SUMMARY

Introduction Schwannomas or neurilemmomas are well demarcated, benign neurogenic lesions arising by a fibroblastic proliferation of the nerve sheath cell (Schwann cell). They usually present as solitary encapsulated lesions with rare occurrence in the upper lip. Non-diagnosed or misdiagnosed schwannomas present a high risk for the tumor to continue growing and exerting pressure on surrounding nerves. These tumours based on their location could lead to facial weakness and paralysis, pressure in ears, tinnitus, hearing loss, balance loss, and could lead to a life-threatening situation.

Case Outline This case is a rare presentation of a schwannoma located in the upper lip of a 21-year-old male patient of Indian origin. The patient complained of a swelling in the mouth with a difficulty in keeping the mouth closed. The swelling was surgically excised and the patient healed completely.

Conclusion This case of occurrence of tumor on the upper lip points to the possibility of considering schwannoma as a possibility in the diagnosis of oral tumors in the future, as the location of the tumor was rare and had a high chance of misdiagnosis.

Keywords: schwannoma; neurinoma; neurilemmoma; oral lesion; head and neck tumors

INTRODUCTION

Schwannoma was first described by Vercay in 1910, who called it neurinoma [1]. But the term neurilemmoma was first coined by Stout in 1935 [1]. Neurilemmoma produces distinct patterns referred to as Antoni A and Antoni B [2]. It has a predilection for head, neck, and surface flexors of the upper and lower extremities [3], with 25–45% of all schwannomas occurring in the head and neck region, the tongue being the most common site [4]. Cranial nerves I and II are not sites for this tumour as they lack Schwann cells [5]. One percent of schwannomas occur in the intraoral region [6, 7]. Among the intraoral lesions, tongue is the most common site, with rare occurrence in the upper lip [7, 8].

On examination it was an ovoid, firm, mobile mass approximately 3 × 2 cm in size, exhibiting a smooth, non-ulcerated, non-erythematous surface. The swelling was in the midline and extended to the labial vestibule (Figure 2). With a provisional diagnosis of traumatic fibroma, an excisional biopsy was performed under local anesthesia. The lesion was encapsulated and this facilitated the meticulous dissec-
tion. The surgical bed was thoroughly cleaned, hemostasis achieved, and wound closure was done (Figure 3).

The gross specimen was yellowish with a smooth, shiny surface (Figure 4). Upon histopathologic examination, both Antoni type A tissue, made up of cells with spindle shaped nuclei arranged in a palisading pattern, and Antoni Type B tissue, showing disorderly arranged cells and fibers, were seen (Figure 5). A diagnosis of schwannoma was made.

The postoperative period was uneventful and the patient is disease free after a year of follow-up.

DISCUSSION

Schwannoma is also known as neurilemmoma, neurinoma, lemmoma, and perineural fibroblastoma [9]. Tissue culture studies by Murray and Stout confirmed the Schwann cell origin when they cultivated the tumour in vitro. They usually present as solitary encapsulated slow growing lesions unless associated with neurofibromatosis. Despite the nerve tissue origin, they are painless. They cause pain only when they cause pain on adjacent nerves, rather than on the nerve of origin [9]. This case showed gradual increase in size and was otherwise asymptomatic. Frequency of lip lesions is comparatively less [8].Infraorbital nerve schwannomas can present as lip masses [10]. Rarely, multinodular neurilemmomas are also seen [11]. Central lesions which cause bony destruction can pres-
ent as unilocular or multinodular radiolucencies that are centered on the inferior alveolar nerve [2]. They may have a true capsule or a pseudocapsule made of fibrous connective tissue [4]. This lesion was encapsulated, which aided in complete removal.

Ultrasound scan with fine needle aspiration biopsy can be diagnostic in 30% and magnetic resonance imaging in 77% of cases [12]. Ultrasound scans show homogenous and hypo-echogenic findings and post-acoustic enhancement. Computed tomography scans show definitely marginated mass with homogenous soft tissue density. Magnetic resonance imaging scans demonstrate a homogenous lesion with low intermediate signal intensity on T1-weighted and high signal intensity on T2-weighted images [13]. This mass was not subject to any such investigation as we relied entirely on our clinical assessment.

Treatment of choice is surgical excision [14]. Recurrence is uncommon [4, 7, 14]. Malignant transformation of schwannomas is rare [7, 15]. Das Gupta and Brasfield [16] reported 8% incidence of malignant schwannomas in the head and neck region. Ghosh et al. [17] reported 13.9% incidence. A provisional diagnosis of traumatic fibroma was made based on the prior history of trauma. Minor salivary gland neoplasms and mesenchymal tumors can also be considered as possibilities.

Histopathology shows two types of tissue – Antoni type A and Antoni type B. The cells in Antoni Type A have elongated or spindle shaped nuclei aligned to form a characteristic palisading pattern. Intercellular fibers are arranged parallel to the nuclei, giving the impression of organoid swirls [4]. Verocay bodies are central, acellular, eosinophilic bodies with reduplicated basement membrane and cytoplasmic processes. Antoni type B tissue shows oval nuclei and disordered cells and fibers with edema fluid and microcysts [9]. There is no myelin as no axis cylinders exist to induce myelin formation by the Schwann cells. Tumor cells also show diffuse positive immunohistochemical reaction for S-100 protein [9]. This lesion exhibited all the classic histopathologic features. If the nerve of origin is visualized, all attempts should be made to isolate it [18]. Here the nerve of origin could not be identified.

It is customary to submit all excised tissue for histopathologic analysis. This case report underscores the importance of the above tradition. Though lesions like the schwanna are the exception rather than the norm, it is becoming of a prudent clinician to be on the lookout for such rare entities.

REFERENCES

Шваном горње усне – приказ болесника и преглед литератури

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САЖЕТАК
Увод Шваноми или неуриноми су јасно ограничени, бениним тумори нервног порекла који настају фибробластном пролиферацијом омотача нерва (Шванових ћелија). Обично се манифестују као солитарне инкапсулиране лезије, а ретко се развијају у горњој усни. Не постављање адекватне дијагнозе шванома представља велики ризик за даљи раст тумора и појаву притиска на суседне нерве. Ови тумори у зависности од локализације могу довести до слабости или парализе, осећаја притиска у уху, тинитуса, губитка слуха и равнотеже, као и по живот опасних стања.

Приказ болесника Овај случај представља ретку манифестацију шванома у горњој усни 21 године старог мушкарца индијског порекла. Пацијент се жалео на отицање у пределу усне дупље и тешкоће да споји усне. Тумор је хируршки уклоњен и рана је уредно зарасла.

Закључак Локализација шванома у горњој усни је изузетно ретка, али се мора разматрати у диференцијалној дијагнози тумора усне дупље јер лако може доћи до превида.

Кључне речи: шваном; неурином; неуринлемом; орални тумор; глава и врат