International Journal of Oral & Maxillofacial Pathology. 2013;4(2):53-57
Available online at http://www.journalgateway.com or www.ijomp.org
ISSN 2231 – 2250

Case Report

An Asymptomatic Palatal Location is Exclusive for Polymorphous Low Grade Adenocarcinoma: A Case Report and Review

Harishchandra Rai, Shaila M, Anitha Dayakar, Lubna Nazneen

Abstract

Polymorphous low grade adenocarcinoma is considered to be a low grade malignancy with a relatively indolent course and a low risk of metastasis and it is found almost exclusively in minor salivary glands particularly on the palate with the history of long duration. We are reporting a case of Polymorphous low grade adenocarcinoma arising on right side of palate in a 47 years old female patient with the duration of ten years. Histopathologically the tumour cells were arranged in Lobular and concentric masses of cells surrounded by thin band of fibrous tissues with peripheral infiltration and connective tissue in few areas was mucinous and hyalinized. Based on the clinical and histopathological findings, diagnosis of polymorphous low grade adenocarcinoma was rendered.

Keywords: Adenocarcinoma; Glandular and Epithelia; Low Grade; Metastasis; Neoplasms, Palate; Salivary Glands.

Harishchandra Rai, Shaila M, Anitha Dayakar, Lubna Nazneen. An Asymptomatic Palatal Location is Exclusive for Polymorphous Low Grade Adenocarcinoma: A Case Report and Review. International Journal of Oral & Maxillofacial Pathology; 2013:4(2):53-57.

Received on: 11/03/2013 Accepted on: 21/07/2013

Introduction

Polymorphous low-grade adenocarcinoma (PLGA) is a recently recognized salivary gland malignancy that was first described by Freedman in 1983 under the name of 'lobular carcinoma', Evans and Batsakis in 1984, eventually coined the term 'Polymorphous low-grade adenocarcinoma.[1] This tumour is considered to be a low grade malignancy with a relatively indolent course and a low risk of metastasis and is second in frequency to mucoepidermoid carcinoma and accounts for 26% of all salivary gland carcinomas. It is thought to originate from the progenitor cells of the distal/terminal duct portions of the salivary gland unit i.e. the intercalated duct reserve cell.[2]

Polymorphous low grade adenocarcinoma is found almost exclusively in minor salivary glands, and is rare in extra oral locations, including major salivary glands with approximately 60% of the cases located in the hard and soft palate followed by 13% of the cases occurring in the buccal mucosa, 10% in the upper lip, 6% in the retro molar area, and 9% in the rest of the oral cavity.[3] It has an approximate female/male ratio of 2:1, it is seen predominantly in patients who are between 30 and 70 years old[4] and rarely occur in patients under the age of 20 years.[5] Here we are presenting a case of PLGA to consider its characteristic features of palatal location with history of long duration.

Case report

A 47 years old female patient reported to our institution with a complaint of swelling in the right side of palate since ten years. History revealed that the onset of these swellings was slow and progressive and the size of swelling was constant since last two to three years. Patient developed a dull aching pain since two to three days, which was continuous and radiating to right side of the face. Intraoral examination revealed a solitary sessile swelling on the palate extending from first maxillary premolar to second maxillary molar on the right side measuring about 4 x 4cm. Mucosa over the swelling was normal in colour. The swelling was firm in consistency, sessile, non- tender, non-fluctuant and non-reducible (Fig 1a). The C.T scan report revealed a well defined cystic lesion (13HU) in the hard palate on right side, eroding adjacent maxilla and causing rare fraction of inferior wall of right maxillary antrum and there was evidence of bulging of lesion into the right nasal cavity. Surgical excision of the lesion with hemi maxillectomy was done and obturator was given to close the surgical defect. There is no recurrence since one year.

