

Polymorphous Low-Grade Adenocarcinoma

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A 60-year-old woman with a 3-year history of a gingivoalveopalatal mass underwent an incision biopsy.

Microscopically, the lesion centered in the stroma is infiltrative (*Figure 1*) and architecturally diverse, having cystic (*Figure 2*), linear or “Indian file” (*Figure 3*), solid and tubular (*Figure 4*) patterns. The cells are uniform in size, round to oval and have bland cytologic features with vesicular nuclei and inconspicuous nucleoli (*Figure 4*). The clinical data and histomorphologic features characterized by architectural diversity yet cytologic blandness lead us to the diagnosis of polymorphous low-grade adenocarcinoma.



Figure 1. Hematoxylin and Eosin (40x) Tumor within the stroma with an infiltrating growth pattern

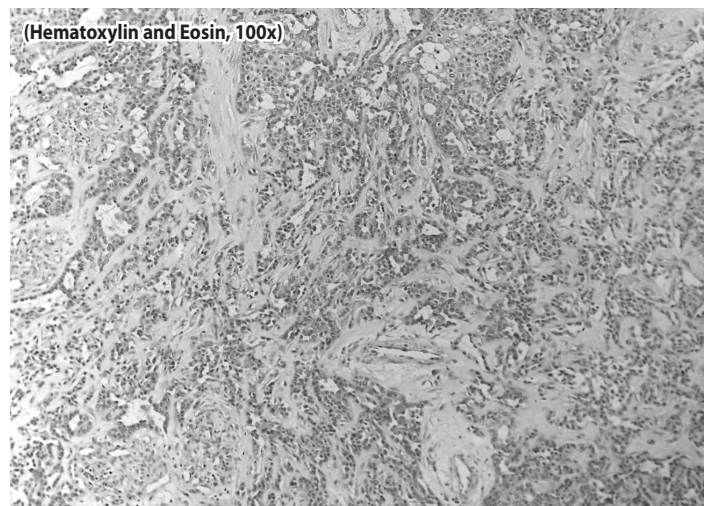


Figure 2. Hematoxylin and Eosin (100x) Tumor showing tubules and small cystic structures.

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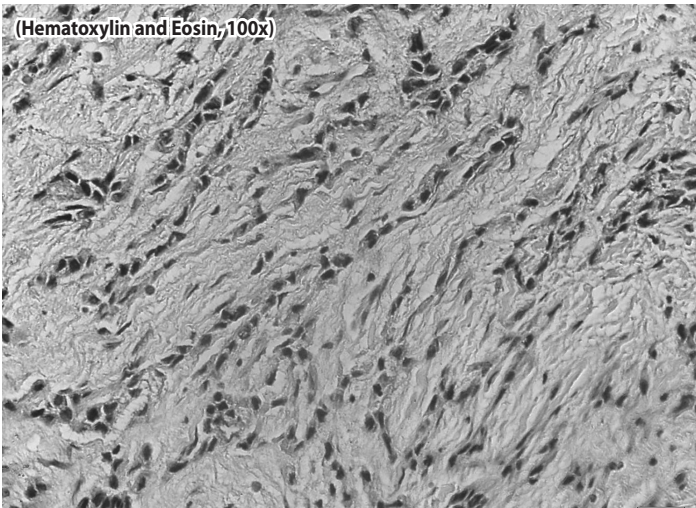


Figure 3. Hematoxylin and Eosin (100x) Tumor cells showing a linear ("Indian-file") arrangement

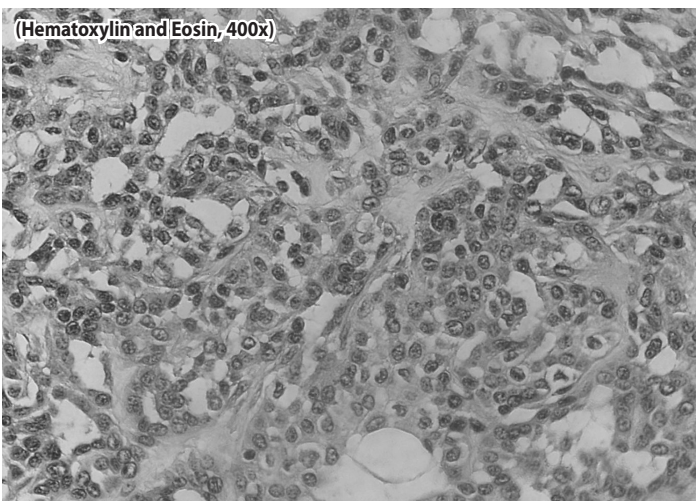


Figure 4. Hematoxylin and Eosin (400x) The round to oval tumor cells are uniform in size with bland nuclei and inconspicuous nucleoli

Polymorphous low-grade adenocarcinoma (PLGA) is a malignant epithelial tumor characterized by cytologic uniformity, morphologic diversity, an infiltrative growth pattern and low metastatic potential.¹ It is the second most common intraoral malignant salivary gland tumor¹ following mucoepidermoid carcinoma. The tumor is found almost exclusively in minor salivary glands and is rare in extraoral locations including major salivary glands.² The tumor affects a wide age range (16 – 95 years; mean 60 years) with only two pediatric cases reported¹ and has a female predilection.^{3,4} It usually presents as a painless mass located within the oral cavity³ 60% of which are located in the palate.¹ They are characteristically unencapsulated although well-circumscribed.³

This entity is architecturally diverse ("polymorphous") even within a single tumor, with solid, tubular, trabecular, cribriform, papillary and

linear patterns being described. Perineural invasion is common although it was not seen in this case. The tumor cells are small to medium sized and uniformly round to polygonal. The nuclei are bland and vesicular with occasional small inconspicuous nucleoli. Mitotic figures can be found occasionally but are never numerous.^{3,8}

The morphologic heterogeneity in small biopsies and frozen section samples can be confused with pleomorphic adenoma and adenoid cystic carcinoma.^{6,7} Glial fibrillary acid protein may help as PLGA is typically non-reactive in contrast to pleomorphic adenoma.² De Araujo and others site that uniformly positive vimentin, CK7 and S100 staining favors PLGA over adenoid cystic carcinoma.⁶ Tumor cytology and histology are quite characteristic - recognizing the constant cytological appearance despite the diversity of architectural tumor patterns should aid one in diagnosing PLGA.

PLGA, despite its infiltrative growth pattern and propensity for perineural invasion usually runs an indolent course. Nodal metastasis and distant spread are rare, occurring in less than 1% of cases.⁴ Seethala and others report that extrapalatal location is associated with a more aggressive clinical course.⁵ Complete surgical excision is the primary treatment with neck dissection reserved for nodal metastasis.¹ One-third of patients may have a local recurrence and lifelong monitoring is suggested. Re-excision is amenable in these cases.^{5,6}

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