



MULTIPLE LEIOMYOMATOUS HAMARTOMAS OF HARD PALATE WITH MIDLINE CLEFT –A CASE REPORT

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ABSTRACT

Hamartomas of the head and neck are described as rarely seen benign neoplasms. They may occur as focal malformations. Hamartomas seen in the oral cavity are rarer and review of literature shows only few cases reported mainly of Japanese and Latin American origins. Whereas those on the palate are still rarer and when present they may often be associated with a cleft palate and have to be differentiated from other tumors.

Our aim is to report an unusual and unique case of Multiple Leiomyomatous Hamartomas of the hard palate associated with a midline cleft palate in a five month old female child. Such tumors should include a detailed evaluation by the surgeons as they may be complicated by the presence of neural tube defects or other associated anomalies. The treatment of oral and palatal hamartomas is surgical, which is often curative without any recurrence.

Key-words: Hamartoma, leiomyomatous, cleft palate.

INTRODUCTION

Hamartomas of the head and neck, especially oral cavity are rare, benign neoplasms or focal malformations. They are composed of tissue elements normally found at the site of their location but grow in a disorganized manner. They are asymptomatic and remain undetected for a long time. Review of literature shows only few cases reported mainly of Japanese and Latin American origins.^[1] These cases showed a wide range in their age of presentation ranging from infancy to the fifth decade. Moreover the hamartomas reported till date presented as a single polypoid, sessile or pedunculated masses.

Such tumors in oral cavity are mostly seen along the tongue.^[2]

Hamartomas on the palate are still rarer and when present may be often associated with a cleft palate and have to be differentiated from other tumors. The present case report consists of an unusual and rare case of Multiple Leiomyomatous Hamartomas of the hard palate associated with a midline cleft palate in a five month old female child. The occurrence, incidence, clinical evaluation, surgical aspects, treatment, histological appearance and possible embryological basis are discussed in the current case report.

CASE HISTORY

A five month old female child was brought with complaints of excessive salivation and difficulty in feeding since birth. The mother also noticed a progressively increasing swelling over the hard palate since birth. On examination of the oral cavity a multilobulated, soft pinkish white mass was seen occupying the whole of hard palate. The mass was obstructing the oral cavity and compressing the tongue, causing difficulty in swallowing. It was associated with a midline cleft palate with no other systemic congenital anomaly.

In the first stage the palatal masses were excised. On gross examination they showed multiple masses together measuring about 5x4x4 centimeters, which were five in number, of which three were pedunculated and two sessile. Largest single mass measured 2.5 centimeters in diameter. Sections were studied for histological diagnosis. The diagnosis of multiple leiomyomatous hamartomas was confirmed on Haematoxylin – Eosin stained slides (Figures 1 and 2). Histologically they showed well circumscribed

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masses covered with stratified squamous keratinizing epithelium with skin adnexae, mainly abundant hair follicles. The core showed dilated blood vessels, smooth muscle bundles, adipose and connective tissue.

DISCUSSION

Hamartomas occur in multiple sites and different organs and are mainly composed of foci or nodules of tissues normally occurring at that site. They are defined as malformations of tissues indigenous to the area resembling and having some features of tumors.^[3] The occurrence of such congenital hamartomas may be as a result of mishap during embryogenesis. They probably have foci in the form of various developmental rests which are vestiges of cell migration and differentiation during embryogenesis. Rarely tumors arise from them. Hamartomas of the oral cavity is a rare tumor. They mostly occur along the tongue. A review of literature by Nava-Villalba et al in 2008 put forth the occurrence of only sixteen cases of leiomyomatous hamartomas of oral cavity in English literature and eight others in Japanese literature.^[1] The age of presentations in their study ranged from infancy to second decade. Most of them occurred on the tongue and only one on the hard palate. All the tumors presented as a single, polypoid, pedunculated or sessile masses less than two centimeters in maximum diameter.

Napier et al in 1996 reported a case of Unusual leiomyomatous hamartoma of the hard palate in a normal five year old girl,^[4] De Biase A et al in 2002,^[5] Zaitoun H et al in 2007,^[6] and Rameshwar Bang et al^[7] in 2010 reported similar hamartomatous lesions on the hard palate. All the cases reported showed occurrence of a single mass on the palate.

The present case also had hamartomatous lesion on the palate but they were unique because of their size and were multiple pedunculated and sessile. The lesion was also associated with a midline complete cleft palate.

Takeyama et al^[8] described a case of hamartoma on hard palate associated with corpus callosum agenesis, microphthalmia and skin malformations. The present case did not have any neural defects and is growing well after a follow up of two years.

The histology of a leiomyomatous hamartoma is very peculiar, with a well delimited subepithelial proliferation of a combination of tissues including smooth muscle fascicles scattered irregularly, blood vessels, lymphatics, adipose tissue and occasionally salivary glands with a fibrous connective tissue stroma and mature fibroblasts.