Histopathological reports showed lobular and concentric masses of cells surrounded by thin band of fibrous tissues (Fig 1b). Connective tissue in few areas was mucinous and hyalinized (Fig 1c). Cribriform

pattern and duct like structures were also evident in few areas and peripheral infiltration of tumor cells in strands or trabecular pattern also was observed (Fig 1d). The tumour cells were round to polygonal in shape with indistinct cell borders and the cytoplasm was scanty with round, oval or spindle shaped nucleus. A thick band of connective tissue was seperating these areas from overlying epithelium. Based on the clinical and histopathological findings, diagnosis of polymorphous low grade adenocarcinoma was rendered.

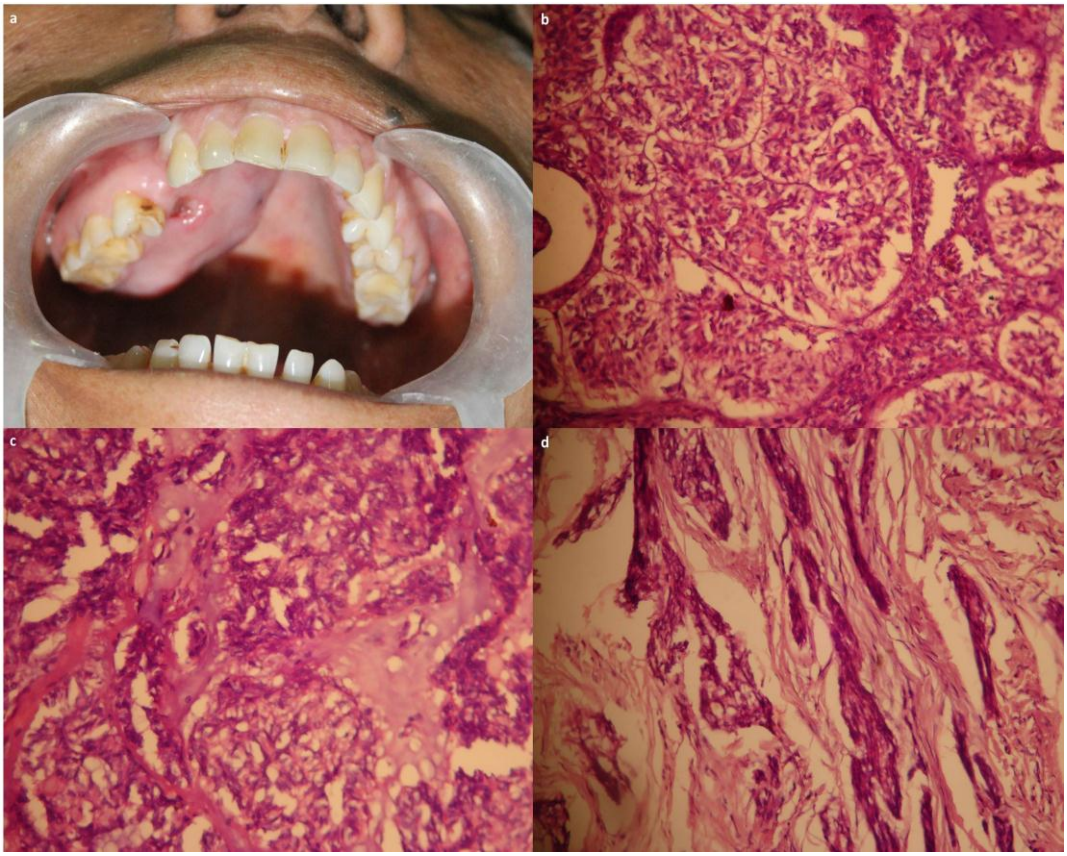

Figure 1: The intraoral photograph showing the lesion on the right postero-lateral part of the hard palate (a). The hematoxylin and eosin stained photomicrograph showing lobular and concentric masses of cells surrounded by thin band of fibrous tissues (b), hyalinized or mucinous connective tissue (c) and peripheral infiltration of tumor cells in strands or trabecular pattern (d).

