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Abstract Pleomorphic adenoma is a benign tumor commonly involving the salivary glands and mainly the parotid gland. It affects the minor salivary glands with the palate being the most common site of occurrence. The involvement of the upper lip is considered rare with very few reports in the literature. Clinically it is a painless, non-erythematous, firm and slowly growing tumor. Microscopically it has a biphasic microscopic appearance with mixed epithelial and stromal components. Although it is benign the rate of malignant transformation is reported as being up to 4 % in long standing cases. The incidence of this tumor has been reported in various gender, ethnic and geographic groups. With the recent reports of the increase in the variations of location and the biological nature of the tumor the need for extensive research is more demanding. This case report presents a systematic approach to long standing pleomorphic adenoma affecting the upper lip.

Keywords Case report · Biopsy · Head and neck neoplasms · Salivary gland tumor · Resection

Introduction

Salivary gland tumors account for approximately 3 % of all head and neck tumors [1]. The most common salivary gland tumor is pleomorphic adenoma (PA) that accounts for 60–65 % of such diseases and mainly affects the major salivary glands [2]. Approximately 85 % of all PAs are located in the parotid glands, 10 % in the minor salivary glands and 5 % in the submandibular glands [1]. The PA most commonly affects the minor salivary glands in the palate, followed by the lips, buccal mucosa, floor of the mouth, tongue, tonsils, pharynx, retromolar area and nasal cavity. The second most common site of origin is the upper lip and is characteristically seen in women in the fourth to sixth decades of life with a history of asymptomatic slow growth over a long period. It is rarely seen in children and adolescents. There is a 2–4 % malignant transformation rate in tumors that are long standing, mostly occurring in the parotid gland [3]. The PA was first described by Minssen in Ahlbom's monograph in 1874 but the etiology of PA has remained unknown. Cytogenetic and molecular studies have reported it as having an epithelial origin with chromosomal abnormalities at 8q12 and 12q25 [4] and uncommon mutations of TP53 [5].

Classically PA is biphasic and is characterized by an admixture of polygonal epithelial and spindle-shaped myoepithelial elements in a variable background stroma that may be mucoid, myxoid, cartilaginous or hyaline. The epithelial elements can be arranged in duct-like structures, sheets, clumps and/or interlacing strands and consist of polygonal, spindle or stellate-shaped cells (hence pleomorphism). Some areas of squamous metaplasia and epithelial pearls may be present. The tumor is not enveloped but is surrounded by a fibrous capsule of varying thickness [5]. The tumor extends through normal glandular parenchyma in the form of finger-like pseudopodia but this is not a sign of malignant transformation.

Case report

A 30-year-old male Nigerian patient presented to the department of oral and maxillofacial surgery, faculty of dentistry, Tripoli University, Libya, with a history of

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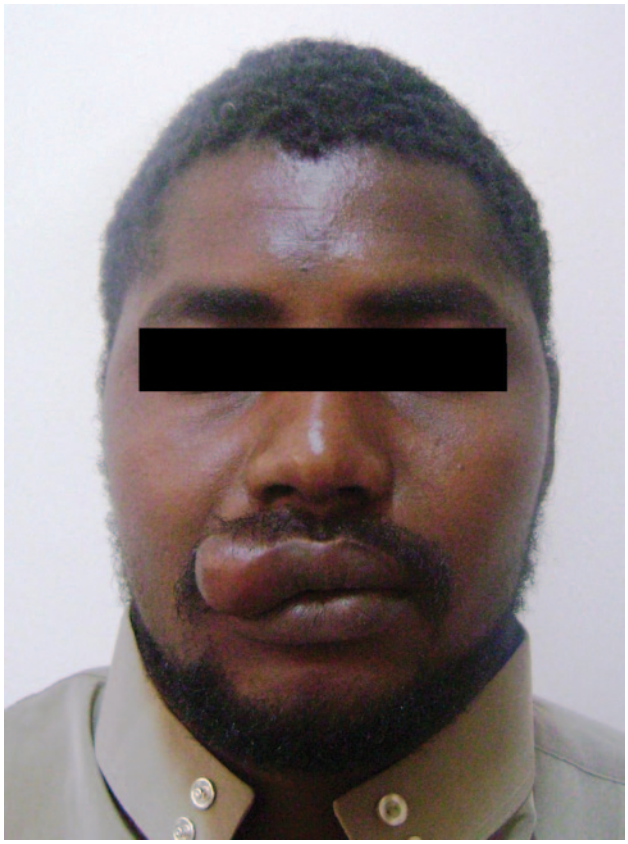

Fig. 1 Preoperative frontal view of the tumor mass in the right side of the upper lip

swelling in the upper lip on the right side for the past 28 months. The presenting size of the swelling was approximately 4×3 cm which had gradually increased from being pea-sized to its present form (Fig. 1). There was no fever or pain during the onset and increase in size of the swelling. On palpation the swelling was firm, non-tender, and predominantly attached to the skin. The overlying skin was stretched and glossy in appearance with a normal temperature. The vermillion-colored border of the affected side was almost obliterated but the physiological movements of the lip were unaffected.

