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Journal

Dermatology Online Journal, 19(8)

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Publication Date

2013

DOI

10.5070/D3198019265

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Peer reviewed

Case Presentation

Polymorphous low-grade adenocarcinoma in the upper lip: a well-described but infrequently recognized tumor.

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Dermatology Online Journal 19 (8): 8

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Abstract

Polymorphous low-grade adenocarcinoma (PLGA) is a rare malignant neoplasm arising almost exclusively in the minor salivary glands. PLGA occurs primarily in the oral cavity, especially in the palate, followed by the oral mucosa and upper lip [1,2]. Conditions involving these locations are often presented at dermatological clinics. Therefore, dermatologists should consider this entity in the differential diagnosis of the oral cavity tumors. We present a case of PLGA in the upper lip.

Case synopsis

A 75-year-old woman was referred to our department for a 3 year-history of a painless and slow-growing nodule in the upper lip. Physical examination revealed a firm nodular and well-circumscribed submucosal lesion, 2 cm in size, with an intact overlying mucosa (Figure 1). Excision of the lesion was performed. Histology showed a solid tumor, not encapsulated, consisting of a proliferation of epithelial cells distributed in different patterns (Figure 2), infiltrating the surrounding fat tissue. The cells were isomorphic and of small to intermediate size. Perineural invasion and mitoses were identified. A diagnosis of PLGA was made. A cervical CT and thorax radiography were normal. A wide local surgical excision and reconstruction with a semilunar advancement flap was performed. Two years after treatment, the patient is free of disease.



Figure 1. Nodular submucosal lesion in the upper lip.

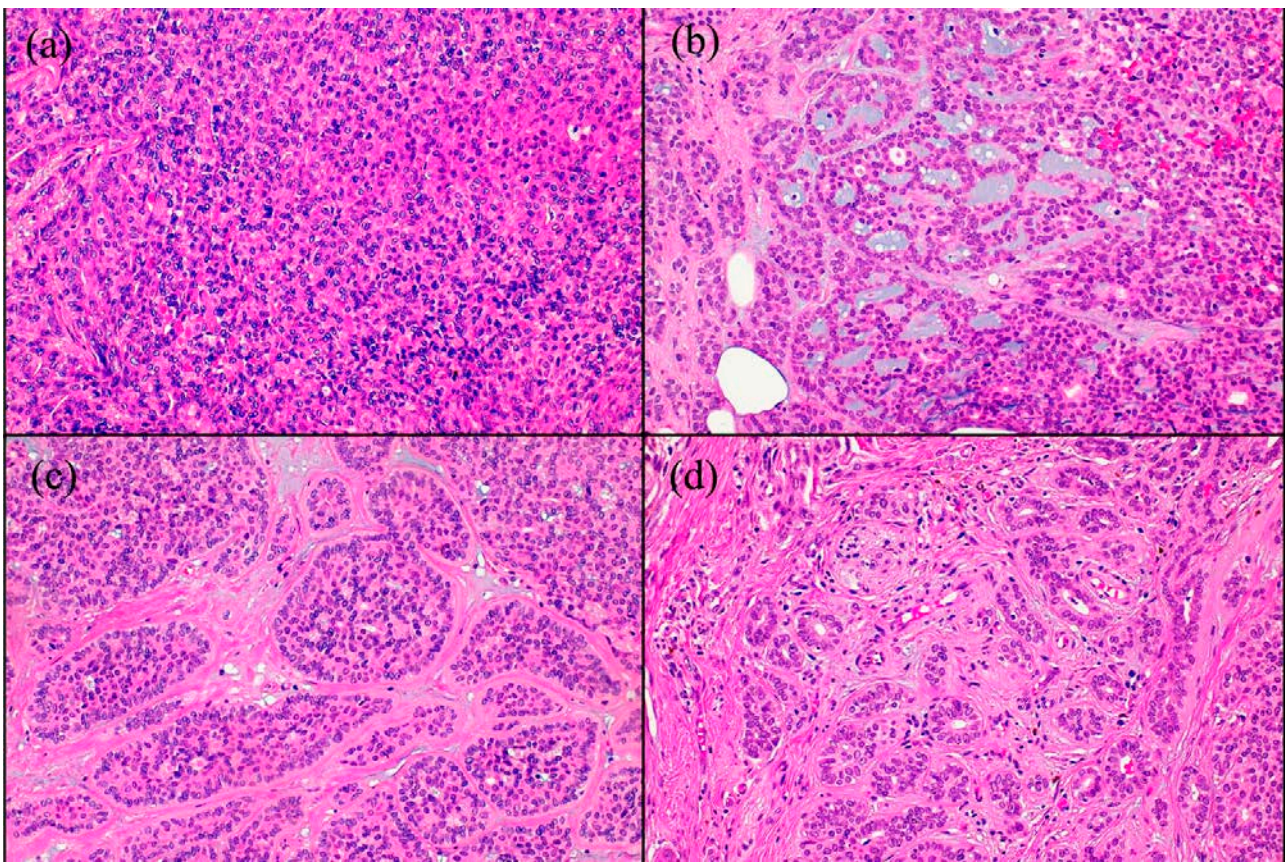


Figure 2. Different histopathological patterns of the tumor (H&E x20). (a) Solid growth pattern. (b) Cribiform architectural arrangement of the tumor cells. (c) Lobular pattern with large solid nests. (d) Tubular and trabecular architecture.

Discussion

The term PLGA was introduced by Evans and Batsakis in 1984 to describe a malignant neoplasm of minor salivary gland origin [1]. Many of the larger series identify PLGA as the second or third most common malignancy of the minor salivary glands [2,3]. There appears to be a female preponderance and the age of presentation typically peaks in the sixth decade of life [4].

The typical presentation is that of an indolent submucosal lump, which may occasionally be painful or even ulcerated [4]. The clinical features will not distinguish PLGA from the other malignant intraoral minor salivary gland tumors and a biopsy is needed to confirm the diagnosis. Histologically, PLGA is characterized by a triad of infiltrative growth, multiple architectural growth patterns that include solid, trabecular, glandular, cribiform, fascicular, cordlike, and papillary (hence the term polymorphous) forms, and cytological uniformity. Because of its morphologic pleomorphism, PLGA has often been misdiagnosed as pleomorphic adenoma or adenoid cystic carcinoma. It is locally invasive and neurotropism is a characteristic feature [4].

Complete local excision with wide margins is the treatment of choice. Late recurrences after 5 years are reported in 9% to 26% of cases despite free surgical margins. Hence, long-term follow-up of at least 15 years is recommended. About 5% to 10% of tumors metastasize to cervical lymph nodes, but distant metastases are unusual. The role of radiotherapy and chemotherapy is not clear in the management of PLGA [4,5].

Since its description, several case series have described this entity, all of them in the oral medicine and surgery literature, with no documented cases in the dermatological journals. PLGA is not mentioned either in most of the dermatology text books. We believe the familiarity with this condition is important in the dermatology setting to facilitate prompt and proper diagnosis and adequate treatment of this well-defined but infrequently recognized disease.

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