CASE REPORT

PLEOMORPHIC ADENOMA OF THE UPPER LIP- A CASE REPORT

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ABSTRACT

Pleomorphic adenoma or benign mixed tumor is the most common salivary gland neoplasm that commonly occurs in major glands, but may also affect the intraoral minor glands. The lips are common mucosal sites, second only to the palate, where they usually present as a freely movable, circumscribed mass. From both diagnostic and therapeutic standpoint, complete surgical removal with adequate margin followed by histopathological evaluation is recommended. The present report describes a case of 25-year-old male with asymptomatic nodular swelling of upper lip which was later diagnosed as pleomorphic adenoma.

KEYWORDS

Pleomorphic adenoma, Upper lip, False capsule, Enucleation

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INTRODUCTION

Salivary Gland tumors though uncommon[1] mostly occurs in major glands with few involving intra oral minor salivary glands[2]. Pleomorphic Adenoma or benign mixed tumor is the most common salivary gland tumor of all salivary neoplasms.[3] Parotid gland followed by submandibular and minor salivary glands are the most common sites.[4] Of all the intraoral Pleomorphic adenomas lips are the most commonly affected sites second only to the palatal region and is observed to be more common on the upper lip.[1]

CASE REPORT

A 25-years-old male came to the Dental OPD, North 24 Parganas District Hospital with a complaint of painless, nodular swelling in the right half of the upper lip. The nodule slowly increased in size during the past four months. There was no history of trauma and his medical and family history was non-contributory. On examination, a 2.5 × 2 cm circumscribed, freely mobile, rubbery mass was elicited in the maxillary right labial mucosa (Fig-1). The overlying mucosa was normal and intact. There were neither neurological abnormalities nor any regional lymphadenopathy. A differential diagnosis of benign tumor of either minor salivary gland or mesenchymal origin was considered. Under local anesthesia via intraoral approach, the tumor was surgically excised with 5 mm clinical margin up to the depth of muscle fascia. This also included the overlying mucosa to ensure complete removal (Fig-2a). Direct primary closure of the resultant wound was done by undermining and advancing the edges (Fig-2b,c). Grossly, a grayish firm tissue piece was received, measuring 2.8 × 2.5 × 1.5 cm (Fig-3a). The cut surface of the lesion was homogeneously white, glistering with occasional minute cystic areas (Fig-3b). On histopathological examination, the sections revealed the presence of a well-circumscribed encapsulated tumor mass being composed of diffuse sheets of epithelial and myoepithelial cells in fibromyxomatous stroma (Fig-4a). The epithelial elements form duct-like structures lined by cuboidal cells oriented around a lumen containing eosinophilic coagulum (Fig-4b). Angular and spindle-shaped myoepithelial cells appeared to spin off the ductal elements (Fig-4c). Squamous cells having typical intercellular bridges and scattered cluster of fat cells were also evident (Fig-4d,e). This confirmed the diagnosis of pleomorphic adenoma. One year post-surgical follow up revealed no locoregional recurrence.

Fig-1: Freely movable, circumscribed, rubbery nodule involving maxillary right labial mucosa.

Fig-2: (a) Surgical excision of the submucosal mass up to the depth of muscle fascia. This included the overlying mucosa; (b, c) Direct primary closure of the wound by undermining and advancing the edges.

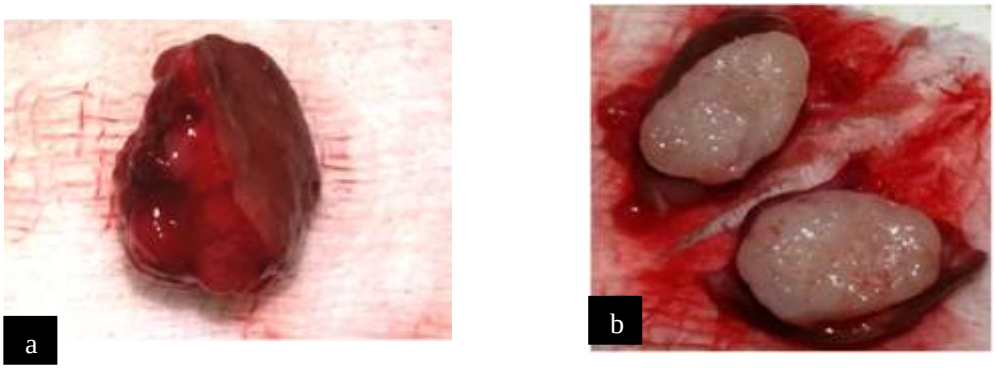

Fig-3: (a) The tumor mass after excision; (b) Gross specimen showing homogeneously white, glistering cut surface with minute cystic areas.

