Solitary Neurofibroma of the Palate
A Very Rare Case

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ABSTRACT
Neurofibroma (NF) is a benign nerve tumor originating from the peripheral nerve sheath. NF can occur as a solitary lesion or as part of the Von Recklinghausen disease syndrome. The solitary type rarely involves the head and neck regions, especially the oral cavity. However, few reports mention that NF appears on the buccal mucosa, floor of the mouth, lips, and gingiva. This paper reports a very rare case of solitary NF in the palate. Has been reported, 37-year-old male patient with a solitary neurofibroma of the palate that is not associated with systemic symptoms. Complete mass extirpation has been carried out using the oral method and the transpalatal approach. This case has a good prognosis but the risk of recurrence, malignancy transformation or associated neurofibromatosis syndrome may occur so observation needs to be done.

Keywords: *Neurofibroma, palate, very rare.*

1. INTRODUCTION

Neurofibroma (NF) is a benign nerve tumor originating from the peripheral nerve sheath and consists of Schwann cells, perineurial-like cells, and fibroblasts. NF can occur as a solitary lesion or as a part of the Von Recklinghausen disease syndrome. The solitary type rarely involves the head and neck region, especially the oral cavity. However, NF in the oral cavity has been reported in the buccal mucosa, floor of the mouth, lips, and gingiva [1]. This paper reports a rare solitary NF in the palate

2. CASE REPORT

A 37 year old male patient with the initial H arrived at the otolaryngology-head and neck policlinic at PKU Muhammadiyah Gombong Hospital with a chief complaint of something blocking the roof of his mouth since 3 months prior. A mass appeared on the palate 1 year prior to the visit and has since grew in size resulting in the blocked feeling. The bump was painless, did not bleed, nor itch.

The patient also reported experiencing nasal congestion since 1 month prior to the visit. Epistaxis, olfactory disturbances, and runny nose was denied. The mass did not interfere with swallowing, nor did it cause shortness of breath or voice change. Severe migraine, double vision, eyeball protrusion, vision disturbances, and watery eyes were denied. Mobile teeth and other oral lesions were denied. Buccal protrusion, soreness, or numbness was denied.

The patient reported a history of a similar lesion that almost invaded the whole oral cavity 10 years ago that interfered with the ability to swallow and breathe. The patient was treated at Sardjito Hospital where the mass was extirpated and an airway was created in the neck. The mass was found to be benign. History of hypertension, diabetes, and weight loss was denied. A history of similar diseases in the family or other parts of the body was denied. History of smoking and consuming alcohol was denied. History of consuming salted fish and food containing preservatives was denied. History of undergoing phototherapy was denied.

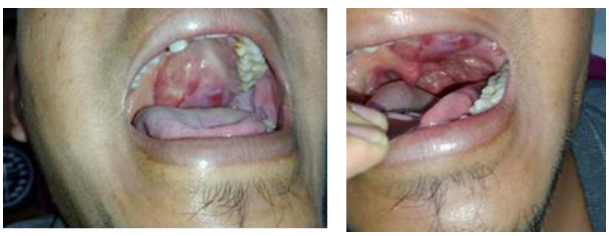
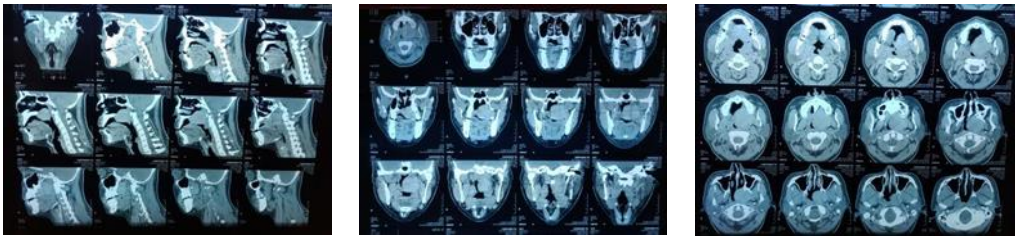


