Polymorphous Low-Grade Adenocarcinoma (PLGA) in an 18-Year-Old Male

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Abstract Polymorphous low-grade adenocarcinoma (PLGA) is a minor salivary gland tumor with a low malignant potential. It is twice more common in females, with a mean age of presentation at 59 years. It is a very slow-growing tumor with mean duration of symptoms that range from 27 months to as long as 40 years. We report a case of a male patient who was found to have PLGA with symptoms since birth. The patient was treated with wide local excision with good results. The lip is a rare location for PLGA, and its occurrence in adolescent age groups is even rarer.

Keywords Polymorphous low-grade adenocarcinoma · Tumor · Salivary gland

Introduction

Polymorphous low-grade adenocarcinoma (PLGA) is a distinctive minor salivary gland neoplasm with indolent course [1]. Since its recognition in WHO classification in 1990, it is recognized as the second most common neoplasm after mucoepidermoid carcinoma. PLGA represents 7–11 % of all minor salivary gland tumors [2, 3]. It is more common in females (2:1) [4]. Mean age of presentation is 59 years; it does not seem to occur in the first or second decade of life [3] and has been rarely reported in adolescence [5]. Average duration of symptoms ranges from a few days to 40 years with a mean of 27 months [3]. We report a case of PLGA of the lip in an 18-year-old patient.

Case Report

An 18-year-old male patient presented with an ulceroproliferative lesion at the left angle of the mouth (Figs. 1a, b). He had pain and bleeding from the lesion. The patient was not addicted to alcohol, smoking, or tobacco chewing. The patient gave a history of having a round solid mass at the same site since birth which ulcerated 3 months ago, making him seek medical attention. He reported undergoing two surgical interventions at the same site of the present tumor 9 years and 2.5 years back, the details of which were not available as these were performed elsewhere. Examination showed an ulceroproliferative lesion measuring about 4 × 3 cm at the left upper lip involving commissure. The lesion was friable, bled on touch, and had irregular margins with an indurated base. There was no clinically detectable cervical lymphadenopathy. CT scan of the same revealed infiltration into the surrounding fat plane without any evidence of cervical lymphadenopathy. A punch biopsy was taken which revealed a malignant round cell tumor in a mucoid background. The patient was treated with a wide local excision and nasolabial flap reconstruction (Figs. 1c, d). Postoperative HPE showed a polymorphous low-grade adenocarcinoma of the minor salivary gland with cells arranged in cords, tubules, and solid nests with mucoid background (Fig. 2). The patient is under follow-up and has no evidence of local recurrence, 1 year after excision.

Discussion

PLGA are minor salivary gland tumors with predilection for intraoral sites. These tumors commonly occur in decreasing order of frequency at the hard palate, soft palate, buccal mucosa, and alveolar ridge [6]. These tumors are common in females with a 2:1 ratio [4]. It is common in the fifth decade of life with mean age at presentation of 59 years with 70 % of patients between 50 and 79 years [3]. Duration of

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Fig. 1 Clinical photograph showing the lesion on the upper lip and angle of the mouth.

a Outside view. **b** Mucosal view postoperative results. **c** Postoperative results from the left side. **d** Postoperative results from the front

symptoms ranges from a few days to 40 years with average duration of 27 months.

PLGA clinically presents with a slow-growing mass which may or may not be accompanied by pain, ulceration, or bleeding [3]; ill-fitting dentures may be incidentally found. There may not be obvious infiltration into surrounding structures. Cervical lymph nodes are usually not involved; however, there are reports of its occurrence. The disease often has a long indolent course, with distant metastasis being rare [1]. Macroscopically, these are solid ovoid masses, usually unencapsulated and typically lying

close to overlying epithelium. The cut surface shows light yellow to tan parenchyma. Tumor necrosis is infrequent.

Microscopically too, these tumors are well circumscribed, unencapsulated with infiltration into the presalivary gland fat tissue, but true infiltration into muscle is uncommon. Tumor cells are seen to invade the remaining salivary gland, separating lobular units and surrounding acini but not actually infiltrating them. Overlying skin is usually not involved even when skin infiltrates, but tumor infiltrates may be seen occasionally. PLGA displays a mixture of growth patterns within a single tumor, like solid islands,

Fig. 2 **a** Resected specimen. **b** Hematoxylin and eosin-stained photomicrograph showing low-power view showing solid fronts of the tumor infiltrating the normal gland (seen at the upper right corner). **c** Low-power view showing cells in cords, tubules, and solid nest with mucoid background. **d** High-power view of the same

glandular proliferates, tubules, trabeculae, cribriform nests, and linear single-cell Indian-file infiltration [3].

Tumor cells are uniformly round to polygonal with indistinct cellular borders and abundant eosinophilic cytoplasm. Nuclei are round containing vesicular nuclear chromatin and inconspicuous nucleoli. Cells are always small to medium sized, regular, and lacking in nuclear atypia. Mitotic figures are infrequent, and tumor necrosis is rarely seen. Tumor cells are arranged concentrically around a nidus, most often nerve bundles producing a targetoid appearance, which is quite characteristic of PLGA. Tumor cells are surrounded by hyalinized eosinophilic stroma and may occasionally show myxoid degeneration. A slate gray–blue stroma, when present, sometimes is characteristic of PLGA. The cytologic differential diagnosis of PLGA includes adenoid cystic carcinoma, pleomorphic adenoma, and monomorphic adenoma. Immunohistochemically moderate to strong diffuse immunoreactivity to cytokeratin, CEA, S-100 protein, and bcl-2 is found [7]. Focal weak reactivity with antismooth muscle actin as well as c-kit is occasionally demonstrated [1]. Glial fibrillary acidic protein (GFAP) is often absent and is used to differentiate PLGA from pleomorphic adenoma which shows strong positivity for GFAP. Reactivity for proliferation markers like Ki-67 and p53 is variable, most of the cells showing weak nuclear reactivity [8].

Treatment

Complete surgical excision including bone, if necessary, is the treatment of choice [9, 10]. No adjuvant therapy is recommended, though frequently postoperative radiotherapy is given. After a successful surgical removal, local recurrence rates of 17 to 24 % with overall survival of 97 % are reported [3]. Metastasis is unusual (9 %). If it occurs, it mainly affects regional lymph nodes. Disease-related deaths are uncommon.

Conclusions

PLGA is a rare neoplasm in adolescents; its occurrence on the lip, local recurrence after surgical excision, and presence

since birth are also rare. The present case demonstrates an unusual presentation and clinical course of this rare neoplasm.

Conflict of interest None is declared.

Consent The patient's consent was obtained for publication of this case report.

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