Schwannoma of the Cheek: Clinical Case and Literature Review

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Abstract

Introduction Schwannoma is a relatively uncommon benign tumor that apparently originates from Schwann cells of peripheral nerves. The most common intraoral site is the tongue followed by the palate, floor of mouth, buccal mucosa, lips and the jaws. The preoperative diagnosis is often difficult, and in the majority of cases, the diagnosis can only be made during surgery and by histological study. The immunohistochemistry reveals that the schwannoma cells test positive for S-100 protein.

Case Report The authors report here a case of an intraoral schwannoma situated in the cheek, treated by complete surgical excision. In the present case the schwannoma presented as a slow growing, circumscribed swelling without any particular features to distinguish it from other benign soft-tissue lesions. The final diagnosis was established based on the clinical, histopathologic and immunohistochemical findings.

Conclusion The presence of schwannoma calls for the careful search for nerve tumors in other parts of the body, although in most cases none may be found. The differentiation of schwannoma from neurofibroma is essential,

because an apparently solitary neurofibroma may be a manifestation of neurofibromatosis.

Keywords Schwannoma · Neurilemmoma · Neurinoma · Cheek · Oral cavity · Buccal mucosa

Introduction

The schwannoma (neurilemmoma) is a neurogenic benign tumor that arises from the Schwann cells of the nerve sheath. There is a large variety of terms for schwannomas in the literature, but only three are still in current use: neurinoma, neurilemmoma and schwannoma. Schwannoma is a relatively uncommon benign, slow-growing, encapsulated tumor that typically arises in association with a nerve trunk and as it grows it pushes the nerve aside. It may arise from cranial and spinal nerve roots or from peripheral nerves, but has a predilection for sensory nerves. Almost 25–45 % of all schwannomas occur in the head and neck region but the development of this tumor in mouth is quite uncommon representing only 1 % of all head and neck region tumors [1, 2]. Other common sites include the flexor surface of upper and lower extremities and less often the mediastinum and peritoneum [3]. Intraoral schwannoma can arise both in soft tissue or bone. Intraosseous schwannomas are most common in the posterior mandible and usually appear as either unilocular or multilocular radiolucencies on radiographs. The most common intraoral site is the tongue followed by the palate, floor of mouth, buccal mucosa, lips and the jaws [4, 5]. The schwannomas in oral soft tissue appear as a smooth submucosal swelling, thus resembling other lesions like mucocoele, fibro epithelial polyp, fibroma, lipoma and benign salivary gland tumors. However, the histological differential diagnosis is made

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with other neural origin lesions, which could be neurofibroma and neuroma, or muscular or fibroblastic origin tumor. The tumor can arise at any age but is most common in young and middle-aged adults without a gender predilection [3]. Conservative surgical excision is the treatment of choice and there is no recurrence if the tumor is completely excised. The prognosis is good and malignant transformation of benign schwannoma has been controversial.

Case Report

A 52-year-old male reported with a diffuse extraoral soft tissue swelling on left cheek below infraorbital region since 6 months.

A clinical examination revealed it to be present lateral to the left nasal wall obliterating the left nasolabial fold with slight disfigurement over the left corner of mouth. The swelling was approximately 4×3 cm in size, oval, firm, non-tender, bimanually palpable, not fixed to deep structures or overlying skin since 6 months (Fig. 1). The patient was symptom-free and reported no history of pain and paresthesia.

Intraoral examination revealed poor oral hygiene, generalized attrition and abrasion and hardness over the mucovestibule in maxillary left canine-premolar region.

A panoramic radiograph revealed a radiolucent soft tissue shadow over maxillary left canine-premolar area (Fig. 2). Fine-needle aspiration failed to yield any material. Surgical intervention was carried out under general anaesthesia. The lesion was attached to the mucobuccal vestibule (Fig. 3). A firm, rubbery, pink mass was removed in toto (Fig. 4). No attachment to the neurovascular bundle was evident. Primary closure was achieved.

