Case Report

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Intraoral Nevus of Ota: A Case Report and Review

Abstract:

The Nevus of Ota is a melanocyte disorder that primarily affects skin and eye regions innervated by the first two caricatures of the trigeminal nerve, the ophthalmic and maxillary divisions. In most instances, pigmentation involves the skin, eyes, and rarely the intraoral mucosa. The oral cavity is not always involved in cases of opthalmic or dermal melanosis. To the best of our knowledge, fifty five cases of Ota nevus involving the oral cavity have been documented. We have summarized all the cases of nevus of Ota involving the oral cavity until today. We are presenting a case of palatal nevus of Ota in a 50-year-old female patient.

Key-words: Palatal Ota, Trigeminal Nerve, Dermal Melanosis

Introduction:

"Nevus of Ota," first reported by Ota and Tanino in 1939, includes the skin near the classification of the first and second estrangements of the trigeminal nerve.[1] The condition happens on account of the failure of melanocytes' migration from the neural crest to the derma-epidermal junction. An entrapped mass of melanocytes imparts it a characteristic gray-blue pigment. The condition is most common in Asian and Indian Subcontinent populations, with an estimated prevalence of 0.014% to 0.034%. In a 5:1 ratio, females are far more affected than males.[2] Presentation on one side of the face is more frequent than on both sides of the facial skin. Apart from dermal and opthalmic involvement, oral mucosal engrossment is rarely visualized.[3,4] Even though intraoral pigmentation of nevus of ota is exceptionally rare, only fifty five cases, including the author of this article, have been described in the English literature (Table 1), and the palate appears to be the most commonly affected intraoral site.[5] Apart from the palate other intraoral nevi of Ota where cases have been documented include the tongue, buccal mucosa, and gingiva, but the palate is the most common presentation, possibly because the origin of the greater and lesser palatine nerves from the maxillary nerve is more proximal than the superior, middle, and anterior alveolar nerves, and involvement of the mandibular division is extremely rare.

Case Report:

A 50-year-old female patient reported to the department of oral medicine and radiology at the Government College of Dentistry, Indore, with the chief complaint of pain and decayed teeth in the upper right posterior tooth region of her jaw for the last ten days. During the course of her current illness, she reported constant, dull aching discomfort in her right upper first molar for the previous two months. This patient's extraoral examination revealed a bluish-gray

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hyperpigmentation present on the right side of the face involving the innervations of divisions one and two of the trigeminal cranial nerve, along with a pigmented area around the right eye, pigmentation of the right eye sclera, the right temporal region, and the right half of the forehead skin since childhood (Figures 1a, 1b, and 1c).

Figure 1a: Extraoral photograph reveals blue-grey skin pigmentation over right side of face involving forehead, periorbital area and cheek region. **Figure 1b:** Hyperpigmentation involving right temporal and zygomatic region upto angle of mouth. Not involving mandibular region. **Figure 1c:** Episcleral involvement in right eye

Even though there was no itchiness or irritation in the pigmented area of the face, the patient stated that the pigmentation had grown in size since childhood. Her prior medical history was non-contributory. An intraoral examination revealed a bluish pigmentation the size of about 0.5x0.5 cm on the right side of the hard palate. (Figure 2)

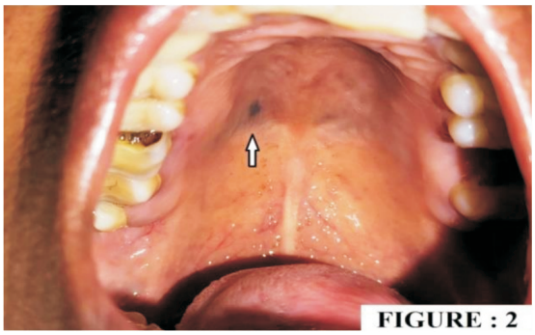

Figure 2: Approximately 0.5x0.5 cm melanotic macule right side of the hard palate just lateral to mid palatine raphae (White arrow)

Our patient was advised to have a biopsy but did not consent to one because she was completely asymptomatic. The presence of characteristic associated pigmentation over the facial skin, eyes, and palate confirms the clinical diagnosis of nevus of Ota. Sturge-Weber syndrome, blue nevus, melasma, Mongolian spot, nevus flammeus, blue nevus, and acquired bilateral nevus of ota like macule (ABNOM) are all possible differentials for nevus of ota (Table 1).

Table (1) Possible differential diagnosis of nevus of Ota [2,10,11]

Differential Diagnosis	Clinical Manifestations
ABNOM	Late onset bilateral symmetrical pigmentation with rare involvement of eye/oral mucosa.
Melasma	Bilateral brownish dark to brown black pigmentation in middle age women without involvement of mucosa.
Mongolian spot	Pigmentation limited to lumbosacral area vanished by 36 years of age.
Nevus of Ito	Pigmentation limited to shoulder area
Nevus flammeus	Affected skin and mucosa more reddish
Blue nevus	Well defined ,elevated lesion, diameter less than 1 cm.
Sturgeweber syndrome	Characteristic port-wine stain one side with disorder of nervous system i.e. seizures.

*ABNOM, acquired bilateral nevus of Ota-like macules

Discussion

Melanocytic nevi in the oral cavity are extremely uncommon in comparison to those in the skin and eyes. Recently, a palate nevus of OTA in association with H. pylori gastritis has also been documented in the literature.[6] On the basis of pigmentation, intensity, and area of involvement, Tanino classified it into four subtypes. It is also interesting to note that in all subtypes of nevus of Ota, there is no involvement of the mandibular region. [7] According to classification, our case will match the characteristics of subset type III.

