Case Report

Rare case of giant myoepithelioma in minor salivary glands of palate in a 9-year-old child

ABSTRACT

Myoepitheliomas (MEs) are extremely rare benign neoplasms composed of ectodermally derived contractile smooth muscle cells (myoepithelial cells). Various tissues such as the salivary glands, breast, larynx, and sweat glands show the presence of these myoepithelial cells. They occur, principally, in the parotid gland and infrequently in minor salivary glands. The term "Myoepitheliomas" was first coined by Sheldon in 1943.It is an uncommon salivary gland tumor which accounts for < 1% of all major and minor salivary gland tumors. Batasakis considers the ME to be "one-sided" variant at the opposite end of the spectrum from the pleomorphic adenoma. There are distinct histological and immunohistochemical characteristics of the tumor which aid in the diagnosis. ME of the palate is uncommon, and only a limited number of cases have been reported in the English literature. It shows a benign clinical course with recurrence in up to 20% of cases without metastasis.The present article sheds light on the presence of ME of minor salivary glands in the palate of 9-year-old child.

Keywords: Myoepithelioma, plasmacytoid, salivary gland tumor

INTRODUCTION

The term "myoepithelioma" was first introduced by Sheldon in 1943.[1] Myoepithelioma (ME) is defined as a benign, solid tumor composed predominantly or entirely of neoplastic cells of myoepithelial differentiation.[2] Normally, myoepithelial cells are essential components of some exocrine glands such as salivary glands, lacrimal glands, sweat glands, and mammary glands.[3] MEs represent 1%–1.5% of all salivary gland neoplasms.[3,4] Most commonly occurs in the parotid glands (40% of all cases) and less frequently in the submandibular gland and the minor salivary glands. MEs of the soft or hard palate account for 21% of all cases and for 93% of intraoral cases.[3,5] In the international literature, the tumor is reported to appear at any age between 8 and 85 years with an average age of 40 years and a peak in the third decade.[4,6] The aim of this article is to describe a case of ME of the palate, focusing on the clinical, histological along with review of the literature.

CASE REPORT

A 9-year-old female child presented with an asymptomatic slowly progressive palatal mass for 1 year (usually noted in the superficial lobe of parotid in the mid-adult years). There was no history of dysphagia, odynophagia, sleep apnea, voice change, weight loss, loss of appetite, and fever. The clinical examination revealed a firm, nontender, nonpulsatile, round, bluish pink mass originating from the

Swapna Patankar, Reshmi Sharma[1], Amod Patankar[1], Venkatesh Kulkarni

Departments of Oral and Maxillofacial Pathology and Oral Microbiology and [1]Oral and Maxillofacial Surgery, Bharati Vidyapeeth Dental College and Hospital, Pune, Maharashtra, India

Address for correspondence: Dr. Swapna Patankar, Bharati Vidyapeeth Dental College and Hospital, Katraj, Pune - 411 046, Maharashtra, India.
E-mail: reshmisharma64@gmail.com

Received: 09 March 2019, **Revised:** 24 May 2019, **Accepted:** 23 March 2020, **Published:** 15 July 2021

Access this article online	
Website: www.njms.in	Quick Response Code
DOI: 10.4103/njms.NJMS_16_19	

How to cite this article: Patankar S, Sharma R, Patankar A, Kulkarni V. Rare case of giant myoepithelioma in minor salivary glands of palate in a 9-year-old child. Natl J Maxillofac Surg 2021;12:284-8.

