Case Report

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Dentistry to Dermatology- A Rare Case of Segmental Odontomaxillary Dysplasia

Abstract:

Jawbones have various properties that result from their peculiar embryonic development. Jawbones play a pivotal role in anchoring teeth. Any disturbance in their development leads to the pathology of the jaw and their surrounding structures. Since these pathologies develop from the jaw they have typical clinical radiographic and histologic features. The case report highlights the importance of prompt diagnosis which will lead to proper treatment and counselling of the patient.

This is a case report of 12 years old female patient who came to the Department of Oral Medicine and Radiology with a chief complaint of caries. Later, on examination, the unusual swelling in the maxilla was noted with the patient being completely unaware and asymptomatic. On correlating various clinical radiographic and histologic features a rarest of rare concealed pathology of jaw bone was unshielded.

Keywords: Segmental Odontomaxillary Dysplasia, Hyperpigmentation, Hypertrichosis, Vertically oriented trabeculae, Becker's nevus

Introduction:

Segmental odontomaxillary dysplasia (SOD) initially was also known as hemimaxillofacial dysplasia is depicted as a peculiar radiographic appearance of altered trabeculae of bone with gingival hyperplasia, tooth anomalies and sometimes with facial asymmetry. This case is eccentric in the sense that the occurrence of segmental odontomaxillary dysplasia is rare but its association with dermatologic finding is even the rarest. Becker's Nevus is a hyperpigmented hyperthypertrichotic patch arising commonly on the skin of the upper trunk, chest, or shoulder of young men. It is often associated with various other anomalies but its occurrence with dental deformity is infrequent.[1] The subsequent case illustrates the multidisciplinary approach of various medical professionals which can be helpful in identifying such a unique disorder. This kind of approach is not only helpful for doctors in predicting the above disorder in future but also to the patient in making them aware of the condition at the earliest.

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Case Presentation:

A 12-year-old female came with the chief complaint of caries in the upper left back region of the jaw in the department of oral medicine and radiology. The patient did not reveal any significant medical and familial history. On intra-oral examination, there was slight gingival swelling seen on the

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left upper back region of the jaw. The patient was completely unaware and asymptomatic about the swelling.A localised elliptical-shaped swelling is seen on the second quadrant with respect to DE and 6 measuring approx. 4x2 cm in diameter extending anteriorly distal to 23 and posteriorly till distal aspect of [26]. Swelling extends medially 5 mm away from the palatal cusp of D to 3mm laterally to the buccal cusp of D. Superior- inferiorly swelling extends from marginal gingiva to the mucogingival junction. The swelling was firm to hard and non-tender and was following the gingival contour throughout. The growth had caused the buccal palatal alveolus expansion. Premolars were missing in the left upper quadrant as shown in Figure 1 A.

Extraoral examination revealed the hyperpigmented area with hypertrichosis in the left lower third of the face as shown in Figure 1 B. A provisional diagnosis of the benign odontogenic tumour was given. As a part of further investigation intraoral periapical radiograph was taken to assess the change in bone. An intraoral periapical radiograph showed missing permanent premolars i.e. 24, 25 with retained 65; root resorption is evident with65. Dense granular and vertically oriented trabeculae pattern can be appreciated.

So based on the radiographic findings the differential diagnosis of Fibrous Dysplasia was givenon seeing the altered trabeculae and dense granular pattern although fibrous dysplasia is not associated with congenital missing tooth or hyperpigmentation with hypertrichosis

Further to enhance our knowledge, various texts were referred and the case was discussed with staff. It was later concluded based on the text available that the lesion was segmental odontomaxillary dysplasia with Becker's Nevus. The path gnomic feature of the entity being missing premolars,unilateral maxillary involvement and vertically oriented trabeculae. Extra oral features associated with Becker's Nevus are hyperpigmentation with hypertrichosis. The case was consulted with dermatologist within the institution and therefore, the patient was advised biopsy of the skin for confirmation (Figure 3 A). Biopsy result favoured the clinical diagnosis of Becker's Nevus. Patient was kept on follow up and was asked to report back as soon as she experiences any discomfort about the intraoral swelling and skin pigmentation. The patient was advised Laser surgery for skin pigmentation. Laser surgery could not be done as the patient was financially unstable and therefore, skin lightening cream was advised and was kept on rigorous follow-up. The patient was also explained about the severity of lesion if left unmonitored. The patient was also enquired about the ipsilateral hypoplasia of breast which is often associated with Becker's Nevus. Fortunately, the patient was not having such intensive findings.

