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Polymorphous low-grade adenocarcinoma arising from the nasopharynx: A rare presentation.

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Abstract

Introduction: Polymorphous low-grade adenocarcinoma is a rare lesion. Nasal fossa and nasopharynx locations have been described in less than 0.5% to 1% of the cases. The incidence of this case is very rare.
Case Presentation: We describe the clinicopathologic features of a case of PLGA arising from the nasopharynx in a 37-year-old Hindu female patient and also review the literature for differential diagnoses of minor salivary tumors arising at unusual locations.
Conclusion: This diagnosis is so rare that is oftened misdiagnosed in masses of the nasopharynx. Accurate diagnosis is important for the prognosis and treatment of the patient.

Introduction

Minor salivary gland malignant neoplasms account for 2% to 4% of head and neck malignant neoplasms, 10% of all oral cavity malignant neoplasms and 15 - 23% of all salivary gland malignant neoplasms.[1] Among the latter, the polymorphous low-grade adenocarcinoma (PLGA) is found, which was first described in 1983 simultaneously by Batsakis, et. al., and Freedman and Lumerman who named it as Terminal Duct Carcinoma and Lobular Carcinoma.[2] Before being described as an entirely different entity, this tumor was diagnosed as Pleomorphic Adenoma or Unspecific Carcinoma or even sometimes as Adenoid Cystic Carcinoma.[3] Another synonym for this tumor is low-grade papillary carcinoma of the palate. Polymorphous low-grade adenocarcinoma (PLGA) is a low-grade malignant tumor of the salivary glands, most often arising from the minor salivary glands with the palate being a common site. Polymorphous low-grade adenocarcinoma is the second most common malignant tumor of the minor salivary glands. It is a rare pathology that affects people in the age range from 30 to 70, with a female predilection in a 2:1 ratio.[4] Sixty percent of the cases

occur on the hard or 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.[4] Other uncommon locations are the major salivary glands, lacrimal glands, tongue, nasopharynx, nasal cavity and as well as the seromucinous glands in other locations. The striking histological feature is architectural diversity combined with benign cytologic features. We describe the clinicopathologic features of a case of PLGA arising from the nasopharynx in a 37-year-old female patient. On light microscopy, varied patterns were seen. The cells were uniform with bland nuclei. Infiltration in surrounding tissue and neural invasion was noted.

Case Presentation: A 37-year-Hindu female presented with a history of nasal intonation, recurrent nasal blockage and blood-stained nasal discharge. On examination there was a 4 by 3 cm mucosa covered, firm mass on the roof and lateral wall of the nasopharynx. Chest x-ray, ultrasonography of the abdomen , thyroid scan (radioactive Iodine 131 uptake) and bone scan were normal. CT scan showed a heterogeneous mass arising from the lateral wall of the nasopharynx without evidence of deeper extension. Subsequently, the patient underwent wide excision of the tumor and tissue was sent for histopathological examination. Grossly, the tumor was well circumscribed but unencapsulated (Figure 1), lobulated and measured 3.2 by 2 by 2.5 cm; cut section was solid, pale white, homogeneous and firm. Microscopically, architectural variability was striking, with the following patterns - tubular, trabecular and papillary (Figures 2A and 2B). Indian file and Cribriform pattern was noted at places. Arrangement of cells around blood vessels was present at places. The glands and tubules were lined by a single layer of cells (Figure 2C). The cells were round to ovoid to polyhedral, with the cytoplasm varying from eosinophilic to clear. Nuclear features were bland with vesicular nucleus, some showing an inconspicuous nucleolus. Hyperchromasia or mitoses were not seen. Lumen showed eosinophilic material at places (Figure 2D), which was variably PAS positive. There were no areas of necrosis. Stroma showed hyalinization at places. An area of neural invasion was noted. A final diagnosis of PLGA of the nasopharynx was made. Postoperative recovery was uneventful. At time of publication, there was no recurrence and the patient is having an uneventful life.

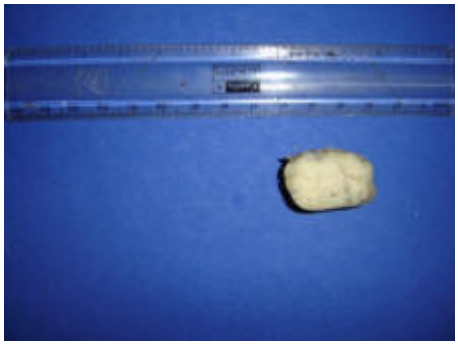
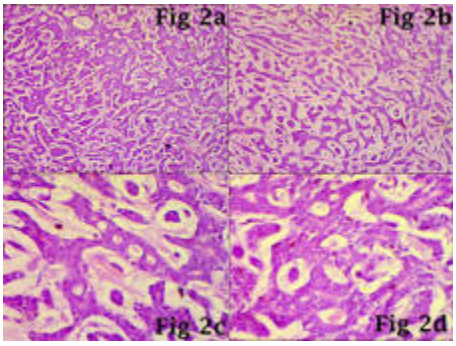