left side of hard palate crossing midline (the lesion was firm in our case, whereas it is usually freely movable if present at any other site in the oral cavity). No ulceration was noted on the surface of swelling Figure 1. The computed tomography (CT) scan showed well-defined heterogeneous mildly enhancing soft-tissue mass arising from hard palate (measuring 4.4 cm × 3.4 cm × 2.2 cm), causing significant bulge in oral cavity with no obvious involvement of soft palate and no bony erosion. There was slight calcification in the center of mass, but no cystic component or fat tissue component within the mass. Based on CT findings, it was diagnosed with a benign palatal mass. The surgical excision of the benign palatal tumor was planned out. Preoperative planning involved replication of tumor in cast [Figure 2]; impression made using medium-body putty material. Tumor impression replicated in the cast was trimmed, following which obturator was fabricated [Figure 3]. The patient was operated through transoral approach under general anesthesia. The mass was totally removed [Figures 4 and 5] with submucosal dissection and histological diagnosis was confirmed. Immediately, after the removal of palatal tumor, the obturator was given for 2 weeks [Figure 6]. The obturator was removed after 15 days of the surgical procedure. The patient was recalled after every 15 days for the next 2 months, to assess the presence of any secondary infectioninfection; Figure 7 is showing the progress after 3 weeks. The patient was kept under observation for the next 6 months.

Histologic features

The tumor is composed of exclusively of neoplastic myoepithelial cells. The tumor shows lobules and sheets of proliferating tumor cells, which are predominantly plasmacytoid [Figure 8]. Tumor cells are round cells with eccentric nuclei with eosinophilic often hyaline appearing cytoplasm giving it a plasmacytoid appearance [Figure 9] and these cells are often referred to as "Hyaline Cells." Little intercellular fibrous stroma is present.

