A Discrete Swelling of the Upper Lip: A Diagnostic and Clinical Stepladder

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Abstract An upper lip swelling can be attributed to multiple etiologies. The clinical presentation and differential diagnosis of an upper lip swelling is described. Histologic examination revealed the presence of a Schwannoma. This rare entity in the upper lip must be considered by the clinician during the evaluation of an upper lip swelling.

Keywords Lip swellings · Schwannoma · Neurilemmoma

Case Presentation

A 28 year old woman reported to the Department of Dentistry with a swelling on the right side of the upper lip of several months duration. The lesion measured 1 × 1 cm with normal overlying mucosa (Fig. 1). On palpation, the

lesion was moderately firm in consistency, fairly well circumscribed and not fixed to the adjacent tissue. It did not blanch on pressure and no pulsation could be felt. No tenderness and altered sensation was elicited. Her dental and medical history was unremarkable and she could not remember any history of trauma to the area. Intraoral examination did not show any dental pathology.

Differential Diagnosis

On the basis of the history and clinical presentation of a slow growing, single, non ulcerated, well circumscribed, freely movable, asymptomatic mass located within the submucosa of the upper lip, it was felt that the lesion was more likely to be benign in nature. A lesion of a minor salivary gland origin, infectious origin (odontogenic/nonodontogenic) and mesenchymal tumour of vascular, smooth muscle or neural origin were among the conditions being considered.

The abundance of minor salivary gland tissue and the propensity for an accidental injury make mucocles or salivary duct cysts among the more commonly seen swellings involving the lips. They typically present as bluish, firm nodular swellings. Often, scarring of the overlying mucosa is observed due to inadvertent repeated rupture of the cysts. A partial obstruction of the salivary duct by a sialolith may lead to a localised soft tissue swelling. Also, it has been observed that mucocles are restricted almost entirely to the lower lip and seldom found on the upper lip. The possibility of the swelling being a mucocle was negated by the presence of a normal overlying mucosa and the absence of a history of trauma to the lip.

Minor salivary gland tumours represent around 10–15 % of all salivary gland neoplasms and are commonly seen in the palate and lips [1]. Accessory salivary gland neoplasms of

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Fig. 1 Clinical photo showing a 1 × 1 cm well circumscribed soft tissue swelling with normal overlying mucosa

the lips are almost universally found on the upper lip and only rarely on the lower lip. This implies that trauma may play no role in the development of salivary gland tumors in this location. Approximately 75 % of salivary gland tumors in the upper lip are benign, among which, the pleomorphic and canalicular adenomas are the most common. Pleomorphic adenomas are relatively common in young adults and the lip is the most common site for the canalicular adenoma (90 % of all cases) [2]. The clinical impression in this case was that this most likely represented a benign minor salivary gland tumor. The prospect of a malignant salivary gland tumor in the upper lip cannot be ruled out definitively, albeit, the lack of fixation and the presence of a well defined border in our patient made this diagnosis highly improbable.

Hemangiomas usually manifest at infancy and childhood, exhibit a rapid proliferative phase, and slowly involute to nonexistent. Superficial lesions are usually deep red or bluish red and seldom well circumscribed. They are readily compressible and fill slowly when released. The short duration of the lesion, non compressibility, absence of blanching and the patient's age make this diagnosis highly unlikely.

Calibre-persistent labial artery, described as a pulsatile elevation in the upper lip is more common in elderly population and is often associated with other vascular lesions such as hemangiomas, arteriovenous malformations and varices [3]. It is often misdiagnosed as a salivary duct cyst and reports of profuse bleeding have been recorded during surgical excision.

Lymphangioma, a benign hamartomatous hyperplasia of the lymphatic vessels most commonly occurs on the tongue, but is seen also on the palate, buccal mucosa, gingiva and lips. The lip involvement is in the form of a nodular, asymmetric, painless swelling. Rarely, it may also present as a diffuse and symmetrical enlargement of the lips.

A diagnosis of a soft tissue infection of the lip (odontogenic/nonodontogenic) is not justifiable owing to the duration of the lesion, total absence of symptoms and a normal dentition. An abscess causing a swelling of the lip is characteristically painful, erythematous, fluctuant and poorly circumscribed.

