Intraoral Atypical Cellular Blue Nevus: A Rare Case Report and Review of Pathogenesis

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ABSTRACT

Nevi are the congenital or developmental malformations seen on the skin and mucosa which usually develop from the nevus cells of surface epithelium. Cutaneous melanocytic nevi are more common than their intraoral counterpart. According to the location of the nevus cells, the nevi are histologically divided into four types: intradermal, junctional, compound and blue nevi. Intraoral Blue nevus is a very rare entity and clinically it manifests as well-circumscribed, slightly raised, asymptomatic, blue-black lesions in palate and gingiva. Mutation of GNAQ gene, encoding the G-protein alpha subunit of cell-surface receptor results in development of Blue nevus. Though blue nevus is a benign neoplasm, it has a high malignant transformation potential. This case of intraoral blue nevus in a 39-year-old male is not only rare of its kind but also had features of cellular atypia which makes it more prone towards transformation into a malignant one.

Key words: Intraoral blue nevus; Atypical; Pathogenesis

Introduction

Nevi are the congenital or developmental malformations seen on the skin as well as mucosa and usually develop from the nevus cells of surface epithelium. Cutaneous melanocytic nevi are more common than their intraoral counterpart. According to the location of the nevus cells in the epithelium and underlying connective tissue, the nevi are histologically divided into four types - intradermal, junctional, compound and blue nevi. Clinically, the intraoral blue nevus manifests as well-circumscribed, slightly raised, asymptomatic, blue-black lesions in palate and gingival. Due to the phenomenon of Tyndall effect, the melanin pigment lying deep within the stroma absorb the longer wavelength light like red, yellow and reflects only the smaller wavelength light i.e. blue to the investigator’s eyes thereby imparting the blue colour to the lesion. Blue nevus has three subtypes viz., common, combined, and cellular. Histopathologically, the blue nevus is composed of pigmented dendritic melanocytic cells arranged in the reticular stroma. Though blue nevus is a benign neoplasm, it has a high malignant transformation potential.

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Case Report

A 39-year old Indian male reported to the outpatient department of a tertiary health care centre with a chief complain of a bluish-black coloured painless swelling in gingiva of left side of the lower jaw. The patient stated that the swelling first appeared about four years ago and slowly increasing in size. His family history was not significant. Patient was a tobacco smoker since last twenty years approximately.

General survey of the patient revealed no such abnormality with all the vital signs within normal limits. Extra oral examination was nil of note. Intra oral examination revealed multiple attrited teeth with extrinsic stains. Examination of the lesion proper revealed a bluish-black 1.2cm X 1.3cm swelling in lingual gingiva in respect to tooth number 35 (Figure 1). On palpation, the lesion was soft to firm in consistency, non-tender and never bleeds on provocation. The borders were not indurated and blanching was absent as noticed in diascopy.

After complete haematological and radiological examination, when the patient was found to be fit for minor surgical procedure, an incisional biopsy was performed (after getting informed consent from the patient) for histopathological confirmation.

Histopathological examination (Figure 2 and 3) revealed the presence of connective tissue covered on the surface by stratified squamous epithelium. The sub epithelial region showed a tumour composed of nests, cords and trabeculae of plump, melanin-producing spindle cells having vesicular nuclei, distinct nucleoli and eosinophilic to clear cytoplasm. At focal areas, cellular atypia was noted. No pagetoid spread on the overlying surface epithelium was seen. Immunohistochemistry was performed and the tumour cells were positive for S100 (Figure-4), HMB-45 (Figure 5) and Melan-A (Figure 6). These microscopic findings were suggestive of atypical cellular blue nevus.

Discussion

Intraoral blue nevus was first reported by Scofield [7]. The cellular blue nevus is very much less common and generally manifests during the second to fourth decades of life. This cellular variant measures 1 to 3
cm in diameter and is usually larger than the common variant. Histopathologically, the cellular blue nevus manifests as a well-defined, highly cellular consisting of aggregate of plump, spindle shaped melanin producing cells within the lamina propria.

Regarding the pathogenesis of blue nevus, various theories have been proposed till date. Among them, the most accepted one is that this blue nevus develop from those latent dendritic melanocytes residing in the stroma during their embryologic migration from the neural crest to the epithelium. During the foetal period, the melanocytes first migrate in the head-neck region at around 70 to 80 days of gestation followed by the entire body. At the end of gestational period, most of the stromal melanocytes disappear. Only few of such melanocytes persist within the stroma of head-neck region and some other areas like presacral area, distal extremities. These melanocytes undergo proliferation and results in dermal melanocytoses.

Some authors have proposed a second theory regarding pathogenesis of blue nevus which states that the mutation of precursor stem cell of the stromal melanocytes results in development of blue nevus. This theory was not widely accepted because CD34, a stem cell marker, is poorly expressed in some blue nevi.
Studies on the molecular pathogenesis of blue nevi explained that the mutations in GNAQ and GNA11 leads to proliferation of dermal melanoblast. In 83% of blue nevi, somatic mutation of GNAQ gene have been identified. Such mutations were noted in the ras-like domain of the protein, there by leading to formation of active GNAQ protein and development of blue nevus.

In studies with transgenic mice, it has been found that there is increased expression of HGF (Hepatocyte Growth Factor) or Hras1 which ultimately leads to proliferation of dermal melanocytes and development of blue nevus.

Conclusion

The differential diagnosis of blue nevi includes amalgam tattoo, vascular malformation and melanoma. In the above said case, there was no amalgam restoration in the vicinity of the lesion. Neither the lesion showed blanching on diascopy. And melanoma usually has a site predilection for maxilla. Hence, the diagnosis of blue nevus was considered as a provisional diagnosis which was confirmed later by histopathological study. However, any atypical blue cellular nevus must be excised and to be followed up regularly as they have a chance of malignant transformation.

References


