Recurrent polymorphous low-grade adenocarcinoma manifesting as a sinonasal mass: A case report

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Abstract
Polymorphous low-grade adenocarcinoma (PLGA) is a malignant neoplasm that tends to follow a benign clinical course. Recurrences are uncommon. We report a case of recurrent PLGA of the paranasal sinuses that manifested as a large mass that filled the entire nasal cavity and left maxillary sinus. To our knowledge, this is the first reported case of a recurrent PLGA of the paranasal sinuses.

Introduction
Polymorphous low-grade adenocarcinoma (PLGA) is a malignant neoplasm that almost always arises in the minor salivary glands. It generally involves the palate, but it has also been reported in the base of the tongue, upper lip, buccal mucosa, tonsil, and retromolar pad. The tumor is fairly common and generally follows a benign clinical course. The prognosis is quite good, explaining the “low-grade” nomenclature. Recurrences are uncommon, as are metastases.

We report a case of a recurrent PLGA that manifested as a large mass in the left midface. To our knowledge, this is the first reported case of a recurrent PLGA of the paranasal sinuses.

Case report
An 84-year-old man presented with a 3-month history of progressive swelling of the left maxilla. The patient had a history of pleomorphic adenoma of the palatal area, which had been treated with a partial surgical excision approximately 4 years earlier. He had done well until he noticed the increasing swelling over his left midmaxillary area, which had caused significant nasal obstruction.

Physical examination revealed the presence of an expansile mass that had arisen from the left maxillary alveolus and extended superiorly to the orbital floor. The face of the maxilla was eroded and displaced anteriorly. The floor of the nasal cavity was replaced by the cystic mass, which extended across the midline and over to the right maxillary sinus remnant. The mass extended superiorly on the left to the orbital floor. Evidence of the previous partial maxillectomy was noticed.

Computed tomography (CT) demonstrated a large complex mass arising from the maxillary sinus, primarily on the left side; extensive bony destruction was evident (figure 1). The inferior orbital wall on the left side was eroded, as was the ethmoid region. Analysis of fine-needle aspiration biopsy revealed the presence of only a few atypical cells.

The patient was brought to the operating room, where a partial maxillectomy and tumor debulking were performed via a lateral rhinotomy approach. The incision was extended along the gingival buccal sulcus in a Caldwell-Luc fashion. Intraoperatively, we noted that a large multilobulated mass had eroded the maxilla, including the maxillary floor, the palatal area, and the alveolar ridge. The tumor extended posteriorly to erode the posterior wall of the maxilla and laterally to involve the lateral wall of the maxilla and the orbital floor. A partial maxillectomy was performed, and the mass was resected. A portion of the capsule was allowed to remain on the orbital floor where erosion was noted. A portion of the palate was resected, and the orbital floor and the periorbita were left intact. The palatal defect was not closed.

The excised tumor specimen measured 3 × 6 × 2 cm. No definitive capsule was identified. The cells were small and lacking in nuclear atypia. Mitotic figures were absent. Under low-power magnification, the central portion of the tumor was solid. Small lobules of cells were located in the periphery of the lesion. Under higher power, most of the tumor cells themselves were bland and relatively uniform. The round and ovoid nuclei were either normal in size or slightly enlarged (figure 2, A). Mitoses were
rare. The histopathologic appearance was most suggestive of a PLGA.

Postoperatively, the patient did well and his nasal congestion was minimal. At the 1-year follow-up, he showed no signs of disease progression.

The pathologic diagnosis of pleomorphic adenoma was originally made 8 years ago. This tissue was subsequently reviewed at our institution, and the specimen was consistent with a diagnosis of PLGA.

Discussion

PLGAs are malignant epithelial tumors characterized by bland, uniform nuclear features, diverse but characteristic architecture, infiltrative growth, and prominent neurotropism (figure 2, B). PLGAs arise almost exclusively in the minor salivary glands. Histologically, one of the characteristic findings is the formation of small tubular structures that have distinct central lumens that are lined with cuboid cells. These structures are sometimes associated with streaming columns that give them a target-like appearance. In addition, cribriform, cystic, and papillary-cystic areas may be seen in the same specimen. The stroma is collagenous, but sometimes hyaline features develop. Psammoma-like calcifications are occasionally seen.

Because of the cells’ prominent neurotropism, perineural infiltration is a frequent finding. In fact, perineural spread is more common in PLGA than in any other salivary gland carcinoma, including adenoid cystic carcinoma. Perivascular invasion is sometimes seen, but it is less common than perineural invasion. The cells often infiltrate bone.

PLGA was first identified as a specific salivary gland adenocarcinoma in 1983 by two independent groups of investigators; Freedman and Lumerman reported 12 cases as lobular carcinoma, and Batsakis et al reported 12 cases of what they called terminal duct carcinoma. The following year, Evans and Batsakis used the term polymorphous low-grade adenocarcinoma to describe this malignancy as a clinically and pathologically discrete entity. In subsequent
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reports, other authors have distinguished papillary and nonpapillary variants of PLGA. The papillary variant is more likely to recur, to metastasize to regional and distant sites, and to result in death.4

Since the original description of PLGA was published, this entity has been recognized with increasing frequency. In their study of a series of 426 minor salivary gland tumors, Waldron et al reported that PLGA represented 11% of all tumors and 26% of malignant tumors.5 Combined data from the Armed Forces Institute of Pathology showed that PLGA accounted for 7.4% of all minor salivary gland tumors and 19.6% of those that were malignant.5

PLGA usually occurs at the junction of the hard and soft palates. Other sites include the buccal mucosa and upper lip.6 Rarer sites that have been reported include the nasopharynx and the nose. Occasionally, the tumor arises in the floor of the mouth; a large tumor in this location may present as a neck mass.

Although the number of cases of PLGA with long-term follow-up is limited, various investigators have followed patients for as long as 11 to 37 years.6 The reported recurrence rate ranges from 0 to 30%, and the rates of both regional and lymphatic metastasis are approximately 10%6.

The appropriate treatment for this disease is wide but conservative surgical resection. There are no data showing any benefit to adjunctive chemotherapy or radiation therapy. Unlike the case with other adenocarcinomas, tumor metastasis and death are rare, which indicates that PLGA is a distinct clinical and pathologic entity.

References