Figure 1. Oral Cavity Examination

General examination from head to toe (excluding otolaryngology-head and neck) of the patient revealed within normal conditions, no generalized nodules, vital signs results were: blood pressure 120-80 mmHg, pulse rate 70x/minute, breathing rate was 22x/minute, temperature was 36℃. The oral examination can be seen in Figure 1, a white and reddish mass was found from the hard palate expanding to the left side of the soft palate and crossed the midline. The shape of the mass was ellipse and it had a smooth surface and a defined border, its' texture was springy. No fluctuation was detected. The mass was painless and upon measurement it was 5cmx3cmx1cm. The adjacent teeth were vital. The uvula was symmetrical, centered, and not hyperemic. Granulated tissue was observed lateral to the uvula, extending from the left palate-glossus arch to the left retromolar trigonum. Tonsil T1-T1 was normal with widened crypts and no detritus were present. Pharynx wall was normal, gag reflex was detected. The tongue was normal, no mass was present, normal mobility was observed. Scar tissue on the anterior of the trachea was found, and there was no enlargement of the lymph nodes.

During examinations using rigid Hopkins nasal endoscope 0º elevation of the mucosa was detected in the posterior aspect of the left nasal cavity. The mucosa was white and reddish in color, had hypervascularization, and slightly blocked the left choana from the inferior. The posterior and lateral wall of the left nasopharynx shifted as a result. No deviation of the septum nor post nasal drip was found, and inferior conchae was eutrophic.

Palatal biopsy state that was proliferation of epithelial squamous cells, hyperkeratosis, acanthosis, and parakeratosis. The epithelial squamous cells were relatively normomorphic. The sub-epithelial tissue was connective tissue and inflammatory cells. No malignancy signs or distinct characteristics was found. The conclusion was squamous hyperplasia (keratosis).



(a) (b) (c)

Figure 2. Head and Neck CT scan with Contrast (a) sagittal view, (b) coronal view, (c) axial view

CT scan images show isodent inhomogeneous lesions (HU=56-71) (Figure 2.a). Its' shape was round, regular, defined border, negative calcifications, negative necrotic centers. Upon measurement its' size was 51x52mm (sagittal cross section), 47x33mm (axial cross section), extended from posterior palate – posterior of the left molar – expanding to the right side, shifted the posterior side of the septum nasal to the nasopharynx. Post contrast administration showed high attenuation (HU=76-94) (Figure 2.c). Visualized intracerebral was within normal range. No enlargement was found on the parotid and submandibular glands. The bones were intact. The Oro nasopharynx tumor gave an impression of malignancy. Examination of thorax imaging found no metastases; however, abdomen hepatobiliary USG was not performed. The patient was diagnosed with recurrent palate tumor and differential diagnosis was keratosis, pleomorphic adenoma, mucoepidermoid carcinoma, adenocarcinoma, lymphoma, neurofibroma, lipoma, fibroma, schwannoma, and hemangioma.

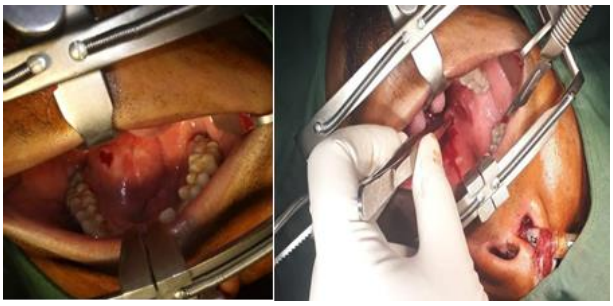


Figure 3. Surgery: Transoral Method with a Mouth Gag and Transpalatal Incision.

The mass was extirpated under general anesthesia using transoral method with a trans palatal approach (Figure 3). The patient's head was put in an extension position, intubated through the nose and the operator was positioned behind the patient's head. A mouth gag and cheek retractor were used to gain maximum surgical field access. An incision was made on the midline of the mass, starting along the posterior of the hard palate to the soft palate while preserving the uvula (Figure 4). A flap was made layer by layer while it was cauterized to control bleeding and reveal the mass.