Leiomyoma, a benign tumour derived from the smooth muscle is uncommon in the oral cavity probably because of the general absence of smooth muscle there except in blood vessel walls and, occasionally, in the circumvallate papillae of the tongue [4]. Most cases are seen in the 3rd decade of life and are described as a slow growing painless lesion. With the lips being one of the more common sites, leiomyoma was considered as a possible diagnosis.

Also considered were the reactive and neoplastic neural lesions (traumatic neuroma, neurofibroma, neurilemmoma). Traumatic or amputation neuroma is not a true neoplasm, but rather an exuberant attempt at repair of a damaged nerve trunk. The oral traumatic neuroma appears as a small nodule near the mental foramen or on the lips or tongue and digital pressure usually causes considerable pain locally [5]. The patient had no history of trauma and the lesion was completely painless.

Neurilemmomas can appear as discrete, non ulcerated nodules and tend to be of the same colour as the normal mucosa [6]. In spite of the fact that they are most common in the head and neck region, the involvement of the upper lip is considered to be extremely rare. Neurofibroma is another common peripheral nerve sheath tumour, consisting of mixture of Schwann cells, perineurial cells and endoneurial fibroblasts. It is seen either as a solitary lesion or as part of the generalised syndrome of neurofibromatosis. Other than the absence of systemic and hereditary factors in the solitary form, no difference exists between the two variants [6].

Conditions such as Orofacial Granulomatosis (Merker-Rosenthal Syndrome, Cheilitis Granulomatosa), Crohn's disease, sarcoidosis, mycobacterial infections (Tuberculosis, leprosy) can present, among other features, as enlargement of the orofacial tissue including the lips [7, 8]. These lip swellings are often diffuse in nature and have a long history of regression and recurrence. Involvement of both lips in association with other systems, such as gastrointestinal tract, is more common. Other conditions such as angioedema, contact allergy, cheilitis glandularis are often associated with causative agents or hereditary factors and may present as enlargement of the labial salivary glands that leads to a nodular texture of the lip and inflamed, erythematous duct orifices on the labial mucosa [9]. In this case, the well circumscribed nature of the lesion, its short duration, and no known exposure to a causative agent does not support the diagnosis of a granulomatous/non granulomatous swelling of the upper lip.

Diagnosis and Management

A biopsy was performed under local anesthesia (2 % lignocaine with 1:100,000 adrenaline). A clean cleavage plane around the lesion could be established with ease and

the lesion was completely encapsulated with a thick capsule (Figs. 2, 3). No vascular or neural bundle was found adherent to the mass. The surgical specimen was sent to the Dept of Pathology for histologic staining and examination. Sections taken from the tumour showed an apparently well circumscribed lesion composed of spindled cells. The tumour showed almost alternate areas of loose oedematous tissue with sparse spindled cells (Antoni B areas) and a more cellular area (Antoni A) composed of many spindle cells in a palisaded arrangement (Fig. 4). At places these cells were palisaded around acellular areas called Verocay bodies (Fig. 5). The cells were monotonous in appearance and showed no atypia. No necrosis or mitosis was noted. On immunostaining, the cells were strongly positive for S100 protein (Fig. 5 inset) and neuron specific enolase. A histopathologic diagnosis of Schwannoma was made.

Discussion

Schwannomas or neurilemmomas are benign tumors of nerve sheath origin. Schwann cells form a thin barrier around each extracranial nerve fibre and wrap larger fibres with an insulating membrane, the myelin sheath, to enhance

Fig. 2 Intraoperative view showing an encapsulated swelling with clean cleavage plane

Fig. 3 Excised lesion

Fig. 4 Low power view of the swelling showing a well circumscribed spindle cell tumor with characteristic alternate cellular areas (Antoni A) and paucicellular areas (Antoni B). (H and E × 200)