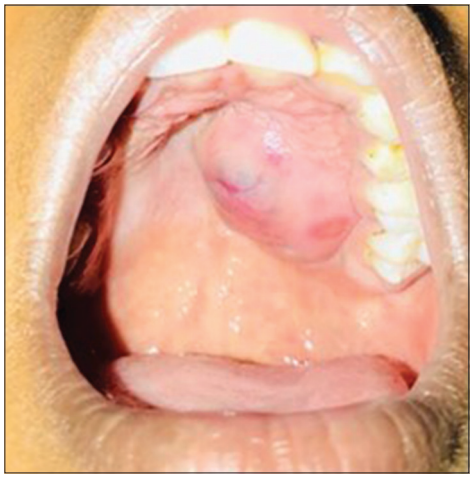

Figure 1: Palatal swelling crossing midline

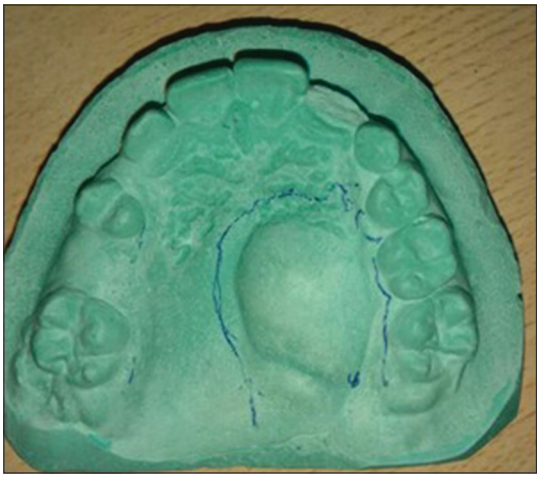

Figure 2: Tumor replicated in cast by taking impression

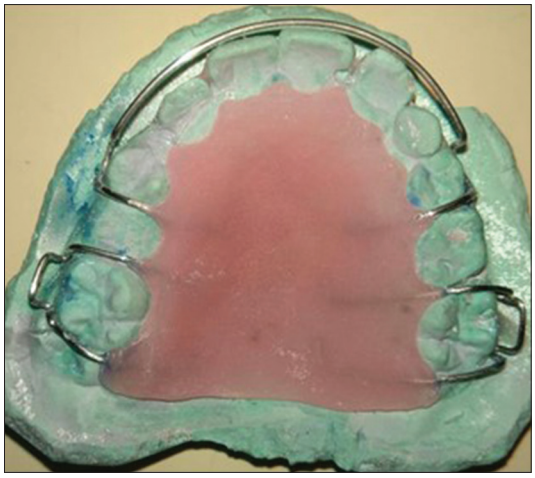

Figure 3: Obturator adapted to caste after the removal of tumor

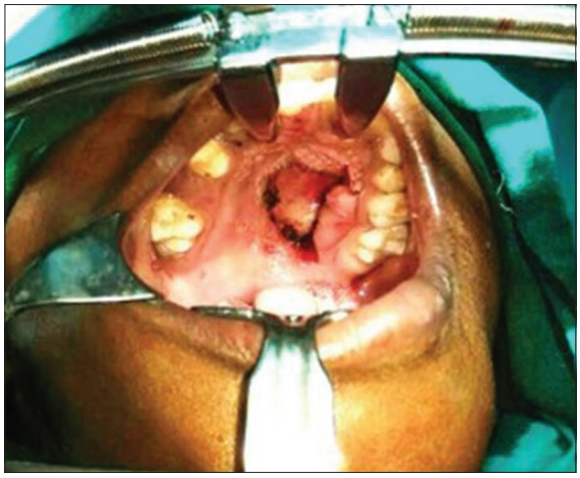

Figure 4: Excision of the tumor

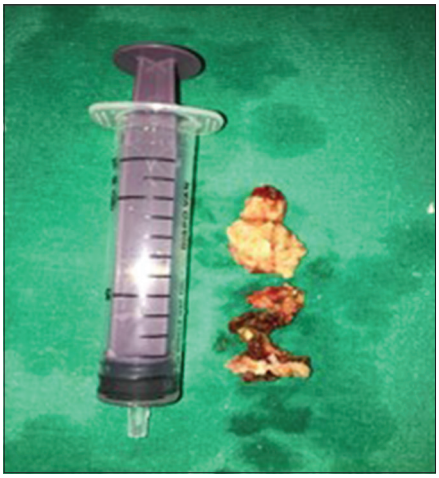

Figure 5: Tumor specimen

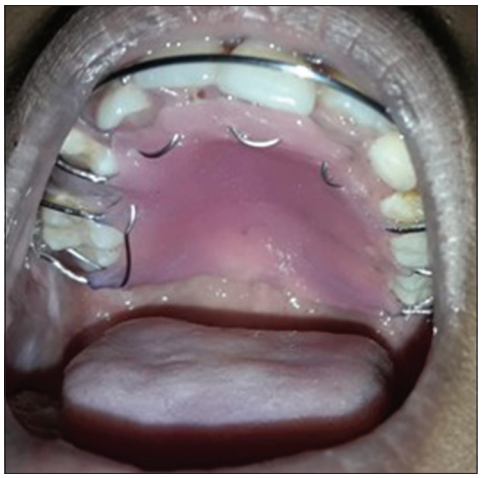

Figure 6: Palate with obturator

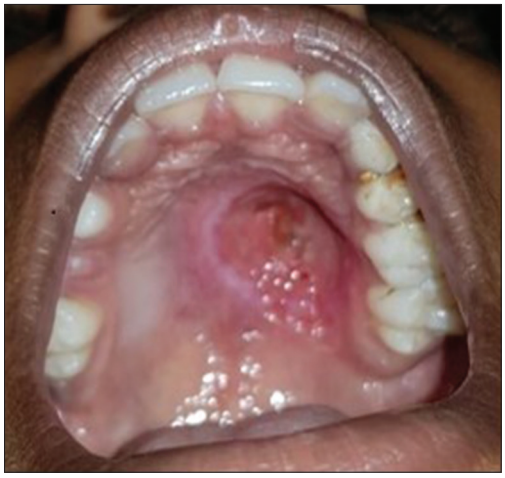

Figure 7: Follow-up after 3 weeks

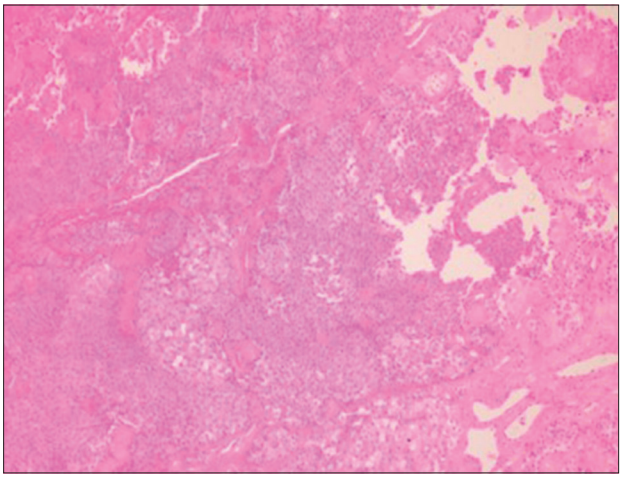

Figure 8: Lobules and sheets of proliferating tumor cells (×10)

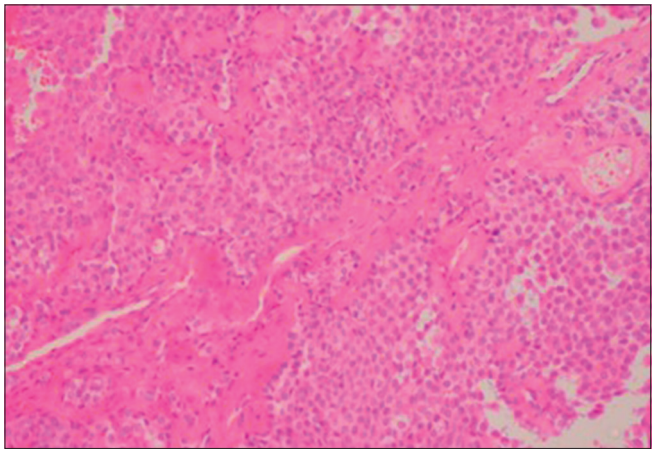

