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SOLITARY NEUROFIBROMA OF THE CHIN

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Abstract

Tumors originated from peripheral nerve tissues are neurofibroma, neurilemoma, plexiform neurofibroma, malignant schwannoma, and granular cell tumor.

Neurofibromas seem to occur in two forms : The first one is circumscribed solitary neurofibroma and the second group is neurofibromatosis or Von Recklinghausen's disease, which is a congenital and familial disease, presenting abnormalities of the skin, nerve system, bones and soft tissue.

A solitary neurofibroma is a benign, relatively circumscribed, noncapsulated tumor which often presents in the skin and subcutaneous tissue as a soft sessile or pedunculated mass.

It may occur anywhere in the head and neck, but the common site of the occurence is the tongue, buccal mucosa, palate in frequency.

Since solitary neurofibroma is a relatively radioresistant and its recurrence rate seems to be low, the treatment of choice is surgical excision.

The author would like to present a case of unusually large solitary neurofibroma occured in the chin, which was successfully treated with surgical excision and reconstructed using deltopectoral flap and tongue flap.

INTRODUCTION

Neurofibromas are developmental disorders of nerve sheaths although they present a tumor-like formations. There is proliferation of tissue of the nerve sheaths in which both the fibrous and the neurolemmal elements participate in different degrees and with remarkable irregularity. It most frequently involve the skin or oral mucosa and does not differ from the disseminated or multiple form of the disease, known as neurofibromatosis or Von Recklinghausen's disease of skin, except that systemic or hereditary factors are usually not present[1].

Crawford[2] required evidence of at least two of the following four clinical findings to make the diagnosis of neurofibromatosis : Café-au-lait spots, familial history, characteristic histologic findings and associated bony lesions.

The structure of the neurofibroma is usually not well circumscribed or encapsulated : the lesion mingles with the surrounding dermal and other tissues or it involves nerves diffusely for variable distance. Nerve fibers often seem dispersed in the tumor mass. There was a striking predilection for the soft tissues of the eyelid, orbit, nose and cheek whereas few lesions were seen in the scalp, mandible, neck, lip, parotid, ear and zygoma.

Histologically, the tumor exhibits considerable variation in histologic structure but is generally composed of proliferation of delicate spindle cells with thin, wavy nuclei, intermingled with neurites in an irregular pattern as well as delicate, intertwining connective tissue fibrils. Cellular and myxoid patterns predominate, organoid features are not present. The treatment of choice is conservative surgical procedure.

In this article, a case of solitary neurofibroma treated by surgical excision and reconstructed using deltopectoral flap and tongue flap will be presented and discussed.

REPORT OF A CASE

A 43-year-old female patient was seen in the department of Oral and Maxillofacial Surgery of Fatima Hospital complaining of a large soft tissue mass on the chin.

The mass on that area was first noticed 20 years ago and it grew very slowly to the size of a large orange measuring 10cmx 10cm. The mass was relatively soft and movable and it had pedunculated shape. Overlying skin retained its normal coloring and surface ulceration was not present (Fig. 1).

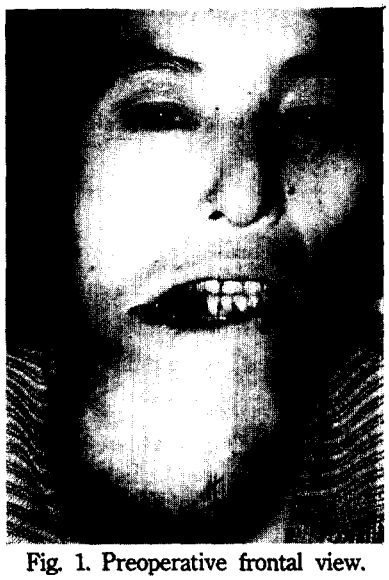

Fig. 1. Preoperative frontal view.

The patient had difficulty in lip closure, and on oral examination several decayed teeth, some amalgam restorations and chronic periodontitis were noticed. Soft tissue mass extended to the right cheek, the vestibule and gingiva of 28, 29, 30 teeth (Fig. 2). Paresthesia, abnormal auditory or ocular findings were not noticed.

Fig. 2. Preoperative intraoral view.

Physical examination failed to reveal additional lesions that might have represented neurofibromas and no menifestations of neurofibromatosis including Café -au-lait pigmentation of the skin were identified[3,4,5]. Roentgen examination revealed no gross abnormality of the bones. A biopsy was obtaind from the chin area of the lesion. The obtained specimen had a uniform grayishwhite color and a rubber-like texture. Microscopic examinations showed the lesional tissue to be a mesenchymal tissue tumor composed of proliferating axon and schwann cells with undulating cell process. The diagnosis was neurofibroma.