Figure 1: Gross Appearance of Removed Tumor



Figures 2A to 2D: Histopathology Appearance of Adenocarcinoma of the Nasopharynx

Enlarged Pictures at End of Document

Discussion

Polymorphous low-grade adenocarcinoma is a rare lesion. In a research study carried out by González Lagunas,[1] a sample of 59 malignant salivary gland tumors was assessed, and no PLGA was found. This lesion is located almost exclusively in the minor salivary glands within the oral cavity, mostly in the hard palate, making its extra oral presentation extremely rare. Nasal fossa and nasopharynx locations have been

described in less than 0.5 – 1% of the cases.[5] There is a clear female predilection, particularly affecting women in their forties and fifties which was simulated with our case. In the series of Castle, et al.,[6] the typical presentation was that of an asymptomatic mass lesion. A small number of patients (13 out of 164 cases) presented with a mass accompanied by pain, bleeding or ulceration. However, this patient did not have a more aggressive disease nor was she more prone to develop recurrences. Grossly, the tumor is usually unencapsulated, well circumscribed, lobular and firm. Microscopically, various architectural patterns could be seen in different areas (glandular, trabecular, tubular, Cribriform, "Indian file" and solid). Luminal eosinophilic material was seen in our case and has not been commonly described. Myxoid change in the background can be seen. In contrast to architectural polymorphism, the nuclei are uniform and bland with absent or negligible mitoses. Castle, et. al.,[6] detected neurotropism in a majority of cases with a targetoid appearance due to concentrically arranged tumor cells around a central nerve twig. The differential diagnoses of PLGA include adenoid cystic carcinoma (ACC) and pleomorphic adenoma. Both ACC and PLGA can have similar architectural patterns; however, the cells in ACC tend to be smaller with hyperchromatic nuclei and coarser chromatin. Mitoses are numerous. It is important to differentiate these two entities, as the prognosis and treatment are significantly different. The infiltrative growth pattern is an important diagnostic clue favoring PLGA when the differential diagnosis includes pleomorphic adenoma. Neurotropism, if present, also favors PLGA. Various immunohistochemical markers have been found to be positive like vimentin, cytokeratin, S-100, CEA, SMA and GFAP.[6] Proliferative markers like Ki-67 and p53 show variability in intensity and percentage of cells positive but generally the intensity is weak.[6] By immunohistochemistry we can differentiate between PLGA and adenocystic carcinoma. The staining qualities of EMA and CEA in the luminal cells were in equal proportion and intensity in adenocystic carcinoma whereas in PLGA, it was dissimilar. The best markers for distinguishing are CD117 and S100 protein. Staining for CD117 is typically strong and more diffuse in adenocystic carcinoma than PLGA., with S100 protein showing the reverse. SMA noted to be strong in adenocystic carcinoma.

Polymorphous low-grade adenocarcinoma described in this report corresponds to the experience described in other countries, with an incidence in the nasopharynx, an unusual site, and slow growth. Histological findings are also similar to those described above: Lobular growth, cord-like infiltration, and pale, ovoid nuclei.

Conclusion

Polymorphous low-grade adenocarcinoma is a rare, malignant neoplasm with occurrence in such an unusual site, with a clinical behavior similar to that of a benign neoplasm, with low symptomatology, which may determine a late diagnosis such as in the case presented. One should consider this diagnosis in patients with a mass in the nasopharynx.

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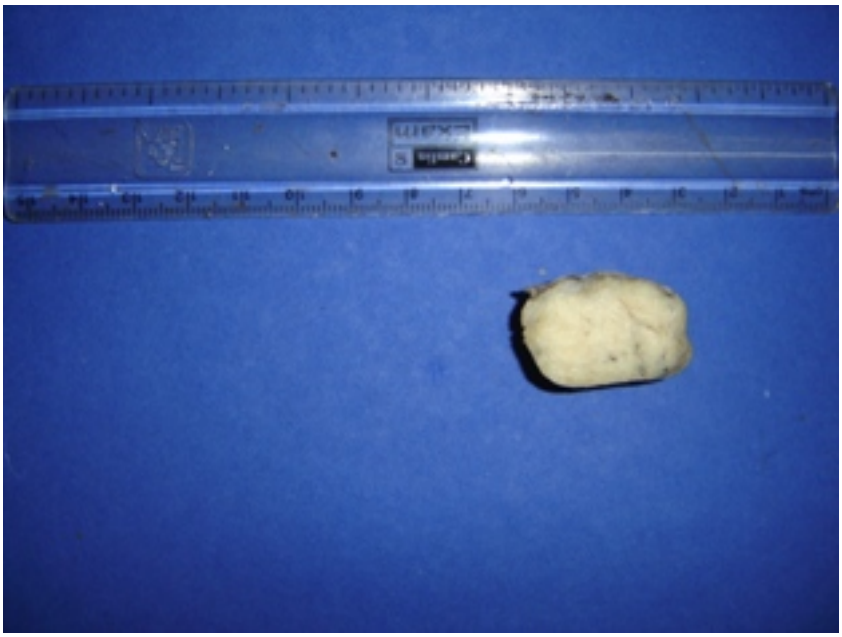