Microscopically, the tissue consisted of encapsulated, well-demarcated tumor lobules composed of spindle-shaped cells with palisading nuclei and prominent Verocay

Fig. 1 Diffuse extraoral soft tissue swelling on cheek obliterating left nasolabial fold

Fig. 2 OPG showing diffuse radiolucency from the region of permanent maxillary left central incisor to first premolar with roots resorption

Fig. 3 Intra operative view of capsulated exposed lesion in the left maxilla

Fig. 4 Encapsulated lesional tissue mass of specimen

Fig. 5 Photomicrograph showing Antoni B tissue and Verocay body (×100, H&E)

Fig. 6 Photomicrograph showing S-100 positivity in tumor cells

bodies. Antoni B tissue represented the predominant microscopic pattern, with only occasional Antoni A areas along with areas of degenerative tissue (Fig. 5). An immunohistochemical stain for S-100 protein was diffusely positive (Fig. 6). Thus a diagnosis of classical schwannoma was established.

The post-operative phase was uneventful. The patient was followed-up for 6 months and had no recurrence of the tumor.

Discussion

Verocay first described neurilemmoma in 1910. He called it “Neurinoma” then. In 1935, the term ‘Neurilemmoma’ was coined by Stout [6].

Oral schwannoma is a rare solitary, slow growing, generally asymptomatic neural tumor that can present itself at any age. However it is more common between the 30 and 50 years of life [7]. William et al. findings showed that in 83 % of the cases studied by them the schwannomas presented in males, while for Lucas there was a greater predilection for females, and for Hatziotis and Asprides; Enzinger and Weiss there was an equal distribution between both sexes [8].

In most of the cases, between 25 and 45 %, extracranial schwannoma occurs in the head and neck region. In the oral cavity the lesion is usually present in soft tissues, more commonly the tongue, followed by the palate and buccal mucosa, and may have clinical aspects similar to other benign lesions like mucocoele, fibromas, lipomas, and benign salivary gland tumors [4]. In some cases the tumor could be intraosseous, being more frequent in the mandible where it may cause bone expansion, pain, and paresthesia. In these cases, differential clinical diagnosis of cysts and odontogenic tumors are commonly formulated [9].

Gallo et al. reported on 157 cases, where 45.2 % of the cases involved the tongue and 13.3 % involved the cheek. Gupta et al. reported 136 cases of schwannoma in the head and neck that consisted of 60 cases in the neck, ten cases in the parotid gland, nine cases in the cheek, eight cases in the tongue, and eight cases in the pharynx. Kun et al. reported in their study that 18 out of 49 cases were in the neck and 11 in the tongue [8].

Wright and Jackson reported 146 cases of schwannoma of the oral cavity soft tissue. Of those, 52 % involved the tongue, 19.86 % the buccal or vestibular mucosa, 8.9 % the soft palate, and the remainder 19.24 % were in the gingivae and lip [4].

Thus, the provisional diagnosis after clinical examination was of benign neoplasm of mesenchymal origin or minor salivary gland neoplasm. At the beginning, no specific hypothesis was formulated concerning schwannoma because it is a rare oral lesion, specifically at the mucobuccal vestibule.

The histological findings of the present case are similar to those reported previously, consisting of a thin fibrous capsule and a tumoral proliferation formed by two types of tissue arrangements: Antoni type A and type B. Schwann cells that are closely packed, forming bundles or arranged in rows with elongated, palisaded nuclei, characterize the Antoni type A tissue. Free bands of amorphous substance between the rows of nuclei constitute the so-called Verocay bodies that under electron microscope appear to be