The intraoral examination included a clinical examination along with panoramic radiography to rule out any associated dental and osseous pathologies. A complete physical examination was carried out to identify any associated pathology. Based on the history and clinical examination, the swelling was solitary without any associated pathology. A provisional diagnosis of fibroma, neurofibroma or PA was made. A fine needle aspiration biopsy (FNAB) was performed to determine the possibility of malignancy due to the long standing course. The results of the FNAB revealed a benign soft tissue neoplasm affecting the upper lip. An incisional biopsy was planned and executed to confirm the diagnosis. The mass was approached from the mucosal surface to avoid any scars on the lip. The histopathological evaluation revealed the biopsy specimen as a PA affecting the upper lip.

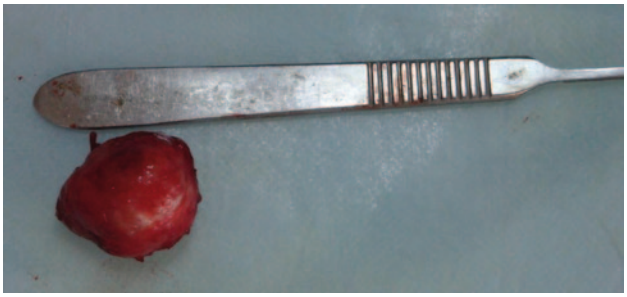

Fig. 2 Surgical specimen of the encapsulated tumor mass after complete excision

As a definitive treatment an excision biopsy was planned using local anesthesia and consent was given for excision of the tumor mass. A vertical incision was placed on the mucosal surface and blunt dissection was performed until the mass was approached. The tumor was well encapsulated and easily separated from the surrounding tissues. The superior aspect of the tumor was almost attached to the skin with surrounding visible hair follicles. A careful separation of the mass was carried out until it was free from the skin and it was removed in a single piece (Fig. 2). The surrounding normal tissues were excised to ensure wide safety margins. Hemostasis was achieved and the tumor bed was irrigated with ample amounts of normal saline and povidine iodine. The thinned out muscle layer was re-approximated and sutured using resorbable polyglactin 910 followed by closure of mucosal layers and the incision. The lip movements were checked and found to be normal but the architecture of the lip was unesthetic due to the long standing presence of the mass leading to thinning of the muscle.

The histopathology report of the surgical specimen confirmed the initial diagnosis with the following report: proliferation of the epithelial and myoepithelial cells in a myxoid background. The epithelial cells were arranged in ducts and small islands, while myoepithelial cells appeared to be spindle-shaped. The supporting stroma appeared myxoid and occasionally myxochondroid in some areas (Fig. 3).

The patient was followed-up regularly for almost 14 months. There were no signs of recurrence and the movements of the lip were unaffected but the architecture of the lip was esthetically unacceptable (Fig. 4). Corrective esthetic surgery to restore the lip architecture was planned after completion of 18 months follow-up but the patient did not respond to the follow-up after 14 months.

Discussion

The PA is the most common tumor of the salivary glands with a high predilection of 85 % in the parotid glands followed by minor salivary and submandibular glands [6]. Although the etiology of this tumor is unknown a few reports on the basis of cytogenetic and molecular stud-

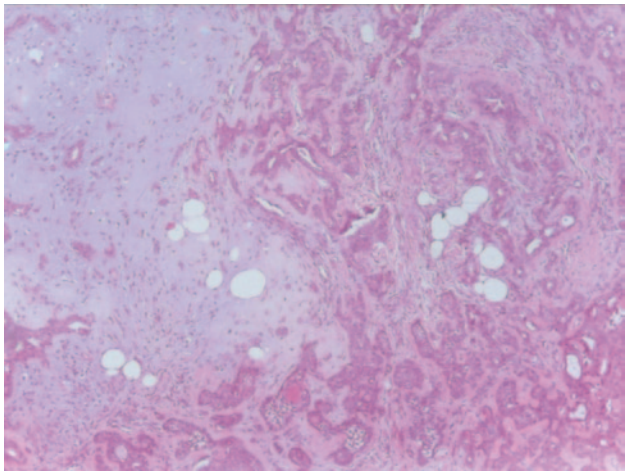

Fig. 3 Photomicrograph showing several ducts surrounded by myoepithelial cells and a myxoid stroma (H & E stain, ×10)

Fig. 4 Postoperative view of the upper lip with severe loss of architecture causing unesthetic appearance

ies suggested chromosomal abnormalities and uncommon mutations of TP53 [5]. The PA is most commonly seen in women in the fourth to sixth decades of life and is exceptionally rare in childhood [7]. The incidence and distribution of PA have been reported to vary with race and geographic region. The most common location for the development of PA in the palate can be attributed to the presence of abundant minor salivary glands in this region. The frequent occurrence of PAs in minor salivary glands is then followed by the upper lip, cheek, tongue, floor of the mouth and rarely the lower lip [8]. The PA is usually a painless, non-tender, non-erythematous, well demarcated and slowly growing tumor with a long course of history. The increase in size severely compromises the esthetics and function of affected areas [9].