DISCUSSION

Salivary gland tumors are uncommon, having an annual incidence of 1.0 to 6.5 cases per 100,000 people worldwide.[1] Most of them (70%) occur in major glands; only few (30%) involve intraoral minor salivary glands.[2] Regardless of the site of origin, pleomorphic adenoma (PA) or benign mixed tumor is the most common salivary gland tumor that accounts for about 60% of all salivary neoplasms.[3] About 80% of pleomorphic adenomas arise in the parotid, 10% in the submandibular gland and 10% in the minor salivary glands of the oral cavity. When they arise in the oral mucosa, the site of predilection is the mucosa over the posterior hard palate and anterior soft palate; otherwise, PA can occur in any location where minor salivary glands exist (e.g. lip, cheek, retromolar region, floor of the mouth, and alveolar mucosa).[4] The lips are commonly affected sites, second only to the palate, and accounting for about 20-40 % of all intraoral PA. Labial tumors are significantly more common in the upper lip, which accounts for about 90% of all lip tumors.[1] PA mainly affects women in their fourth to sixth decade of life.[5] In the lips, however, they tend to occur at an earlier age than it does at other sites.[6] Although PA has a natural history of asymptomatic slow growth over a long period, some cases exhibiting rapid growth have been reported, especially in the palate. Small tumors typically form smooth, mobile, firm lumps but larger tumors tend to become bossellated and may attenuate the overlying skin or mucosa.[7]

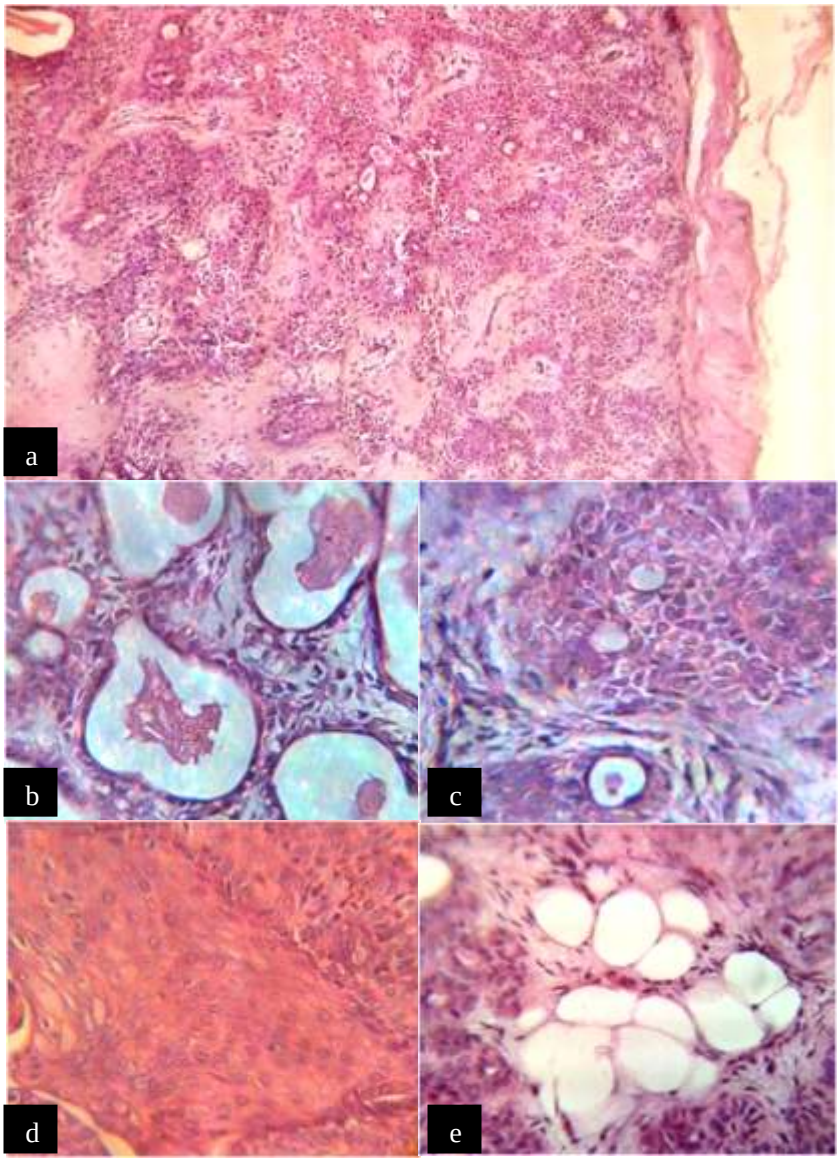

Fig-4: (a) Low-power photomicrograph revealing an encapsulated tumor mass composed of diffuse sheets of epithelial and myoepithelial cells in fibromyxomatous stroma (H&E, x4); (b) Tubular structures lined by cuboidal cells oriented around a lumen containing eosinophilic coagulum (H&E, x40); (c) Angual and spindle-shaped myoepithelial cells spinning off the ductal elements (H&E, x40); (d) Squamous cells having typical intercellular bridges (H&E, x10); (e) Cluster of fat cells (H&E, x40).