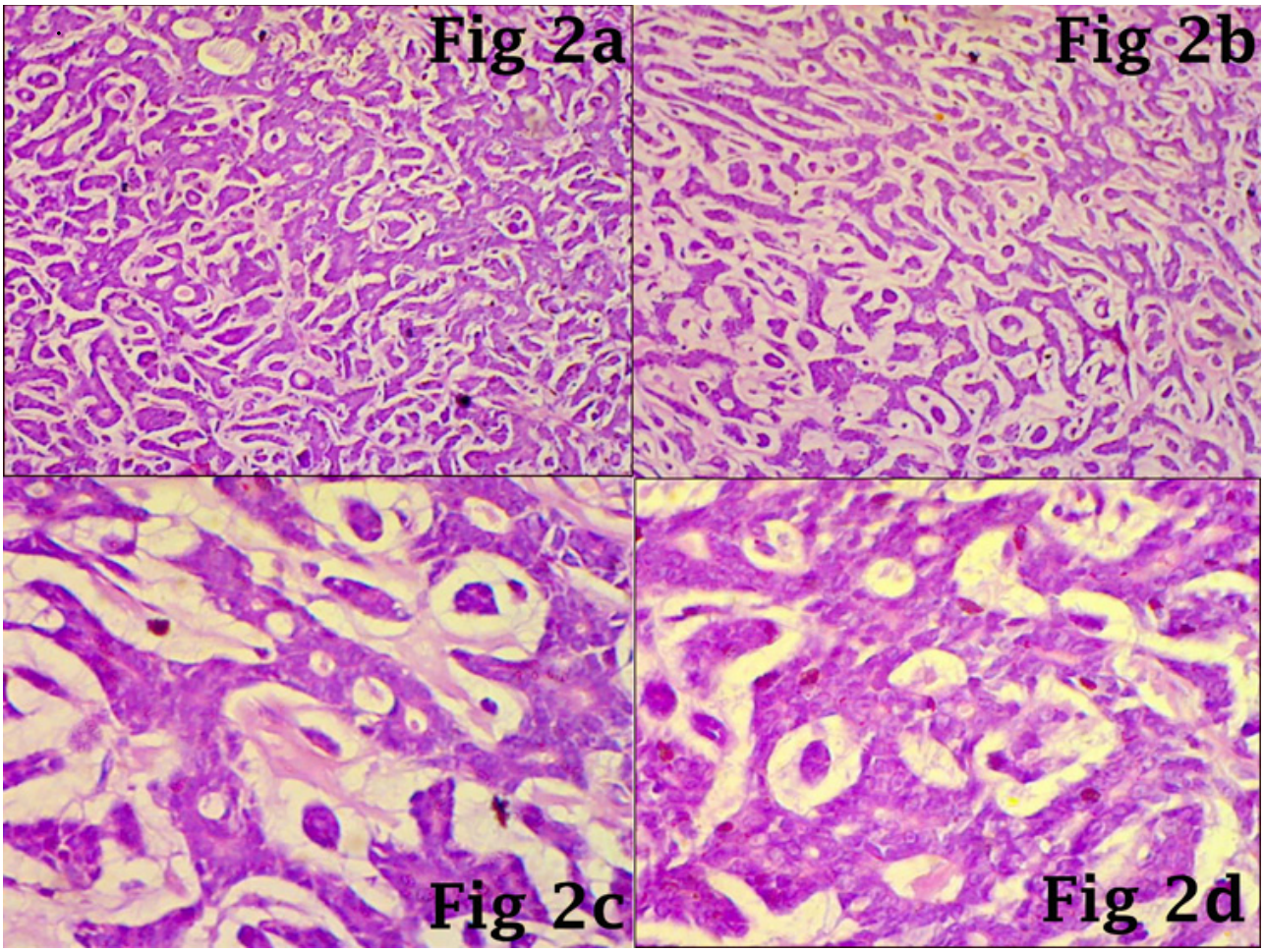
Fig 2a
Fig 2b
Fig 2c
Fig 2d