composed of thin cytoplasmic processes with small amounts of collagen and basal laminar material showing frequent reduplication. Marx and Stern have referred the term ‘Verocay body’ to a structure that results due to occurrence of two clusters of palisaded nuclei around an eosinophilic mass, and the eosinophilic area consists of cytoplasmic material and replicated basal membrane [7]. While in the Antoni B tissue it has less number of cells and less organization, where the fusiform cells are widely separated, dispersed in a loose and random fashion with a network of delicate reticulated fibers. According to Enzinger and Weiss [3], the vasculature in Schwannoma is generally not prominent, but in older tumors some changes occur, like inflammation, fibrosis, and nuclear atypia. Whereas, According to Marx and Stern, schwannomas often have a prominent vascular component with dilated, irregular vessels and thick fibrotic walls. In the present case, the histopathologic analysis revealed a majority of Antoni B pattern for the whole specimen. Considering the nonorganization of the tumoral Schwann cells forming the Verocay bodies, that are classical structures of this lesion, immunohistochemical staining for S-100 was done which was diffusely positive. According to the literature review, immunohistochemical staining is essential in the diagnosis of this neoplasm. Most parts of the tumoral cells in schwannoma show immunohistochemical positive reaction for S-100 protein. According to Chrysomali et al. [6] the tumoral cells with Antoni A show greater intensity scores compared to Antoni B tumor pattern.

Microscopically, both schwannoma and the neurofibroma contain elongated cells with irregular nuclei lying between bundles of collagen fibers. They differ histologically and histogenetically, as the schwannoma is derived from the Schwann cells and the neurofibroma from the fibroblasts of the perineurium. Neurofibroma is unencapsulated, consisting of a mixture of Schwann cells, perineurial cells, and endoneurial fibroblasts [3, 6].

From the immunohistochemical viewpoint, all neural origin tumors show positivity for S-100 protein, but immunohistochemical examination could assist in lesion differentiation. Chrysomali et al. observed an intense positive reaction to S-100 in schwannoma and palisaded encapsulated neuroma. Intensive reaction to CD57 is observed in traumatic neuroma, while capsular epithelial membrane antigen (EMA), and CD34 stainings are observed in schwannoma [6].

Passador-Santos et al. studied S-100 protein, EMA, laminin, fibronectin, and collagens I and II in neural benign neoplasms. They found that schwannoma cells were intensively positive for S-100 regardless of the growth pattern. EMA was seen only in the capsule staining perineurial cells and laminin stained basement membrane around tumor cells. Fibronectin and collagens I and III

were expressed, especially in the capsule. In neurofibroma, the cells expressed S-100 whereas EMA was expressed only in a few scattered cells. Laminin and fibronectin showed a diffuse positivity. Collagens I and III showed a fibrillar arrangement with diffuse positivity [10].

The treatment for benign schwannoma consists of total surgical lesion removal. According to Asaumi et al., ultrasonography, CT, and MRI may be helpful diagnostic and treatment tools, for the estimation of tumor margins, the lesion composition, and the determination of whether there is infiltration to surrounding structures or not. The recurrences as well as the malignant transformation are rare events [11].

The presence of schwannoma calls for the careful search for nerve tumors in other parts of the body, although in most cases none may be found. The differentiation of schwannoma from neurofibroma is essential, because an apparently solitary neurofibroma may be a manifestation of neurofibromatosis [4].

The treatment of choice is pericapsular excision [7]. If the nerve of origin is visualized, an attempt should be made to separate it carefully to preserve function, although this is sometimes not possible. In our case the connection with the nerve could not be seen, as occurs in other locations of the oral cavity such as the upper lip. The schwannoma should be extirpated in its entirety to avoid tumor recurrence, even if the nerve of origin cannot be preserved.

The prognosis is very good since it does not usually recur, and malignant transformation is rare, although authors have mentioned a malignant transformation rate of 8–13.9 % [12]. Malignant schwannomas differ histologically from the benign type in their higher mitotic rate, the presence of necrosis, their infiltrative appearance and irregular positivity for the S-100 protein [13]. Since it usually develops in the extremities, malignant schwannoma is very rare in the oral cavity, although Hamakawa et al. [14] described a case in the mandible with parotid and lung metastasis. Kun et al. [15], described six cases in the maxillofacial region, two of these had malignant transformation.

Conclusion

The schwannoma is a benign tumor that is not often encountered in clinical practice. It should not be discarded when observing a submucosal soft tissue swelling of oral cavity, as in the presented case. The final diagnosis should be made after histopathological examination and in some cases after immunohistochemical analysis. The definitive treatment is the total removal of the lesion.

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