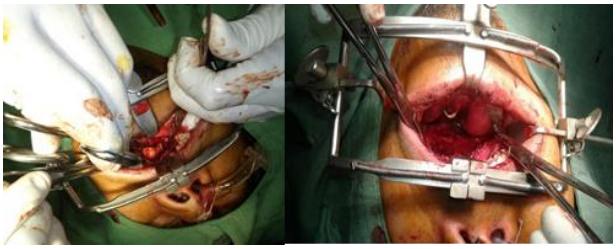


Figure 4. Surgery: Identified and Extracted Mass

The mass was released from adjacent healthy tissues. Because of its' large size, partial removal with cauterization was applied to maximize view of the surgical field. A frozen section biopsy of the tissue was done and the results show the mass was benign. Releasing the mass from adjacent healthy tissue was continued to the cranial direction, the left minor palatal artery was identified at the hard palate junction and bleeding was controlled. The remaining mass was removed using the metzenbaum scissors and maximally cleaned from the healthy tissue using takahasi forceps (Figure 4). The mucosa of the posterior nasal cavity and nasopharynx border was preserved. After the removal of the mass and the bleeding was controlled, the cavity left by the mass was rinsed with NaCl solution and packed with spongostan to minimize dead space, afterwards the flap was closed and stitched layer by layer

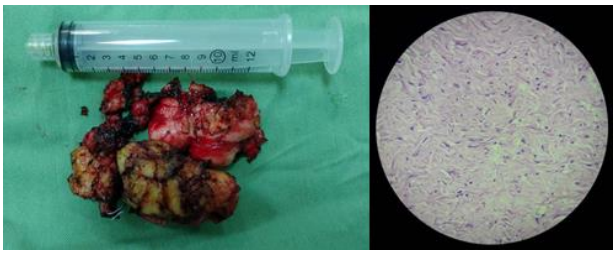


Figure 5. (a) Specimen and (b) Anatomical Pathology Results

Three brownish-white specimens with sizes ranging from 2,5x2x1 cm (smallest) to 4x3x2 cm (largest) were obtained (Figure 5.a). Anatomical pathology examinations showed mesenchyme tumor tissue in the form of proliferation of fibrous connective tissue, partially hyalinized (Figure 5.b). Tumor cells was relatively normal in morphology. The nucleus was spindle with pointed ends. No malignancy was found. It was concluded that the mass was neurofibroma. Immunohistochemistry examination was not performed. After surgery the patient did not exhibit nasal voice, was able to swallow water and solid food, no shortness of breath was observed, and surgical wound was normal. The nasogastric tube was removed after 1 week during post-surgery appointment.

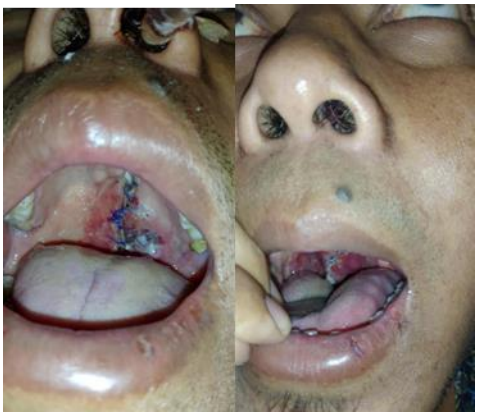


Figure 6. Post-surgery

3. DISCUSSION

Neurofibroma is a benign tumor originating from the peripheral nerve sheath. The two main forms of neurofibroma are periphery or neurofibromatosis type I (NF-I) and central or neurofibromatosis type II (NF-II).[1] NF-I was first described in 1982 by Recklinghausen, a German anatomy specialist, as a classic form of the von Recklinghausen skin disease. The diagnostic features of NF-I are multiple NF lesions across the body, several hyperpigmented macules on the skin, optical glioma, pigmented hamartoma of the iris, distinct osseous lesions such as sphenoid dysplasia and immediate family history with neurofibromatosis. NF-II has a lower prevalence compared to NF-I and is characterized with the presence of bilateral acoustic neuroma, and is possibly linked to multiple endocrine neoplasia III [2].

Neurofibroma is relatively common compared to other neural lesions affecting the head and neck, with a prevalence of 37%. This may be a result of nerve richness in the head and neck area [3]. However NF in the oral cavity and oropharynx is rare, ranging from 2% to 7% of cases, and mostly linked with NF-I [4]. The most commonly affected nerve of the oral cavity are the trigeminal nerve and the superior cervical nerve. The most common affected site is the tongue, followed by the buccal mucosa, floor of the mouth, lips, and gingiva [3]. Solitary NF lesions on the palate is a rare variant [5]. The tumor discussed in this paper is most likely a benign tumor considering the results of anatomical pathology, anatomical area, and is linked to a syndrome.

