination tests were negative.

Preliminary clinical impression was of a submandibular salivary gland neoplasm with multiple differential diagnosis including mucoepidermoid carcinoma, lymphoepithelial lesion, branchial cyst, submandibular dermoid, external laryngocoele, etc. Large-bore needle aspiration was performed revealing a cystic lesion with thick dirty cheesy white content and histopathological findings suggestive of the presence of ductal elements, epidermoid cells and keratin content without any sign of dysplasia or atypical cells. The lesion being abnormally large in size, extensive and relatively rapidly growing, further investigation was carried out. Contrast-enhanced computed tomography (CECT) neck and corroborating magnetic resonance imaging (MRI) findings [Figure 2a-d] were suggestive of a benign submandibular salivary gland cystic lesion with well-capsulated outline and well-maintained outer fatty layer. Based on clinical, imaging and HPE findings, the lesion was diagnosed as epidermoid cyst in submandibular gland.

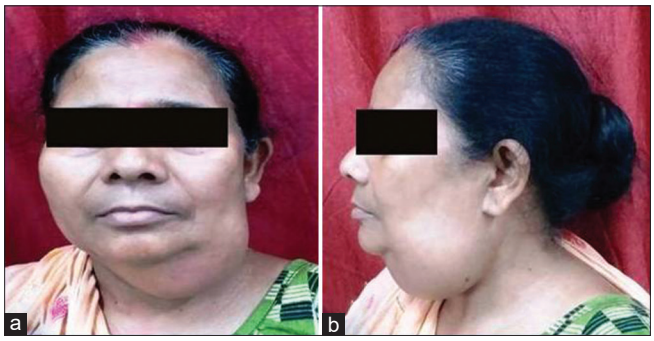

Figure 1: (a and b) Pre-operative photographs of the patient

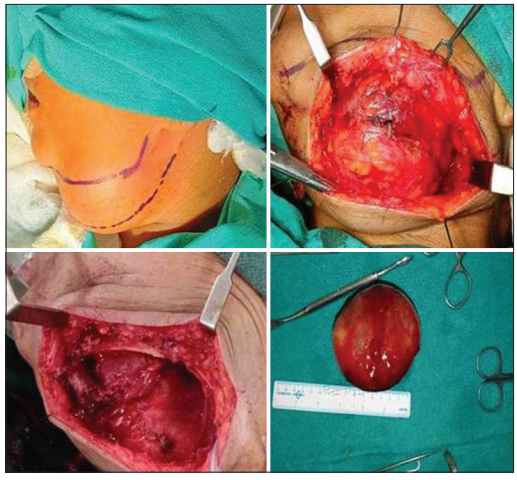

Figure 3: Surgical photographs

Written informed consent of the patient was taken for surgical excision under general anaesthesia, which was carried out using upper cervical crease incision [Figure 3]. Intraoperative frozen section HPE of enucleated specimen along with regional lymph nodes was done. The report revealed malignant transformation with no involvement of regional lymph node. Oncosurgical team was activated and further surgical clearance was completed as per protocol including modified supraomohyoid neck dissection. Postoperative definitive HPE also confirmed the lesion to be epidermoid cyst of submandibular gland with malignant transformation into SCC [Figure 4a and b]. One-year postoperative follow-up was uneventful without any complication or manifestation of recurrence.

Discussion

Epidermoid cysts are simple benign developmental lesions, lined by stratified squamous epithelium with cystic cavity filled with keratinic cheesy material. They usually arise from developmental ectodermal remnants or traumatic implantation of epithelial cells. Approximately 7% of cases are found in the head-and-neck region and 1.5% of cases in oral cavity but are extremely rare in the major salivary glands, especially in submandibular gland.[1,2,6] The exact histopathogenesis is speculative but thought to develop from entrapped branchial pouch epithelial remnants present in major salivary glands.[2]