Fig. 5 Schwannoma showing palisaded arrangement of spindle cells forming Verocay bodies. (H and E × 400) Inset shows the tumor cells to be positive for S100. (IHC S100 × 400)

nerve conductance. Any nerve covered with a Schwann cell sheath can be a point of origin for this tumour. This tumour was first described by Verocay in 1908 and was named as “neurinoma”. Subsequently, other names like, solitary nerve sheath tumour, perineural fibroblast tumour, neurilemmoma and schwannoma have been proposed by other workers, of which, the latter two are the most popular. The head and neck region, with its complex innervation is associated with nearly 45 % of all cases [10]. Large nerves such as the vagus and the cervical sympathetic chain are commonly involved. The oral cavity and the lips are highly innervated, yet, intraoral occurrences are quite uncommon and account for barely 1 % of these tumors [11]. Das Gupta and associates [12] reported only 17 cases of intraoral involvement in a total of 136 schwannomas of the head and

neck. Hatziotis and Asprides in a review of intraoral neurilemmomas, found only 7 in lip and 6 in buccal mucosa [13]. The intraoral lesions show a predilection for the tongue, palate and buccal mucosa. Involvement of the upper lips is extremely rare and only 8 cases have been reported [14]. A solitary, slowly enlarging, freely mobile, smooth surfaced mass is almost always its signature presentation. Most oral schwannomas are asymptomatic and quite a few are discovered during routine clinical examination. When the nerve of origin is small, its association with a given tumor may not be easily obvious. In this case, the cutaneous branch of the infraorbital nerve distributed throughout the upper lip is the most likely origin of the schwannoma. In a few patients with large lesions, interference with speech or swallowing has been reported [15].

Aids like ultrasound (USG), computed tomography (CT) and magnetic resonance imaging (MRI) have been used to diagnose schwannomas in soft tissues. MRI is usually the imaging modality of choice and a schwannoma appears hypointense or isointense relative to muscle on T1—weighted images [16]. Intraoral schwannomas in soft tissue usually appear as a smooth submucosal swelling mimicking other lesions like mucoceles, benign salivary gland neoplasms, vascular and smooth muscle tumours. A clinical diagnosis is rarely made and as this is an infrequent tumour it is not usually suspected in the oral cavity. Kun and coworkers were able to make a corrective preoperative diagnosis in the neck in only 4 of a total of 49 cases and concluded that it was very difficult to make the diagnosis based on diagnostic imaging techniques [17]. Also, upper lip schwannomas present as relatively small lesions and the use of imaging modalities should not be considered as routine or necessary [16].

Intense S-100 protein immunoreactivity for cytoplasmic and nuclear patterns indicates neural origin [18]. These staining results and the associated H&E microscopic findings confirmed the diagnosis of schwannoma in the reported case. As neurofibromas show a much greater propensity for malignant transformation, a histological differentiation between schwannoma and neurofibroma becomes essential because an apparently “solitary” neurofibroma may be a manifestation of neurofibromatosis. There are 5 histological variants of schwannoma and a mixture of Antoni A along with Verocay bodies and Antoni B patterns is pathognomic of this lesion [19]. Hyperchromatism and cellular atypia seen in ancient schwannoma variant can be misdiagnosed as soft tissue sarcoma [20]. Even as the malignant transformation of a schwannoma has been reported in a few isolated cases, it is considered to be extremely rare in the oral cavity [21, 22].

Surgical excision of the lesion is the treatment of choice. The encapsulated form is enucleated easily, whereas the nonencapsulated requires normal tissue margins to avoid

relapse. Recurrence is rare and is usually attributed to a multinodular variant and incomplete surgical resection [23].

Follow-Up

The presence of schwannoma calls for a careful search for nerve tumors in other parts of the body. At a subsequent follow up, the patient denied a family history of neurofibromatosis. Examination by a physician did not reveal any such lesions in other parts of the body. No recurrence has been observed in the upper lip since 8 months.

Conclusion

In summary, we present a case of a patient with a mass of the upper lip resembling an adenoma, which on biopsy proved to be a schwannoma. Although the involvement of the upper lip is very rare, this entity must be considered during the evaluation of a lip swelling.

Conflict of interest None.