No racial or sex inclination has been found in oral NF. This tumor can occur in a wide range of ages from 10 months to 70 years old, with an average of 45 years old [6]. The direct cause of solitary NF is unknown, however it is hypothesized that this disease is a hyperplastic hamartomata's malformation as opposed to a neoplastic condition. It is assumed that NF originates from Schwann cells, perineural cells, and fibroblasts [7]. The patient in this report was a 37 year old male, not the average age of NF. When linked to the possibility of recurrence, it was

traced that NF of this patient first appeared when the patient was 27 years of age.

Oral lesions are usually slow in progression, are painless, distinctively enlarged and stemmed without mucosal ulceration. The color of the mucosa varies from normal to red or yellow. When pressure is applied sometimes pain appears, and the mass usually disrupts physiological functions. Oral NF is a non-odontogenic tumor with differential diagnoses such as Schwannoma, fibroma, traumatic lesion, neuroma, lipoma, or salivary gland tumors [8]. This patient experienced slowly progressing complaints without neurological deficits, disturbance of speech, swallowing, and breathing.

Microscopically, NF and schwannoma are similar as both contain elongated cells with unorganized nucleuses between collagen fibers. The origins of schwannoma is Schwann cells around the nerves while NF originates from perineurium fibroblasts. A schwannoma consists of a mixture of Schwann cells, perineural cells, and endoneuria fibroblasts, and it is capsuled unlike NF. Immunohistochemistry analysis is able to confirm NF and differentiate NF from other benign neural lesions. NF is immunopositivity towards S-100 protein in 85-100% of cases [8]. Anatomical pathology of the specimens from this case revealed a benign tumor supporting neurofibroma, however due to limitations of facilities immunohistochemistry analyses was not performed.

It has been explained above that ultrasonography, CT scan, and MRI are diagnostic modalities to determine tumor margins and to view infiltration to its' surrounding structures. If bone erosion was found during CT scan, malignancy would be suspected [9]. A CT scan with contrast was ordered for this patient and malignancy was suspected with a relatively large size, however bone erosion was not detected. Although clinical examination leans towards a suspicion of benign tumor, the definitive diagnosis of the patient was based on histopathologic examination.

Treatment of solitary NF is total excision of the mass. However recurrence is prevalent because the tumor is non-capsulated and the disease is infiltrative in nature [10]. This may be prevented by total resection with tumor-free margins. Recurrence happen in 20% of NF patients who underwent total resection and increases to 44% in patients with subtotal resection [11]. Total surgical excision in the head and neck may present a challenge as NF is infiltrative, and difficulty increase when multiple lesions are present. Risk of neurological deficits increase in cases of multiple lesions. Tracheotomy may be useful in large oral or oropharynx NF and decreases difficulty in anesthesia. The most common complication during surgery is bleeding [12]. A transoral total resection using trans palatal approach and frozen section. Anesthesia was achieved per nasal without the need for tracheostomy.

Malignant transformation of benign NF is rare and are usually linked to schwannoma radiotherapy [13]. The most common malignancy is neurofibrosarcoma, which occurs on 5% of cases. Neurofibrosarcoma is an aggressive tumor with a poor prognosis [14]. Malignant transformation rarely occurs on small solitary NF. However, patients with neurofibroma type I have a 8-13% risk of malignant peripheral nerve sheath tumor (MPNST). Most MPNST cases arises in plexiform neurofibroma compared to solitary neurofibroma, and appears as a larger metastasized tumor. The prognosis of this malignant tumor linked to NF-I is very poor with a 21% 5-year life expectancy [15]. Regular observation after surgery is critical to detect recurrence or malignant transformation and its' link to generalized syndromes.

4. CONCLUSION

It was reported that a 37 year old male patient was diagnosed with neurofibroma of the roof of the mouth (palate) which was solitary and not connected to any systemic symptoms. The possibility of recurrence was found, with a time frame of over 10 years. Mass extirpation was performed orally using trans palatal method.

Solitary neurofibroma of the oral cavity is very rare, however they have been found on the tongue, buccal, floor of the mouth, lips, and gingiva. This case is a rare case of solitary NF on the palate with a good prognosis. The risk of malignant transformation, neurofibromatosis syndrome and recurrence are possible thus observation is required

CONSENT FOR PUBLICATION OF THE IMAGE

Author declare that the patient or guardian agreement to published her images on this journal.

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