Discussion

Polymorphous low grade adenocarcinoma is a rare lesion and the clinical behaviour of this lesion differs from other malignant salivary gland neoplasm as it is characterized by slow rate of growth, absence of symptoms, minimal aggressiveness, low metastatic potential and favourable prognosis. Although locally invasive, pain and ulceration are not frequent features but may be the consequence of trauma or pressure from oral prosthesis.[6] PLGA accounts for 7–11% of all neoplasms occurring in minor salivary glands.[7] The palate is the most frequent location, but it has been described also in other locations, such as the lip, tongue, buccal mucosa, floor of the mouth, maxillary sinus, pharynx, retromolar region, parotid, submandibular gland and sublingual gland.[8-15] High density of glandular tissues in the palate and in particular the junction of hard and soft palates renders this location the most frequent site for minor salivary gland neoplasms.[6]

In the largest study of 164 PLGA cases from armed force institute of pathology (AFIP) files, all were in minor salivary glands with palate being the commonest site.[16] In a large study on 80 cases, 11.3% cases were diagnosed as PLGA with palate as commonest site, 27.9% had experienced symptoms for longer than one year and 13.1% were asymptomatic.[17] Some of the cases showing similar features like our case are summarised in Table 1.

The appearance of PLGA in major salivary glands is extremely rare, and it has only

been referred in few reports, with similar clinical features as PLGA of minor salivary glands. The frequency of regional lymph node metastasis is 6–10%.[7,21] Metastases to the paraesophageal lymph nodes, lungs, orbit and skin have been reported, but overall distant metastases develop in less than one percent of cases.[5] It is often the result of inadequate local control of disease with the lung being the most frequently involved site.[6] A rare intraosseous case also have been described.[22]

Author	Location of the lesion	Duration
Gupta S et al (2011)	Palate	8 years
Kulkarni et al (2011)	Palate	2 years
Sunil et al (2010)	Hard palate extending to retromolar region	5 years
Pintor et al(2007)	Palate	8 years
Kumar et al (2004)	Junction of hard and soft palate	2 years
Crean et al (1996)	Palate	3 years
	Retromolar region	7 years

Table 1: A reported cases of polymorphous low grade adenocarcinoma of palate

Macroscopically, they are firm to solid ovoid masses, typically lying in close proximity to the overlying surface epithelium, and they are characteristically unencapsulated, although well circumscribed. Surface ulceration is not common. The cut surface is light-yellow to tan[16], without a specific gross appearance.[23] The histology of PLGA comprises its infiltrative growth patterns, varied cytologic features, and the presence of matrix material. They are seen to infiltrate into perisalivary gland adipose connective tissue, but true skeletal muscle invasion is uncommon. When present, skeletal muscle involvement usually presents as a compression of the muscle fibers. Infiltration into the adjacent salivary gland is quite common, however. The surface epithelium is usually intact, but on occasion is ulcerated, when intact, the surface epithelium is usually uninvolved by tumour.[23]

PLGA may display a striking mixture of growth patterns within a single tumour, including solid lobules, glandular profiles, tubules, trabeculae, cribriform nests, a linear, single cell "Indian-file" infiltration.[2,6,20,] Tumour cells are often arranged concentrically around a central nidus, creating a "targetoid" appearance. The nidus is often a small nerve bundle (neurotropism), and is quite characteristic for PLGA.[6,19,23] Papillary foci can be rarely seen, but when present represent only a minor component and are not the dominant pattern.[23] Tumour consists of cuboidal/columnar isomorphic cells with spindle shaped nuclei. Nucleoli appear small and inconspicuous and the chromatin varies from vesicular to stippled. The tumour stroma varies from mucoid to hyaline[2,19] and perineural invasion is common.[2] Metaplastic changes, including squamous, sebaceous, or oncocytic alteration, are only rarely identified.[19,23]

Separation of PLGA from pleomorphic adenoma and adenoid cystic carcinoma are the main diagnostic dilemmas. Clinical features of pleomorphic adenoma affecting minor salivary glands overlap with those of PLGA and include absence of symptoms, slow growth, firm consistency, smooth texture as well as palatal predilection. Pleomorphic adenoma is usually circumscribed and variably encapsulated.[6] In the minor glands, however, it can be nonencapsulated and mimic an invasive neoplasm. The recognition of typical chondromyxoid matrix and the presence of hyaline, plasmacytoid cells are sufficient to secure a diagnosis of pleomorphic adenoma.[23]

