

## Papillary Cystadenocarcinoma In The Retromolar Area – A Brief Case Report –

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Received : May 11, 2005  
Accepted : October 21, 2005

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Salivary gland papillary cystadenocarcinomas are rare lesions, and particularly when they are found in the oral cavity. They have been recognized as being low grade carcinomas of the salivary glands. The author reports here on a case of recurrent papillary cystadenocarcinoma in the right retromolar area. The initial mass had multicystic and papillary structures with low grade features. The recurred mass showed basically the same histologic features. However the layer of papillae and the solid portion were increased and the tumor cells were larger and more pleomorphic with prominent nucleoli and frequent mitoses. Focal comedo type tumor necrosis and spindle cell proliferations in the surrounding soft tissue were present. This is a very rare report of a minor salivary gland papillary cystadenocarcinoma in a Korean, and the morphologic dedifferentiation was accompanied by the clinical recurrence.

**Key Words** : Papillary cystadenocarcinomas; Salivary gland; Retromolar

Papillary cystadenocarcinomas of the salivary gland are very rare and distinct neoplasms.<sup>1-3</sup> They had been reported as an atypical type of adenocarcinoma, a malignant papillary cystadenoma, a low grade papillary adenocarcinoma or a mucus producing adenopapillary carcinoma.<sup>3-8</sup> The WHO classification defined papillary cystadenocarcinoma as a low grade malignant tumor that's characterized by cysts and papillary endocystic projections.<sup>2</sup>

Histopathologically, this tumor is characterized by prominent cystic, and frequently papillary and invasive growth, but it lacks the common features of the cystic variants of the other salivary gland carcinomas such as mucoepidermoid carcinomas, acinic cell carcinomas, and etc.<sup>2,8</sup>

Microscopically, single or multilayered epithelial tumor cells line the cystic space and papillae, and this occasionally forms the cribriform pattern.<sup>1,8-10</sup> The predominant tumor cells are composed of small cuboidal cells, large cuboidal cells, tall columnar cells and a mixture of these cell types. In some tumors, the large cuboidal cells occasionally show either an apocrine or oncocytic appearance,<sup>9</sup> and mucous secreting cells can also be present.<sup>10</sup> Usually, mitotic figures are rarely present.<sup>1,7,8,11</sup> Perineural invasion or vascular invasion is also rarely seen.

The author reports here on a rare case of recurrent papillary cystadenocarcinoma in the retromolar area.

### CASE REPORT

A 55-year-old female visited our hospital complaining of an enlarging mass that was detected two months ago in the oral cavity. The mass was firm and painless with a bleeding tendency. The physical examination, chest X-ray and other laboratory examination were unremarkable. Computed tomography (CT) scans showed a 0.5 cm-sized mass in the right retromolar trigone area with no significant lymphadenopathy. There was no evidence of metastasis. The patient underwent a total mass removal.

Macroscopically, the resected specimen consisted of several pieces of soft tissue, that measured about 1.0 cc in total volume. Microscopic examination showed several fragments of papillary fronds of variable sizes and shapes with small foci of solid growth. The tumor cells were small and cuboidal with hyperchromatic nuclei, mild atypia and occasional small nucleoli (Fig. 1). The cytoplasm showed partial oncocytic change or a hobnail appearance. Mitotic figures were rare.

The patient did not receive any more adjuvant chemotherapy or radiation. Thereafter, the patient experienced occasional bleeding and mild aching pain at the operating site. 17 months after the initial resection, the patient felt a slowly growing mobile mass with mild swelling in the same area. CT scans showed a 1.5 cm sized well-defined heterogeneous enhancing mass. There was no

evidence of metastasis to other sites including the head and neck area. The patient underwent a total excision of the recurrent mass.

Macroscopically, this specimen was a soft tumor that was partially covered with skeletal muscle and fascia with some fragments of bone. The cut surface was composed of multiple cystic spaces, and the specimen measured  $1.5 \times 1.5 \times 1.0$  cm. It was partially circumscribed with a yellowish gray granular surface. Focal hemorrhage and necrosis were noted (Fig. 2).