Treatment: Reassurance was provided regarding the benign nature of the condition. The patient was placed on periodic recall to monitor the growth and development of the maxillary bone and teeth. Patient was advised to undergo LASER for extraoral hyperpigmented area but due to financial crisis patient did not get it done. Hence, patient was advised skin lightening cream

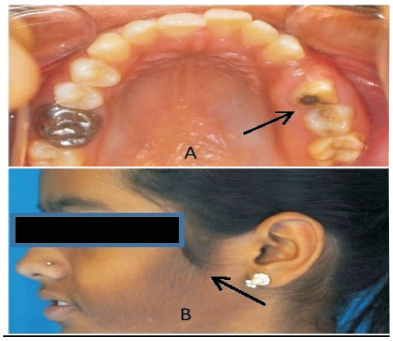

Figure1 A: Showing intraoral affected site
Figure1 B: Showing hyperpigmented area with hypertrichosis

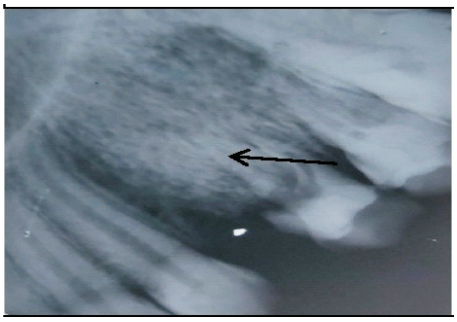

Figure2: Showing vertically oriented trabeculae

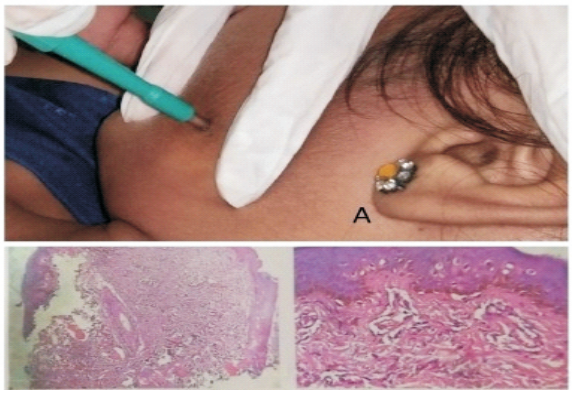

Figure 3 A: Site of biopsy
Figure 3 B: Confirmed biopsy report showing Becker's Nevus

Discussion:

The aetiology was SOD has been unknown. Clinically, patients with SOD have a non-progressive, unilateral buccal-lingual enlargement of the posterior maxilla with accompanying gingival enlargement, usually in early childhood. The average age of patients is 11.4 years. Dental abnormalities are noted, which include over-retained primary teeth, congenitally missing teeth, delayed eruption (frequently involving premolars), increased spacing between teeth, and altered tooth size.[2,3,4] The primary teeth (molars, in particular) may have abnormal coronal and radicular morphology. Often the teeth are smaller or larger than normal with elongated or splayed roots and enlarged or atrophic pulp chambers.4 It has been suggested that patients with SOD and hypertrichosis are always male. But in our case, it was female who was affected. Radiographically, ill-defined sclerosis or coarse bony trabeculation with vertical orientation is noted in the posterior maxilla, often with reduction of the maxillary sinuses.5The above case also highlightsthe importance of simple intraoral periapical radiograph in diagnosing such rare conditions .The above condition is infrequent and perhaps under-recognized condition affecting the maxilla and associated structures. Lack of extensive knowledge among dental and medical professionals assessing a patient with SOD may lead to incorrect or delayed diagnosis with unnecessary treatment and patient confusion.

Patient's perspective- Patients was completely asymptomatic about the condition initially and later on was counseled about the rarity and need of follow up. Although patient was cooperative, since the hyperpigmented area present extra orally was stagnant and was not increasing in size, patient refused to undergo LASER surgery and therefore was advised skin lightening cream.

Reference:

1. Miles DA, Lovas JL, Cohen MM. Hemimaxillofacial dysplasia: a newly recognized disorder of facial asymmetry, hypertrichosis of the facial skin, unilateral enlargement of the maxilla, and hypoplastic teeth in two patients. Journal of Oral Surgery, Oral Medicine and Oral Pathology 1987;64(4):445-8.
2. Agrawal A, Daniel J, Vijeev V. Segmental odontomaxillary dysplasia. Journal of Indian Society ofPedodontics and Preventive Dentistry 2014;32(1):83-6.
3. Azevedo RS, da-Silveira LJ, Moliterno LF, Miranda AM, de-Almeida OP, Pires FR. Segmental odontomaxillarydysplasia: report of a case emphasizing histopathological, immune histochemical and scanning electron microscopic features. Journal of Oral Science 2013;55(3):259-62.
4. Whitt JC, Rokos JW, Dunlap CL, Barker BF. Segmental odontomaxillary dysplasia: report of a series of 5 cases with long-term follow-up. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics2011;112(2):29-47.
5. Smith MH, Cohen DM, Katz J, Bhattacharyya I, Islam NM. Segmental odontomaxillary dysplasia: An underrecognized entity. The Journal of the American Dental Association 2018;149(2):153-62.