Figure 9: Tumor cells having round regular nuclei with moderate pink cytoplasm at many places giving a plasmacytoid appearance (H and E, ×40)

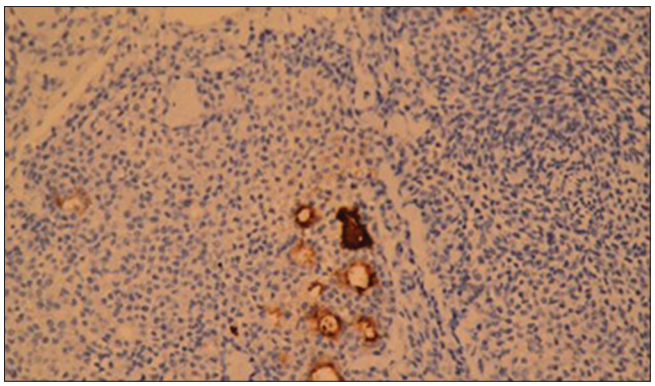

Figure 10: Embryonic membrane antigen positive in salivary ducts, negative in tumor cells

Myoepithelial differentiation of tumor cells is confirmed with immunohistochemistry using markers cytokeratin – CK-5 and CK-6, embryonic membrane antigen (EMA) [Figure 10], and smooth muscle actin (SMA) [Figure 11]. SMA shows focal positivity in tumor cells. EMA shows positivity in salivary ducts and is negative in tumor cells. Cytokeratin shows positivity in tumor cells that is CK-5 [Figure 12] and CK-6 [Figure 13] shows diffuse positivity in tumor cells.

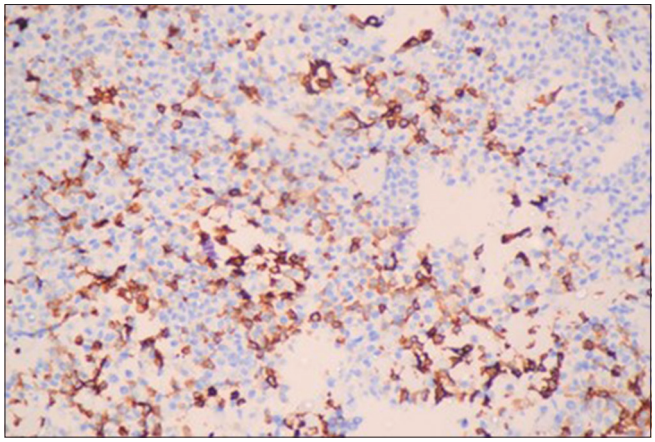

Figure 11: Smooth muscle actin focal positivity in tumor cells (×20)