Histologically, the tumor was composed of various sized cystic spaces in about 70% of the lesion, and there were some small solid islands with invasive borders. The cystic lumen was occupied by numerous papillary projections. The shape of papillae was variable from epithelial tufts to large arborescent structures (Fig. 3A). The cyst walls and papillae were lined by large cuboidal and columnar cells in single to multi layers. The tumor cells had moderate pleomorphism with irregular nuclear membranes, vesicular chromatin and prominent eosinophilic nucleoli (Fig.

3B). The mitotic count was 15-20/10HPF with a few atypical mitotic figures. Therefore, the recurred tumor was considered to have transformed into a higher grade as compared with the previous tumor. Foci of comedo-type tumor necrosis and reactive spindle cell proliferations in the surrounding soft tissue were observed. Some foamy macrophages were present. No vascular or perineural invasion was present.

PAS and mucicarmine stains showed no mucin or eosinophilic granular materials in the cytoplasm of the tumor cells. The tumor cells were immunohistochemically positive for cytokeratin, and they were negative for vimentin, S-100 protein, smooth muscle actin and CEA.

## DISCUSSION

Pleomorphic adenoma is the most common benign tumor

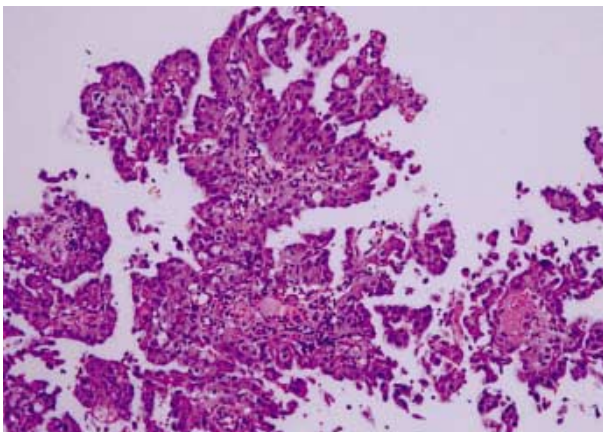


Fig. 1. The initial mass shows papillary fronds with small cuboidal cells with mild atypia.

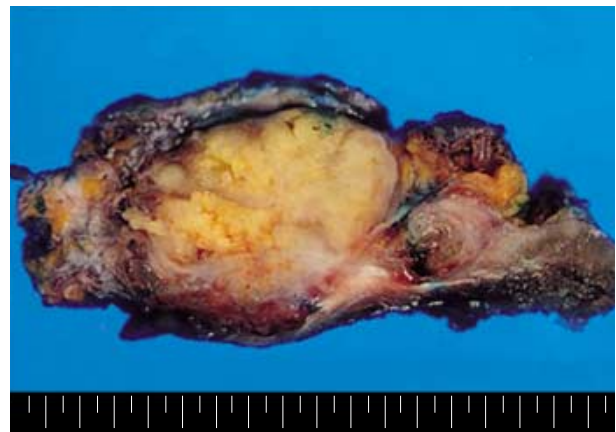


Fig. 2. On cut surface, mass shows multiple cysts and yellowish gray granular surface.

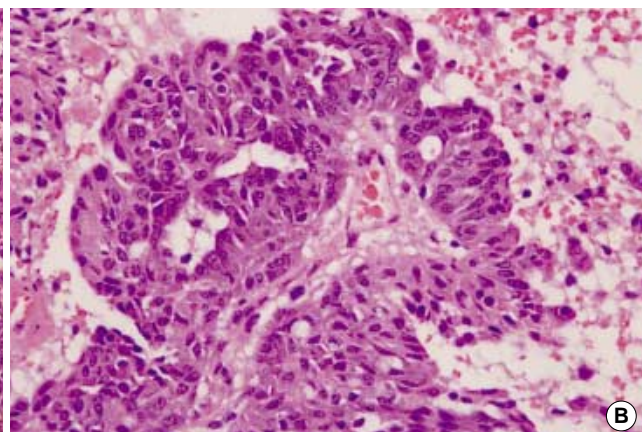
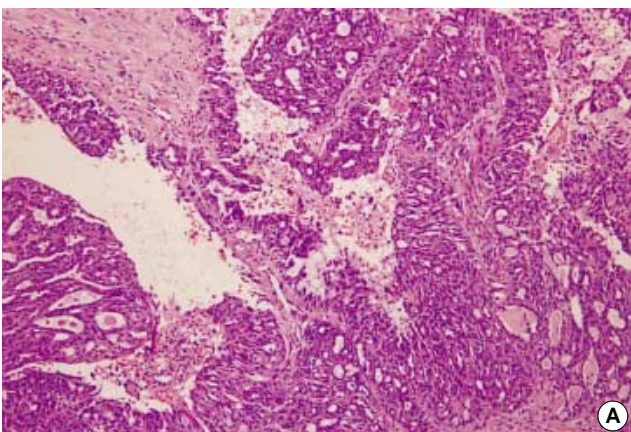


