ABSTRACT

Preoperative diagnosis of malignant salivary gland tumors is difficult as radiological imaging procedures have low sensitivity rate for detecting malignancy in parotid gland tumors. With careful and detailed analysis of cytological features, guided fine needle aspiration cytology can prove to be a reliable diagnostic modality that can help in differential diagnosis of cystic parotid malignancies from cystic benign tumors and non-neoplastic lesions. Papillary cystic variant of acinic cell carcinoma is a rare, cystic, primary neoplasm of salivary gland that occurs commonly in parotid. The tumor shows high local recurrence rate and has poorer prognosis compared to classic acinic cell carcinoma. Preoperative cytological diagnosis of papillary cystic variant of acinic cell carcinoma of parotid, in the case described here was the basis for adequate and timely surgical management with good results.

Key words: Acinic cell carcinoma; cytological diagnosis; papillary cystic variant

Introduction

Papillary cystic variant of acinic cell carcinoma (ACC-PCV) occurs in younger patients as compared to the classic type, which presents in fifth decade of life. Acinic cell carcinoma (ACC) is a low grade malignancy with 5-year survival of 90% while ACC-PCV is universally fatal in 10 years. This emphasizes the need for its prompt and accurate preoperative diagnosis. Fine-needle aspiration cytology (FNAC) under radiological guidance can yield diagnostic cytology material from the predominantly cystic tumor. The report describes a case of a young male in whom preoperative cytological diagnosis helped surgeons perform total parotidectomy with prophylactic neck dissection in the same sitting.

Case Report

A 30-year-old male presented with progressively enlarging preauricular right cheek swelling for the last 4 months. He did not have any other complaints or past history of major systemic illness and was nonreactive for human immunodeficiency virus (HIV).

On local examination, the 5 cm × 4 cm swelling over the right parotid region extended from the molar region to the posterior border of the mandible and from the middle of the tragus to 0.5 cm below the right angle of the mandible. It was cystic, tense, and immobile [Figure 1a]. Facial nerve function was normal. There was no swelling on the
contralateral side. His oral examination did not reveal any abnormality.

With clinical diagnosis of benign cystic tumor favoring pleomorphic adenoma, the patient was referred for FNAC study. 6 mL of clear, yellow fluid was obtained without any regression in the size of the swelling on multiple aspirations. The aspirated fluid was centrifuged; smears were made and stained with hematoxylin-eosin, Papanicolaou, and May-Grünwald-Giemsa stains. As the smears were paucicellular and inconclusive, ultrasonography (USG)-guided FNAC was advised. Ultrasonographic finding of a heterogeneous lesion involving deep and superficial lobe of the right parotid was thought to be suggestive of pleomorphic adenoma.

Cytology smears from USG-guided aspirate showed moderate cellularity of cells that formed large monolayered sheets, small groups forming papillae and single cells in a clean hemorrhagic background [Figure 1b]. Cells had well-defined cell borders with finely vacuolated or granular cytoplasm. Nondescript small cuboidal cells and oncocytic cells were also seen. Nuclei were mostly round to oval with pale, fine granular chromatin and showed conspicuous nucleoli and mild anisokaryosis at places [Figure 2a]. No acinar structure or bare nucleus was seen. Necrotic debris or mucin was absent in smear background.

Taking into consideration the cystic nature of the swelling, clear fluid aspirate and myriad cells with bland nuclear morphology forming papillae, cytodiagnosis of ACC favoring papillary cystic variant was offered. Preoperative computed tomography (CT) scan revealed a lobulated cystic lesion in superficial portion and multiple solid moderately enhancing nodules extending in its deep lobe reaching up to parapharyngeal fat with enlarged right jugular nodes. Under general anesthesia, total parotidectomy with prophylactic neck dissection was done with preservation of the facial nerve. Lymph node dissection up to level V was carried out.

Surgical specimen sent for histopathology examination included the parotid gland with a solid cystic tumor mass removed in pieces, aggregating 5 cm × 4 cm × 2 cm in size, along with submandibular gland and cervical lymph nodes up to level V.