The differential diagnostics of these lesions most commonly include lipoma, fibroma, neurofibroma, myoepithelioma and basal cell carcinoma. These lesions are more commonly seen in the oral soft tissues. The long standing history suggests the inclusion of malignant tumors such as rhabdomyosarcoma or carcinoma expleomorphic adenoma [3]. Osseous changes have been reported in the minor salivary glands [8, 10].

The management of PA consists of resection with wide safety margins and FNAB is a very useful and quick diagnostic aid that can be performed in a small clinical setting. The FNAB examines the cytological appearance of the tumor to distinguish whether the lesion is benign or malignant. Clinicians should be aware of the limitations of FNAB as it has varying rates of sensitivity and specificity. As a process of determining the definite diagnosis, an incisional biopsy should be planned. The incisional biopsy is the gold standard technique to confirm the clinical diagnosis in such lesions. It cannot be performed in the major salivary glands due to the associated potential complications. Hence it is best to perform FNAB as the initial diagnostic measure. A resection along with wide safety margins is the best treatment modality [11]. The incision should be made vertically on the labial mucosa. Deep horizontal incisions on the labial mucosa should be avoided to prevent severe fibrosis mucosa leading to shortening of the lip. Blunt dissection should be performed for careful separation of the surrounding tissues. Any severance from the muscle will result in an abnormal and unesthetic function of the lip and the affected area. The tumor is easily identified by the thick capsule covering the mass which usually confirms the benign nature of the tumor. However, this is not a confirmed feature unless proven by the histopathological examination. Care should be taken to prevent damage or rupture of the capsule as a recurrence of 45 % and malignant transformation of 4 % have been reported in cases of PA. This is mainly attributed to rupture of the capsule and inadequate clearance of the tumor cells from the surrounding tissues [12]. The complete excision of the tumor along with surrounding normal tissues should be carried out to prevent recurrence. The use of frozen sections to determine the safety margins is an excellent aid to prevent recurrence and malignant transformation [3, 9, 11].

A typical PA has a biphasic microscopic appearance with epithelial and stromal components. Ductal epithelial glands make up glandular structures and cystic structures of various sizes. Myoepithelial cells can be of very different cytomorphological appearances, such as spindle-shaped, plasmacytoid, squamoid, star-shaped and basaloid. These cells can be distributed separately or may form islands, solid sheets or trabeculae. Myxochondroid changes are the most common change in the stroma; however, the stroma can be loose myxoid or dense hyalinized depending on the rate of collagenization [6]. The

cartilaginous differentiation areas commonly seen in tumors originating from the parotid and submaxillary glands are seen much less frequently in tumors originating from the minor salivary glands. The ductal epithelial component is positive for keratin and epithelial membrane antigen (EMA) by immunohistochemical investigations [12].

In conclusion, the PA should be treated aggressively to avoid increasing the rate of recurrence. The occurrence of this tumor in rare sites, such as the base of the tongue, external auditory canal, rhinopharynx, upper lip and lower lip suggests the widespread involvement in the oral tissues apart from its common locations [1, 8, 13]. The uncertain biological behavior of the tumor with the inclusion of osseous elements [10] and increase in the rate of malignant transformation [3, 11] should alert clinician to an intensive management protocol. Performing an incisional biopsy of the tumor enables planning of optimal surgical management but cannot be performed in the major salivary glands due to its limitations and the associated complications. The FNAB is the best diagnostic aid in major salivary glands although it has limitations. More research is needed to identify the etiology and study the biological behavior due to the reported variants and changing clinical patterns. The optimal management includes aggressive surgical treatment and the long-term follow-up to prevent recurrence. A long-term follow-up enables identification of recurrence along with esthetic changes at the tumor site [13].

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None.

Ethical approval
All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5).

Informed consent
Informed consent was obtained from the patient for the surgical management, follow-up and the exhibition of the eyes in case of publication or display for scientific purpose.

Conflict of interest
Irfan Mohammed, Abir Saghali, Saravanan Ramasamy and Abdurahman Elmezwghi declare that they have no conflict of interest.

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