Adenoid Cystic Carcinoma (ACC) resembles PLGA in age, gender and palatal predilection, perineural invasion as well as slow rate of growth, variability of growth patterns and infiltrative borders. In contrast, low-grade, dull pain is a frequent complaint with ACC and palatal tumours may appear ulcerated. Both tumours may recur locally; however, ACC is more aggressive with a higher proliferative index, carries a greater potential for distant versus regional metastasis and has a worse prognosis.[6] The cytoplasmic staining of ACC is pale to clear staining unlike in PLGA which is eosinophilic to amphophilic, the nuclei are more hyper chromatic and more angular. There is the accumulation of basophilic pools of glycosaminoglycans in the cribriform areas, which is not typical of PLGA.[2,24] A

preponderance of cribriform elements and the presence of a tubular pattern with double-layered ducts composed of small angular hyperchromatic cells, however, are characteristic of adenoid cystic carcinoma.[6,24] There have been several reports recently that C-KIT (CD117) is expressed in adenoid cystic carcinoma, but not in PLGA.

Application of immunohisto-chemistry in the diagnosis of PLGA and its differentiation from other salivary gland tumours has been extensively examined. Studies have shown expression of markers such as glial fibrillary acid protein (GFAP) in PA; vimentin, S-100; high and low molecular weight cytokeratins and epithelial membrane antigen (EMA) in PLGA, and Ki67, c-kit (CD117), alpha smooth muscle actin and carcinoembryonic antigen (CEA) in ACC.[6]

The primary management of PLGA is surgical. Wide local excision with good margins is recommended to achieve local control. This can be performed with little morbidity for small intraoral tumours. Larger tumours may need reconstruction as appropriate. Unless a major resection with reconstruction is planned, an elective neck dissection is not warranted. The overall prognosis is good. Perineural invasion does not appear to affect the prognosis.[2]

Conclusion

Polymorphous low grade adenocarcinoma is a rare malignant salivary gland tumor, with a clinical behaviour similar to that of a benign neoplasm with low symptomatology and long duration. Due to the benign nature of the tumor,most of the time PLGA will be considered as one of the differential diagnosis for benign salivary gland tumors.

Acknowledgement

We would like to acknowledge all the staff members in the Department of Oral and maxillofacial Pathology for their support and guidance.

Author Affiliations

1.Dr.Harish Chandra Rai, Professor and Head, 2.Dr.Shaila M, Reader, 3.Dr.Anitha Dayakar, Reader, 4.Dr.Lubna Nazneen, Post Graduate Student, Department of Oral and Maxillofacial Pathology, KVG Dental College and Hospital, Rajiv Gandhi University of Health Sciences , Sullia, Dakshin Karnataka, India.