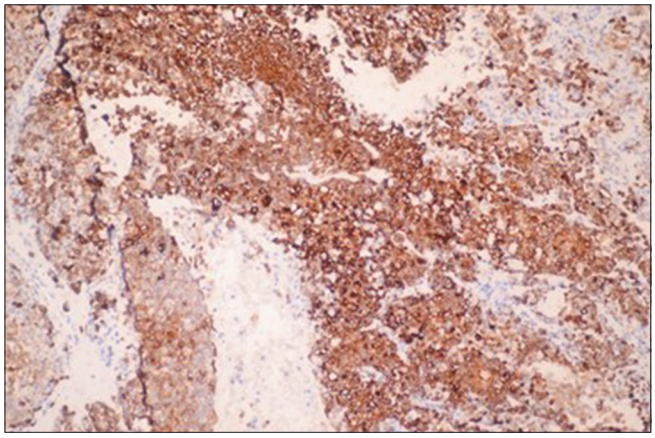

Figure 12: Cytokeratin CK-5: Positive in tumor cells

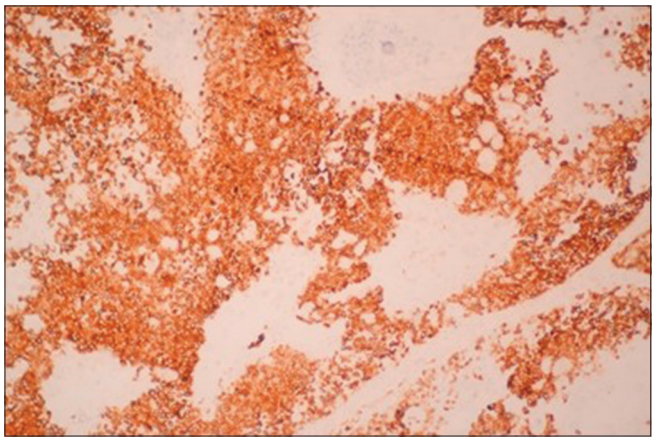

Figure 13: Diffuse positivity in tumor cells

DISCUSSION

MEs show four different morphological patterns which include nonmyxoid (solid), myxoid (pleomorphic adenoma like), reticular (canalicular like), and mixed. The cellular patterns of MEs consist of plasmacytoid cells, spindle cells, epithelioid cells, and clear-cell patterns which do not account for differences in recurrence rate, biological behavior, or the patient age. In the oral cavity, the plasmacytoid cell type is more commonly seen, whereas spindle cell type is more frequently seen in the superficial lobe of the parotid gland.[7] Myoepithelial cells are most commonly seen in salivary glands, unlike in this particular case, where it was in the palate. It is also seen in extra salivary gland tissues such as the breast, skin, lung, and larynx. MEs occurring in both salivary and extrasalivary tissues showed similar morphological and immunohistological characteristics. ME should be differentiated from its malignant counterpart that is, malignant ME, which is more aggressive and show recurrence even after adequate treatment. Histopathologically, the presence of cellular atypia, cellular pleomorphism, cellular necrosis, increased mitotic figures, invasive growth pattern, or combination invading the surrounding connective tissue are categorized as malignant ME. MEs of palate needs to be differentiated from other tumors of palate-like pleomorphic adenoma, neurinomas, hemangiomas, malignant tumors, metastatic tumors, lymphoma, solitary fibrous tumor, nerve sheath tumors, fibrous histiocytoma, paraganglioma, leiomyoma, leiomyosarcoma, hemangiopericytoma, and other inflammatory diseases. Many of these lesions share common clinical and radiological features, and hence, biopsy is needed for confirmation of the diagnosis of ME.[8] MEs show varying enhancement patterns on CT as faint enhancement, no significant enhancement, or marked enhancement. Factors influencing the enhancement pattern of ME include histological component, stroma, vascularity, and histological cell type. The cellular ME with fibrous stroma being more vascular showed more enhancement than those ME being rich in the myxoid stromal component. In our case, the tumor showed mild heterogeneous enhancement after contrast administration. Enhancement patterns may have a role in differentiating the slow-growing well-demarcated masses of the soft palate.[9]