References

1. Speight PM, Barrett AW (2002) Salivary gland tumours. *Oral Dis* 8(5):229–240
2. D'Silva NJ, Benian R, Flint A, Cordell KG (2008) Soft tissue swelling of the upper lip. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 105:271–273
3. Rosdy NM, Firth NA, Rich AM (2010) Caliber-persistent labial artery: often misdiagnosed as a mucocele. *Int J Oral Maxillofac Surg* 39:1230–1233
4. Orsini G, Fioroni M, Rubicini C, Piattelli A (2001) Leiomyoma of the lip: report of a case. *J Oral Maxillofac Surg* 59:80–83
5. Subhashraj K, Balanand S, Pajiniammalle S (2009) Ancient schwannoma arising from the mental nerve. A case report and review. *Med Oral Patol Oral Cir Buccal* 14:E12–E14
6. Chrysomali E, Papanicolaou SI, Dekker NP, Regezi JA (1997) Benign neural tumors of the oral cavity. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 84:381–390
7. Fatahzadeh M, Rinaggio J (2006) Diagnosis of systemic sarcoidosis prompted by orofacial manifestations; a review of literature. *J Am Dent Assoc* 137:54–60
8. Sciubba JJ, Said-Al-Naief N (2003) Orofacial granulomatosis: presentation, pathology and management of 13 cases. *J Oral Pathol Med* 32:576–585
9. Jham BC, Meiller TF, King M, Scheper MA (2008) A diffuse but subtle swelling of the upper lip. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 106:773–777
10. Cohen M, Wang MB (2009) Schwannoma of the tongue: two case reports and review of the literature. *Eur Arch Otorhinolaryngol* 266:1823–1828
11. Arda HN, Akdogan O, Arda N, Sarikaya Y (2003) An unusual site for an intraoral schwannoma: a case report. *Am J Otolaryngol* 24:348–350
12. DasGupta TK, Brasfield RD, Strong EW et al (1969) Benign solitary schwannomas (neurilemmomas). *Cancer* 24:355–356

13. Hatziotis J, Asprides H (1989) Neurilemmoma of the mandible: report of a case. *Br J Oral Maxillofac Surg* 27:512–516
14. Yang SW, Lin CY (2003) Schwannoma of the upper lip: case report and literature review. *Am J Otolaryngol* 24:351–354
15. Rosen FS, Pou AM, Quinn FB Jr (2002) Obstructive supraglottic schwannoma: a case report and review of the literature. *Laryngoscope* 112:997–1002
16. Asaumi J, Konouchi H, Kishi K (2000) Schwannoma of the upper lip. Ultrasound, CT and MRI findings. *J Oral Maxillofac Surg* 58:1173–1175
17. Kun Z, Qi DY, Zhang KH (1993) A comparison between the clinical behaviour of neurilemmomas in the neck and oral and maxillofacial region. *J Oral Maxillofac Surg* 51:769–771
18. Okada H, Tanaka S, Tajima H, Akimoto Y, Kaneda T, Yamamoto H (2012) Schwannoma arising from the sublingual gland. *Ann Diagn Pathol* 16:141–144
19. Kljancienko J, Caillaud JM, Lagace R (2006) Cytohistologic correlations in schwannomas (neurilemmomas), including “ancient”, cellular and epitheloid variants. *Diagn Cytopathol* 34:517–522
20. Humber CC, Copete MA, Hohn FI (2011) Ancient schwannoma of the upper lip: case report with distinct histologic features and review of literature. *J Oral Maxillofac Surg* 69:e118–e122
21. Hamakawa H, Kayahara H, Sumida T, Tanioka H (1998) Mandibular malignant schwannoma with multiple spinal metastases: a case report and a review of literature. *J Oral Maxillofac Surg* 56:1191–1196
22. Woodruff JM, Selig AM, Crowley K et al (1994) Schwannoma (neurilemmoma) with malignant transformation. *Am J Surg Pathol* 18:882–895
23. Hashiba Y, Nozaki S, Yoshizawa K, Noguchi N, Nakagawa K, Yamamoto E (2007) Recurrent multinodular neurilemmoma of the female upper lip. *Int J Oral Maxillofac Surg* 30:171–173