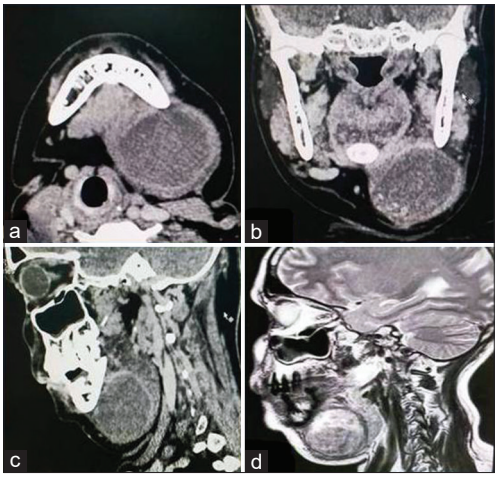

Figure 2: (a-d) Pre-operative CECT and MRI images. CECT = Contrast-enhanced computed tomography; MRI = Magnetic resonance imaging

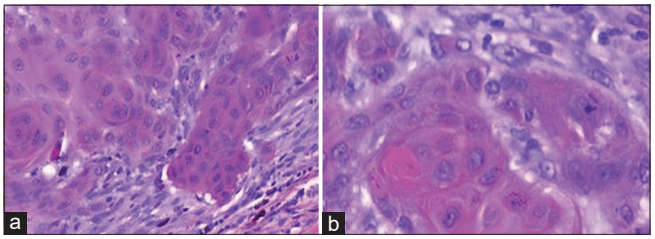

Figure 4: (a and b) Atypical cells with nuclear hyperchromatism and invasion in H and E stain at ×100 and ×400

Malignant transformation of epidermoid cyst into SCC is rare and less than a 100 case reports have been published in the English literature.[4,7,8] Frank *et al*. in their literature review summarised that 54.8% of cases of malignant transformation are seen in the head-and-neck region, mean duration of cyst presence was 92.6 months, mean cystic size at the time of diagnosis was 5 cm and most common presenting symptom is a rapidly growing lesion (48.6%).[3] The aetiology for malignant transformation is uncertain and thought to be long-standing chronic local irritation, infection, inflammation, direct trauma, etc.[3,7] Alkul *et al*. in 2022 pointed out that malignant transformation should be suspected when the lesion shows rapid growth, is larger in size (>2 cm), heterogeneous content, failure to respond to antibiotics, associated pain and erythema, etc.[4] Bhatt *et al*. in 2008 published a case report of SCC arising in the lining of an epidermoid cyst within sublingual gland.[9] No such case report describing malignant transformation in submandibular epidermoid cyst lining could be traced in published English literature till date.

Considering the rarity of the phenomenon, clinical course and ideal line of treatment are not established. The present case showing clinical features of suspicion, multiple advanced diagnostic modalities including CECT, MRI, fine-needle aspiration cytology, HPE and cytology were performed without any result suggesting malignancy. Positron emission tomography scan and immunohistochemistry could have been other viable options if availability and cost-effectiveness factors are not considered. However, intraoperative frozen section HPE was performed and based on the definitive features of malignant transformation, surgical process was redirected accordingly which was pre-empted and necessary written informed consent was taken. Whenever any suspicious clinical manifestation, i.e., rapid growth, large size, etc., is encountered as in the present case, it is strongly recommended to section the specimen and get the HPE done.[3] The potential for malignant transformation of submandibular gland epidermoid cyst should not be ignored.

Once the diagnosis is established, the treatment is essentially surgical excision with 4–6 mm margin with or without clearance of regional lymph nodes.[3-5] Adjuvant radiotherapy or chemotherapy is considered when there is invasion into adjacent tissue, surgical excision is incomplete/doubtful or presence of distant metastasis on subsequent investigations. Frank *et al*. in their literature review, concluded that there was no recurrence on follow-up in 39 cases who underwent wide-local excision (with or without radiotherapy) out of 42 cases and three cases with distant metastasis had negative outcome irrespective of treatment.[3] In this reported case, wide-local excision with modified supraomohyoid neck dissection (including contralateral level IA nodes) was performed. The patient is under follow-up uneventfully for one year without any suspicious manifestations.

Conclusion

Rare incidence of large epidermoid cyst in submandibular salivary gland and further malignant transformation into SCC presents with diagnostic complexity.[10] This case is unique of its kind and occurrence with no previous literature report. Whenever we encounter any such large, relatively fast-growing submandibular gland lesion, irrespective of benign investigatory findings, it is strongly recommended to pre-empt possibility of malignant transformation and go for definitive HPE to get positive patient outcome. It is also important to consult oncosurgical team in advance and to have transparent and effective communication with the patient and/or patient caretakers.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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