Fig. 3. The recurred mass shows several fragments of papillary fronds (A) with large cuboidal and columnar cells with moderate pleomorphism, vesicular nuclei and prominent nucleoli (B).

for the intraoral minor salivary glands, and mucoepidermoid carcinoma is predominant among the malignant tumors. The most common site of the cystadenocarcinoma of the minor salivary gland is the lip, and this is followed by buccal area, palate, tongue and retromolar area. The retromolar area is a very rare tumor site, especially for papillary cystadenocarcinoma.<sup>1,9,11</sup>

The salivary gland papillary cystadenocarcinomas arise in the age from 20 to 86 years (mean, 58.8 years), and men and women are equally affected. The patients usually present with a slow growing, asymptomatic mass.<sup>1,9</sup> A bleeding tendency was seen in this present patient, and this was due to erosion of the surface epithelium despite the small size of the initial mass about 0.5 cm in diameter, yet this is a rare symptom. At recurrence, the mass measured 1.5 × 1.5 × 1.0 cm. The reported sizes of papillary cystadenocarcinomas have ranged from 0.4 to 6.0 cm (mean: 2.2-2.4 cm).<sup>1,9</sup>

The malignancy of papillary cystadenocarcinomas is represented by its infiltrative growth, regional lymph node metastasis, foci of solid growth and cytologic atypia. Most of the papillary cystadenocarcinomas usually have low-grade cytological features with relatively mild pleomorphism and rare mitotic counts.<sup>1,2,8-10</sup>

Conspicuous intraluminal papillary architectures is observed in 75% of these tumors. The morphology of the papillae has been described to be quite variable from simple linear strands to complex structures.<sup>1,2,9</sup> However, the above features are not unique for papillary cystadenocarcinoma, and these cystic spaces can be observed in other salivary gland tumors such as mucoepidermoid carcinoma and acinic cell carcinoma.<sup>2,8,11</sup> In the present case, the tumor cells had no eosinophilic granules on PAS staining, and this ruled out acinic cell carcinoma.<sup>8,11</sup> The mucoepidermoid carcinomas were ruled out, because there were no characteristic combinations of mucous, intermediate, and epidermoid cells in the present case.<sup>1,8</sup>

The predominant cell populations of papillary cystadenocarcinomas are small cuboidal cells, large cuboidal cells, tall columnar cells and a mixture of these cell types.<sup>1,8,9</sup> In the present case, the tumor cells were mainly small cuboidal shape in the initial mass, but they became larger cuboidal and tall columnar cells in the recurred mass. The microscopic feature of the predominantly columnar cells was reported to be associated with high grade variants with local recurrence.<sup>10,12,13</sup> Other high grade features were the more atypical and frequent mitoses, multiple foci of necrosis, thick fibrovascular cores of papillae, solid components, and the absence of papillary growth.<sup>7,8,10,12-14</sup> Up to now, the histological criteria that allow prediction of the clinical outcome

have not been well established, and more follow up data is needed.<sup>7</sup>

In the present case, the recurred mass had higher grade features than did the initial one. The tumor cells were larger in size, more pleomorphic with more prominent nucleoli and they had brisk mitotic activity with increased layers of papillae and solid portions. Foci of comedo type necrosis were observed in the cystic portion, and foci of spindle cell proliferations were present in the surrounding soft tissue area.

The papillary cystadenocarcinomas have usually been reported to have a good prognosis.<sup>1,2,8</sup> However, a close follow-up would be necessary in this case when considering the features that suggest the high grade transformation as described above.

In conclusion, the author report here on a rare case of papillary cystadenocarcinoma arising in the retromolar area. Its histologic features were basically similar to salivary gland papillary cystadenocarcinomas of the other common sites. Although papillary cystadenocarcinomas usually show low grade features, the transformation to high grade features with clinical recurrence was peculiar in this case, and this required a close follow-up.

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