References

1. Singh R, Sheikh S, Pallagatti S, Aggarwal A, Singh B, Puri N. Polymorphous low grade adenocarcinoma- A rare case report. Int J Cont Dent 2010;1(3):89-92.
2. Sunil S, Sreenivasan BS, Titus J, et al. Polymorphous low grade adenocarcinoma- case report and review of literature. Oral Maxillofac Pathol J 2010;1(2):77-82.
3. Pintor MF, Figueroa L, Martinez B. Polymorphous low grade adenocarcinoma: review and case report. Med Oral Pathol Oral Cir Bucal 2007;12(8):549-51.
4. Clayton JR, Pogrel MA, Regezi JA. Simultaneous multifocal polymorphous low grade adenocarcinoma- report of two cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1995;80:71-7.
5. Kumar M, Stivaros N, Barrett AW, et al. Polymorphous low grade adenocarcinoma-a rare and aggressive entity in adolescence. Br J Oral Maxillofac Surg 2004;42:195-9.
6. Fatahzadeh M. Polymorphous low grade adenocarcinoma: case report and review of diagnostic challenges. Arch Orofacial Sci 2012;7(2):92-100.
7. Anwar MM, Khan S. Polymorphous low grade adenocarcinoma in oral cavity: a case report. Bangaladesh J Otorhinolaryngol 2010;16(1):66-9.
8. Gupta R, Gupta K, Gupta R. Polymorphous low grade adenocarcinoma of the tongue: a case report. J Med Case Repor 2009;3:9313-5.
9. Etİt D, Altınel D, Bayol U, et al. Polymorphous low grade adenocarcinoma located in the maxillary sinus. Turk Patoloji Derg 2012;28:274-7.
10. Kaul R, Gulati G, Kaushik G, Raina S. Polymorphous low grade adenocarcinoma- an unusual presentation. Online J Health Alli Sci 2010;9(2):18-19.
11. Takubo K, Doi R, Kidani K, et al. Polymorphous low grade adenocarcinoma arising at the retromolar region: A rare case of high grade malignancy. Yonago Acta medica 2007;50:17-22.
12. Arathi N, Bage AM. Polymorphous low grade adenocarcinoma of parotid gland: A rare occurrence. Indian J Pathol Microbiol 2009;52(1):103-5.
13. Godoy LR, Suarez L, Mosqueda A, Meneses A. Polymorphous low grade adenocarcinoma of the parotid gland: case report and review of the literature.

Med Oral Patol Oral Cir Bucal 2007;12:30-3.
14. Rathod V, Rathod C. Polymorphous low grade adenocarcinoma of submandibular salivary gland tumour- A case report. J Indian Dent Assoc 2011;5(1):41-2.
15. Potluri A, Prasad J, Levine S, Bastaki JM. Polymorphous low grade adenocarcinoma: a case report. Dentomaxillofac Radiol 2013;42(2). doi:10.1259/dm fr/14804843.
16. Castle JT, Thompson LDR, Frommelt RA, Wenig BM, Kessler HP. Polymorphous low grade adenocarcinoma: A clinic-pathologic study of 164 cases. Cancer 1999;86(2):207-19.
17. Jansisyanont P, Blanchaert Jr RH, Ord RA. Intraoral minor salivary gland neoplasm: a single institution experience of 80 cases. Int J Oral Maxillofac Surg 2002;31:257–61.
18. Gupta S, Kumar CA, Raghav N. Polymorphous low grade adenocarcinoma of the palate: Report of a case and review of literature. Int J Head Neck Surg 2011;2(1):57-60.
19. Kulkarni M, Gabhane M, Mahajan A. Polymorphous low grade adenocarcinoma: A case report. Indian J Dent Sci 2011;3(2):23-5.
20. Crean SJ, Bryant C, Bennett J, Harris M. Four cases of polymorphous low grade adenocarcinoma. Int J Oral Maxillofac Surg 1996;25:40-4.
21. Pinto PX, Coleman N. Regional metastasis in polymorphous low grade adenocarcinoma-Report of a case. Int J Oral Maxillofac Surg 1997;26:447-9.
22. Sato T, Indo H, Takasaki T, et al. A rare case of intraosseous polymorphous low grade adenocarcinoma (PLGA) of the maxilla. Dentomaxillofac Radiol 2001;30:184-7.
23. Lester D, Thompson R. Polymorphous low grade adenocarcinoma. Pathology Case Reviews 2004;9(6):259-63.
24. Paleri V, Robinson M, Bradley P. Polymorphous low grade adenocarcinoma of the head and neck. Curr Opin Otolaryngol Head Neck Surg 2008;16:163-9.

Corresponding Author
Dr. Harishchandra Rai,
Professor and Head,
Department of Oral & Maxillofacial Pathology,
K.V.G Dental College and Hospital,
Sullia, Dakshin Kannada, Karnataka, India.
Ph: 09448254745
E-Mail: hrishrai@gmail.com

Source of Support: Nil, Conflict of Interest: None Declared.