The patient was admitted to our department for operation. The review of medical, social and familial history yielded no significant findings. Findings of physical examination, the results of routine laboratory studies, chest X-rays and electrocardiogram were noncontributory.

Uuder general anesthesia with Halothane, N_2O and oxygen, the bulging mass was excised and the defective area was reconstructed with tongue flap intraorally and right deltopectoral flap extraorally (Fig. 3, Fig. 4). The gross description of the lesion was that of irregular rounded mass measuring approximately 10cm×10cm×8cm. The wound and flaps healed uneventfully but due to shallow vestibule and slight contraction in the right cheek, the patient underwent vestibuloplasty and scar revision 8 months later (Fig. 5, Fig. 6).

The color of the deltopectoral flap on the chin had improved. So it matches well with the face at present time (Fig. 7, Fig. 8). Mandibular denture was delivered after the vestibuloplasty and the patient is now able to use it without much discomfort.

Fig. 3. Raising tongue flap.

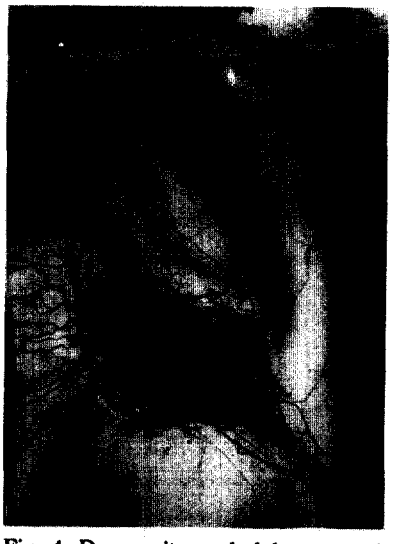

Fig. 4. Donor site and delto-pectoral flap.

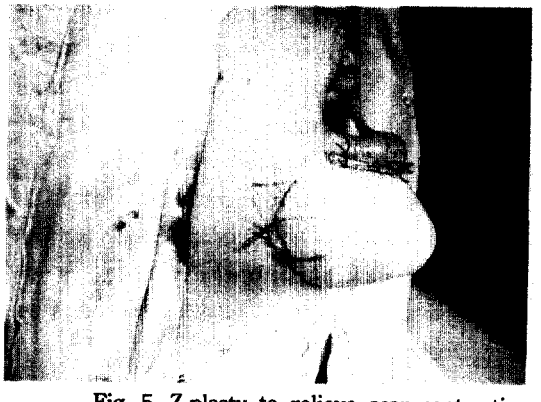

Fig. 5. Z-plasty to relieve scar contraction.

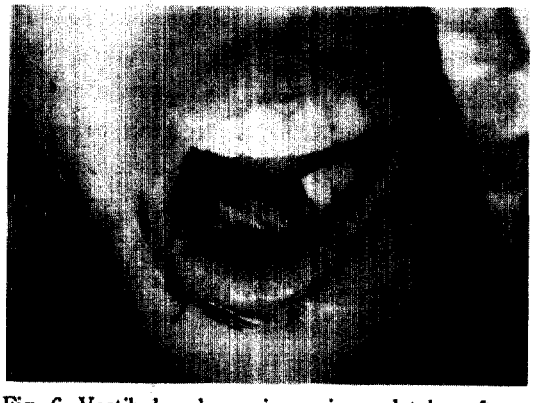

Fig. 6. Vestibular deepening using palatal graft.

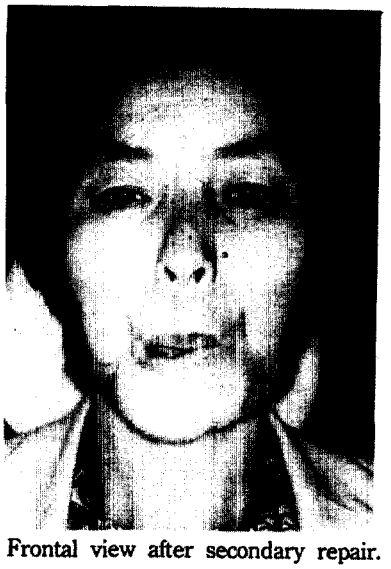

Fig. 7. Frontal view after secondary repair.

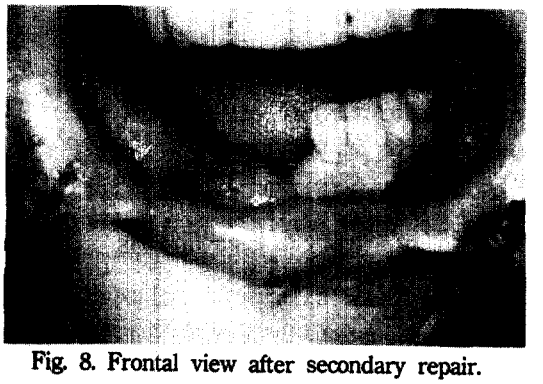

Fig. 8. Frontal view after secondary repair.