They can be named differently depending on the predominant component as vascular, neurovascular, lymphatic, fatty, glial, rhabdomyomatous or leiomyomatous hamartomas. Such lesions have to be differentiated from other intraoral lesions like congenital epulis, choristoma of tongue, fibroepithelial polyps, teratoma and others.^[9] The histology is obvious and diagnostic. In some difficult cases immunohistochemistry for smooth muscle may be helpful.^[10]

The treatment of oral and palatal hamartomas is surgical which is curative without any recurrence. The cases associated with cleft palate can undergo definitive palatoplasty at a later age probably after a follow up of nine to twelve months.

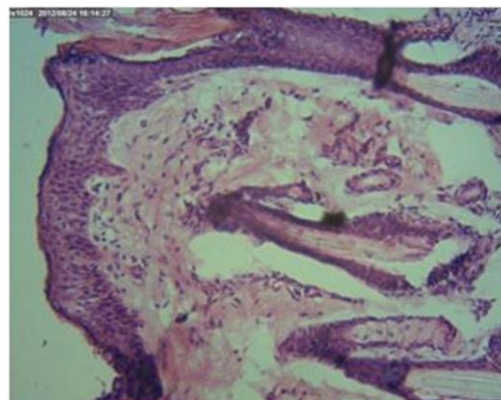


Figure 1: Haematoxylin and Eosin stained slide of multiple leiomyomatous hamartomas of palate showing covering skin with adnexae.

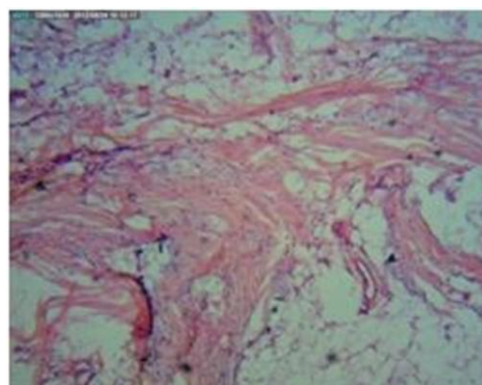


Figure 2: Haematoxylin and Eosin stained slide of multiple leiomyomatous hamartomas of palate showing core with smooth muscle bundles and adipose tissue.

CONCLUSION

A rare and probably unique case of Multiple Leiomyomatous Hamartomas of hard palate associated with midline complete cleft palate in

infancy is reported. Such tumors on the hard palate should warrant the attention of the surgeon for detailed evaluation as they may be complicated by presence of neural defects or other associated anomalies. Other clinical as well as histological differential diagnoses should be ruled out. A cautious surgical plan with proper follow up will definitely give best results.

REFERENCES

- 1] Nava-Villalba M, Ocampo-Acosta F, Seamandura-Pacheco A, Aldape – Barrios BC: Leiomyomatous hamartoma : report of two cases and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008,105: 39.
- 2] Miyamoto Y, Nagayama M, Hayashi Y. A cleft palate child with lobulated tongue and lingual hamartoma: Report of a case. *J Oral Maxillofac Surg.* 1991;49:644.
- 3] Harada H, Morimatsu M, Kusakawa J, Kameyama T. A hamartoma-like mass on the palate?.A possible discussion regarding the components of a pigmented naevus and hyperplastic salivary gland. *J Laryngol Otol.* 1997;111:296.
- 4] Napier SS, Devine JC, Rennie JS, Lamey P-J. Unusual leiomyomatous hamartoma of the hard palate. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996;82(3):305.
- 5] De Biase A, Cicconetti A, Agrillo A, Matteini C, Della Rocca C. Unusual vascular hamartoma of the hard palate. A case report. *Minerva Stomatologica.* 2002;54:157.
- 6] Zaitoun H, Triantfyllou A. Smooth muscle hamartoma of the hard palate. *J Oral Pathol Med* 2007;36(4):245.
- 7] Bang RL, Burezq H, Al-Najjadah I. Vomerine hamartoma in a cleft palate child. *Indian J Plast Surg* 2010;43:216.
- 8] Takeyama J, Miyota H, Nagasaka H. Hamartoma on the hard palate associated with corpus callosum agenesis, microphthalmia, and skin malformation. *J Oral Maxillofac Surg* 2007;65(3):553.
- 9] Liang CC, Lai JP, Lui CC. Cleft palate with congenital midline teratoma. *Ann Plast Surg.* 2003;50:550.
- 10] Baden E, Doyle JL, Lederman DA. Leiomyoma of the oral cavity: a light microscopic and immunohistochemical study with review of the literature from 1884 to 1992. *Eur J Cancer B Oral Oncol* 1994;30B (1):1.