Sections from the tumor showed papillary cystic pattern along with solid and micro cystic areas surrounded by dense collagenous stroma and lymphocytic infiltrate. Tumor was seen infiltrating the fibrous capsule. Acinic, granular, oncocytic and small cuboidal cells were identified. Nuclei were placed eccentrically with normal nucleocytoplasmic ratio and showed bland nuclear chromatin with single nucleolus. Hobnailing of luminal cells was prominent [Figure 2b]. No atypical mitoses were identified. Mucin or necrosis was absent. There was no vascular or perineural invasion. Submandibular salivary gland was normal and none of the subcentimetric lymph nodes showed evidence of tumor metastasis. Diagnosis of papillary cystic variant of ACC was confirmed on histology.

According to American Joint Committee on Cancer (AJCC) staging criteria, the tumor stage was T3N0M0, that corresponds stage II in cancer staging. Recovery of the patient was uneventful. The oncologist did not recommend postoperative chemotherapy or radiotherapy to the patient who remains under lifelong clinical follow-up.

Discussion

ACC constitutes 2.5-4% of parotid gland tumors and papillary cystic variant accounts for one-fourth of various architectural patterns of acinic cell carcinoma (ACC) that makes it very rare. ACC-PCV occurs in young age and shows a female

![Figure 1: (a) Photograph of patient showing right preauricular swelling (b) FNAC smear showing cells in cohesive sheets and papillary fragments. (H and E, ×200)](image1.png)

![Figure 2: (a) FNAC smear showing vacuolated cells (single arrow) and cells with granular cytoplasm (double headed arrow) Inset showing cells with oncocytic cytoplasm (MGG, ×200) (b) Section of parotid gland- solid cystic tumor showing microcystic solid area with papillae. Inset showing hobnailled cells. (H and E, ×200)](image2.png)
The standard treatment of primary salivary neoplasm is positivity. These features are absent in ACC-PCV. Intracytoplasmic lumina in a mucoid background with mucin with bland nucleus and abundant vacuolated cytoplasm show in ACC-PCV. In papillary cystadenocarcinoma, tumor cells squamous, hybrid cells, and mucophages that are not seen necrotic background with the presence of malignant. Mucoepidermoid carcinoma shows dirty mucinous and papillary cystadenocarcinoma.

Interpretation of cytological diagnosis of ACC-PCV is a diagnostic challenge due to:
1. Dilution of cell content with cyst fluid,
2. Change in cell morphology due to suspension in cyst fluid,
3. Polymorphous cell population with lack of predominant cell component,
4. Cytoarchitectural difference from classic ACC.

FNAC yields hypocellular fluid from largely cystic tumor. Diagnostic material can be obtained from the solid portion of a cystic tumor under sonographic guidance. Smear shows monolayered sheets, papillary clusters, and a few single cells. Polymorphous cell population includes granular cells, vacuolated cells, and nondescript small cuboidal cells with no acinar structure and no naked nucleus. Metaplastic oncocytic and squamoid cells are often seen. A detailed and careful interpretation of the cytological findings is of utmost importance for the diagnosis of this rare tumor.

On cytology, ACC-PCV needs to be differentiated from two cystic malignant tumors, that is, mucoepidermoid carcinoma and papillary cystadenocarcinoma.

Mucoepidermoid carcinoma shows dirty mucinous and necrotic background with the presence of malignant squamous, hybrid cells, and mucophages that are not seen in ACC-PCV. In papillary cystadenocarcinoma, tumor cells with bland nucleus and abundant vacuolated cytoplasm show intracytoplasmic lumina in a mucoid background with mucin positivity. These features are absent in ACC-PCV.

The standard treatment of primary salivary neoplasm is complete surgical resection of the tumor. ACC comes under low-grade salivary gland tumor and its papillary cystic variant is high grade and universally fatal in 10 years. National Comprehensive Cancer Network (NCCN) has recommended elective neck dissection for this high-grade variant.

Conclusion

To conclude, FNAC remains the primary modality for diagnosis of papillary cystic variant of ACC. Preoperative cytological diagnosis and associated imaging features can help surgeons to plan the extent of surgical treatment as complete surgical excision is the treatment of choice for most other malignant cystic parotid tumors, ACC-PCV being a high-grade variant of ACC with poor prognosis is treated with surgical resection of tumor with prophylactic neck dissection in a single operation in young patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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