DISCUSSION

Tumors arising from peripheral nerve tissue are neurofibroma, neurilemoma, plexiform neurofibroma, malignant schwannoma and granular cell tumor.

A neurofibroma is a relatively benign circumscribed but not encapsulated schwann cell neoplasm. It most frequently involves the skin or oral mucosa and does not differ from the disseminated or multiple form of the disease known as neurofibromatosis, or Von Recklinghausen's disease of skin, except that systemic or hereditary factors are usually not present.

Some authors, such as Adekeye[6], Lorson[7], Rittersma[8], Shapira[9], Winters[10] and others reported neurofibromatosis with head and neck lesion. Gnepp, Hunt and Kim reported central neurofibroma of the jaw bone. One article by Vincent and Williams[11], skeletal involvement in neurofibromatosis had been reported to occur in 51 to 71 percent of patients.

Neurofibromas may occur in any of the oral structures including the tongue, the buccal mucosa, the alveolar ridge, the gingiva, the lips, the palate, the floor of the mouth, and the pharyngomaxillary space [12~15]. The overall frequency of oral manifestations are generally reported to be between 4% and 7%. However in a recent investigation by Shapiro and coworkers[16] which included both oral and panoramic radiographic examinations, the frequency of oral manifestations of neurofibromatosis was 72%. Emmanuel, Oladepo, Adekeye et al[6] in the review of 28 patients with neurofibromas of the head and neck, reported that the incidence was greater in males (60.7 %) than in females (39.3%). The duration of clinical symptoms ascertained in 17 patients ranged from 5 months to 15 years with a mean of 6.3 years. There was a positive family history in 4 (14.3%) patients.

According to Shklar and Meyer[17], classification of benign neurogenic tumors of the oral cavity are the neurofibroma, schwanoma, and traumatic neuroma. It is important for the clinician to distinguish between the neurofibroma and the schwannoma, as the potential clinical behavior of the two can be different. The neurofibroma is an unencapsulated neoplasm containing small nerve fascicles in an unorganized collagenous matrix. The lack of encapsulation allows the tumor to infiltrate contiguous tissue thus making a complete surgical removal difficult[18]. The solitary neurofibroma is a benign, slowly growing, relatively circumscribed, but nonencapsulated neoplasm originating in a nerve and consisting of schwann cells, perineural cells and varying amounts of mature collagen[19].

Histologically the neurofibroma exhibits considerable variation in histologic structure but is generally composed of a proliferation of delicate spindle cells with thin, wavy nuclei intermingling with neurites in an irregular pattern as well as delicate intertwining connecutive tissue fibrils. Cellular and myxoid patterns predominate, organoid features are not present. Melanocytes may sometimes be found in the tumor and mast cells are common. The treatment for solitary neurofibromas is surgical excision. X-ray radiation is of no value and recurrence rate seems to be low. Lesions which have undergone sarcomatous transformation have a poor prognosis, although occasional survivals are recorded following surgical removal of the tumor.

SUMMARY

A case of solitary neurofibroma of the chin occured in 43-year-old female patient is reported. The tumor mass was surgically excised, and the defect area was reconstructed with tongue flap and deltopectoral flap.

The patient had an unremarkable postoperative course and is currently in 3 years of postoperative period sustaining good appearance and function without recurrence.

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이부에 발생한 신경섬유종

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장세홍 · 안재진 · 정민원

국문초록

말초 신경 조직에서 발생되는 종양은 신경섬유종, 신경초종, 과립세포종, 악성신경초종 등이 있다.

신경섬유종은 2가지 형태로 분류될 수 있다. 첫째 형태는 단독형 신경섬유종이고, 둘째는 신경섬유종 혹은 Von Recklinghausen's 병으로 불리우는데 이는 주로 피부, 신경조직, 뼈, 연조직에 기형을 유발할 수 있고, 선천적이며, 가족력을 나타낸다.

단독형 신경섬유종은 양성이며, 경계는 명확하나 capsule에 의해 싸여 있지 않으며, 피부나 피하조직에 sessile 혹은 pedunculated 형태로 나타난다. 두경부의 어느 부위에나 나타날 수 있으며, 구강내의 호발 부위는 혀, 협점막, 구개 순이다.

단독형 신경섬유종은 방사선에 의한 치료에 효과가 적으며, 재발율이 낮고, 치료는 주로 외과적 절제술로 제거를 하고, 결손부를 삼각흉피판(D-P flap)과 설피판(tongue flap)으로 수복하여 현재까지 합병증 없이 양호한 결과를 얻었다.