Ulceration of the nodular mass may occur, but the presence of ulcer provides no clue to the invasiveness of the tumor.

Differential diagnosis of such clinically benign mass of the upper lip can be classified into three groups-salivary gland tumors, mesenchymal tumors, and infections (Table-1).[8] Among benign salivary gland neoplasms of upper lip, only the occurrence of canalicular adenoma exceeds that of PA. Canalicular adenoma usually presents as a slow growing, painless, firm to fluctuant, pink or bluish mass with an inherent tendency of multifocality. Acinic cell adenocarcinoma is the most common malignant salivary gland neoplasm of the upper lip whereas mucoepidermoid carcinoma is more common in lower lip.[9] Mucocele and malignant neoplasms commonly affect lower lip and their incidence in upper lip is rare. In younger age, a blue swelling of upper lip that blanches under digital pressure may be suspected as hemangiomas or lymphangiomas. Few cases of dermoid and epidermoid cyst and a rare occurrence of oral cysticercosis of upper lip have also been reported.[10-12]

Grossly, PAs are usually well-defined, ovoid or round masses having smooth, sometimes bosselated surface. Their consistency varies from hard to rubbery or soft and fluctuant. The cut surface is typically white and resembles a cut potato. Bluish

TABLE

Salivary gland tumors	Pleomorphic adenoma Canalicular adenoma Mucoepidermoid carcinomas
Mesenchymal tumors	Lipoma Leiomyoma Nerve tumors (neurofibroma, neurilemmoma/schwannoma) Benign fibrous histiocytoma Oral focal mucinosis Granular cell tumor
Infections	Tuberculosis Syphilitic gumma (if the lesion presents as an ulcerated mass) Deep fungal infections (e.g. histoplasmosis, cryptococcosis, blastomycosis, coccidioidomycosis)

Table 1: Differential diagnosis of clinically benign solitary mass of the upper lip.[8]

gritty areas representing cartilage-like material and gelatinous or glistening component representing myxomatous differentiation may be seen. Older and larger tumors often show cystic regions.[13]

Microscopically, the most significant histologic features relate to the capsule of the tumor. Most tumors are encapsulated, but there is wide variation in both the thickness and presence of the capsule. In predominantly mucoid tumors and those that arise in minor glands, the capsule may be absent or poorly formed. Many tumors show local extension of finger-like tumor processes into or beyond the capsule. PA is characterized by diverse architectural rather than cellular pleomorphism. Epithelial and modified myoepithelial elements intermingle most commonly with tissue of mucoid, myxoid or chondroid appearances. There is often proliferation of epithelial cells in strands or sheets about tubular or ductlike structures. These tubular elements are composed of cuboidal cells and may contain eosinophilic coagulum.[14] Spindle-shaped myoepithelial cells may appear to spin off the ductal elements.[2] In other areas the tumor cells may assume a stellate, polyhedral, squamous, plasmacytoid, or clear form. In some tumors the stroma becomes densely hyalinized; in others, chondroid, adipose, and even osseous stromal elements are encountered.

Although the mobility, lack of pain and ulceration may raise the suspicion of benignity, biopsy is recommended for definitive diagnosis. For intraoral tumors, the type of biopsy usually depends upon the size and location of the tumor. In palate, a deep incisional biopsy of the mass is recommended in its center to establish a firm diagnosis prior to planning definitive surgery. In other mucosal sites, diagnostic biopsy and curative surgery are one and the same and a peripheral excision with adequate free margin is all that need. For lip tumors, this must include overlying mucosa but should not include overlying skin as the muscle fascia of orbicularis oris is an effective anatomical barrier. In any site, enucleation, or a "shelling out" of a PA, is contraindicated.[13] As these tumors are known to have microscopic pseudopod-like extension into the surrounding tissue due to 'dehiscence' in the false capsule, a "conservative" enucleation would almost ensure residual tumor cells and set the patient for multifocal recurrences. Most patients with recurrence will have it within 18 months. Long-term follow-up is recommended, as the risk of recurrence may remain lifelong for such patients.[15]

CONCLUSION

Pleomorphic adenoma must always be in the differential list of a well-defined, painless, mobile mass of upper lip. Complete surgical removal followed by histopathological confirmation is the mainstay of management. Long-term follow-up is recommended as PA may notoriously be recurrent particularly while arising in intraoral minor salivary glands.

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