Histologically, pleomorphic adenoma shows the presence of chondromyxoid matrix, which is considered most specific for the diagnosis while this feature is absent in ME along with the absence of glanduloductal differentiation. Peripheral nerve sheath tumor should be differentiated from spindle cell variant of ME, while clear-cell adenocarcinoma and mucoepidermoid carcinoma should be considered in the differential diagnosis of clear-cell variant of ME.[1] Simple surgical excision is the treatment of choice for benign ME if parotid gland is involved. If any other oral mucosal site is involved, excision with 0.5 mm margin peripherally and deep margins of the periosteum or muscle fascia as appropriate, can be considered. Excision is mostly curative and there is very little chance of recurrence

in benign ME. In case, recurrence is there, and it should be considered a potential unrecognized malignancy.

CONCLUSION

The epithelial-myoepithelial carcinoma, polymorphous low-grade adenocarcinoma, and adenoid cystic carcinoma, as well as inflammatory conditions, must be considered before ruling out the diagnosis of benign ME of the palate, based on their characteristic histopathological features. The treatment of choice for MEs is complete surgical excision with margins of the nonlesional area. The recurrence is rarely seen even after 10 years of surgery. Recurrence is rare for benign myoepithelial tumors, while the overall prognosis of myoepithelial carcinoma is poor[10]. Radiation therapy is used only in cases where surgical intervention is not feasible. The prognosis of this present lesion is favorable as the child is in a growing age group. It is advised to keep the patient under observation for the next 2 years.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sheldon WH. So-called mixed tumours of the salivary glands. Arch Pathol 1943;35:1-20.
2. Perez DE, Lopes MA, de Almeida OP, Jorge J, Kowalski LP. Plasmacytoid myoepithelioma of the palate in a child. Int J Paediatr Dent 2007;17:223-7.
3. Ferri E, Pavon I, Armato E, Cavaleri S, Capuzzo P, Ianniello F. Myoepithelioma of a minor salivary gland of the cheek: Case report. Acta Otorhinolaryngol Ital 2006;26:43-6.
4. Cuadra Zelaya F, Quezada Rivera D, Tapia Vazquez JL, Paez Valencia C, Gaitán Cepeda LA. Plasmacytoid myoepithelioma of the palate. Report of one case and review of the literature. Med Oral Patol Oral Cir Bucal 2007;12:E552-5.
5. Piattelli A, Fioroni M, Rubini C. Myoepithelioma of the gingiva. Report of a case. J Periodontol 1999;70:683-7.
6. Astarci HM, Celik A, Sungu N, Ustun H. Cystic clear cell myoepithelioma of the parotid gland. A case report. Oral Maxillofac Surg 2009;13:45-8.
7. Kanazawa H, Furuya T, Watanabe T, Kato J. Plasmacytoid myoepithelioma of the palate. J Oral Maxillofac Surg 1999;57:857-60.
8. Ellis GL, Auclair PL, editors. Epithelial neoplasms. In: Tumors of the Salivary Glands. Annapolis, Md, USA: ARP Press; 2008. p. 123-33.
9. Hiwatashi A, Matsumoto S, Kamoi I, Yamashita H, Nakashima A. Imaging features of myoepithelioma arising from the hard palate. A case report. Acta Radiol 2000;41:417-9.
10. Ren J, Liu Z, Liu X, Li Y, Zhang X, Li Z, *et al*. Primary myoepithelial carcinoma of palate. World J Surg Oncol 2011;9:104.