There aren't many cases of ota nevus developing into malignant melanoma involving the dermis and the ocular aspect. Similar to incidence in females, nevus of ota conversion to malignant melanoma is also common in this gender as compared to males. Dermal nevus of ota into malignant melanoma has been reported in twelve cases, while in the case of ocular aspect it is sixteen with female predominance. The MAP kinase pathway has been nevied to eight genes, including BRAF and NRAS.A G-coupled protein mutation causes continuous activity of these receptors.[9] Females had higher levels of G-protein-coupled kinases, which may be the cause of female dominance compared to males.[10] Studies on whole-exome sequencing in dermal benign nevi of the ota have suggested mutations in the GNAQ, MMP10, BAP1, COL4A4, FN3K, and PDL3 genes. [11]

This being a deep dermal pigmentation, various treatment modalities like cryotherapy, surgical excision, chemical peeling, dermabrasion, and lasers are being used to cure this disfigurement. In the current treatment scenario, Q-switched

ns-domain laser has emerged as a first-line treatment modality for nevus of Ota, but it is associated with a number of complications for patients post-operatively. In an attempt to minimize hyper pigmentation, hypo pigmentation, and scar formation, the Pico second pulse alexandrite laser is also being used, as it requires a short treatment with a short duration of the treatment, resulting in fewer complications in comparison to the Q-switched ns-domain.[12]

Table (2) Documented cases of Intraoral Nevus of Ota

Ref No.	Gender	Age (yrs)	Intraoral location	Country/City Name	Remark
1.	Male	16	Buccal mucosa right side	UK	White
2.	Female	35	Along palate midline and left border of tongue	USA	Negro
3.	Male	45	Palate (midline)	USA	Negro
4.	Male	30	Palate bilateral	Japan	
5.	Male	23	Right buccal mucosa	Brazil	White boy
6	Female	16	Right side of the palate	India (Varanasi,UP)	Hindu
7.	Female	43	Left side of the palate	Non-oriental	White
8	Female	27	Buccal mucosa	Non-oriental	White
9	Female	63	Buccal mucosa		
10	Female	59	Palate bilaterally more on left side	USA	White
11	Female	30	Palate	India	
12	Female	30	Palate bilaterally	India	
13	Female	32	Palate	India	
14	Female	26	Left buccal mucosa and palate	Germany	White
15	Female	32	Hard palate	India (Karnataka)	
16	Male	33	Bilaterally (more right side)	India (Karnataka)	
17. 18 19	Nd	Between 8 to 37	Palate	India (Karnataka)	
20	Female (Daughter)	3	Palate	China	
21	Female (Mother)	46	Palate	China	
22	Male	21	Along hard palate palate midline	Brazil	White
23	Female	18	Hard palate,right labial mucosa,facial aspect of maxillary gingiva	Indian	
24	Male	22	Left side of hard palate	Indian	
25	Male	21	Hard palate	Iran	
26	Female	23	Left buccal mucosa	India (Manglore)	
27	Female	36	Hard palate right side	India (Mysore)	
28	Female	Not Documented	Hard palate	Yemen	
29	Female	27	Palate and buccal mucosa	Argentina	
30	Male	30	Hard palate bilaterally	India (Udaipur,Rajasthan)	
31	Female	25	Left buccal mucosa	India (Moradabad,UP)	
32	Female	26	Midline hard palate	India(AP)	
33	Male	22	Hard palate left side	India(Kerela)	
34	Male	24	Hard palate right side	India (WB)	
35	Female	42	Hard palate	Japan	
36	Male	56	Soft palate left side	India (Jodhpur,Rajasthan)	
37	Male	34	Hard palate	India (New delhi)	
38	Female	20	Hard palate right side, Maxilarry marginal and attached gingiva and right lateral border tongue	India (Gujrat)	
39	Male	12	Left buccal mucosa	USA	White
40	Female	20	Left buccal mucosa	China	
41	Male	52	Posterior hard palate right side	Bulgaria	White
42	Female	35	Hard palate right side	India	
43	Male	28	Hard palate left side	India	
44	Female	6	Hard palate right side	India (Banglore)	
45	Female	19	Hard palate right side	India (UP)	
46	Male	40	Posterior glandular zone of hard palate	Mexico	
47	Female	22	Hard palate both side	India (TN)	
48	Female	48	Hard palate left side	UK(Liverpool)	White
49	Female	30	Hard palate mid palatine region	India (Maharashtra)	
50	Male	12	Hard palate left side	India(AP)	
51	Female	36	Palate	India(Assam)	
52	Female	18	Hard palate right side	India(New Delhi)	
53	Female	22	Hard palate midline	India(New Delhi)	
54	Female	13	Hard palate , gingiva	India(New Delhi)	
55	Female	50	Hard palate right side	India (Indore,MP)	Present case

Ref. No.	Intraoral Article Published
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Conclusion:

Lack of familiarity with the nevus of Ota is obvious as it is a rare disorder. Dental or medical doctors might get confused by such patches of the face or palate with hemangiomas or other pigmented lesions and refer patients to undergo unnecessary ultrasound or angiography. While, in certain cases, unrecognition can also put patients at risk for melanoma and Opthalmic consequences.

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