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Case Report

Pathology Section

Acinic Cell Carcinoma Papillary-Cystic Variant: Diagnostic Pitfalls in Fine Needle Aspiration Cytology

UMA KUMAR

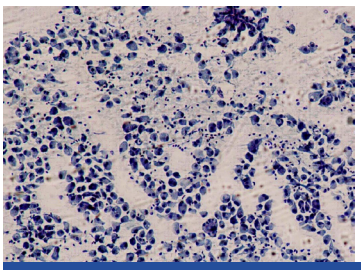
ABSTRACT

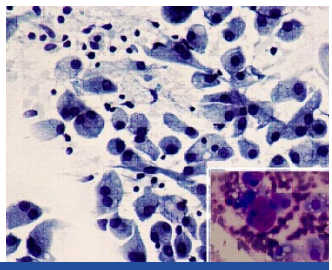
Acinic cell carcinoma is a rare tumour comprising 1%-3% of all salivary gland neoplasms. Acinic Cell Carcinoma Papillary Cystic Variant (ACC-PCV) is a distinct subtype and shows variegated appearance on cytology. It is important to differentiate it from other malignant lesions because of its poor prognosis. We describe a case of 20-year-old female with swelling on left cheek for the last four months. Fine needle aspiration was done and diagnosed as intermediate grade mucoepidermoid carcinoma on cytology. Histological study of the resected tumour showed features of ACC-PCV. We are presenting this case to illustrate the diagnostic problems encountered in cytology and important points to be kept in mind while reporting FNA of salivary gland tumours.

Keywords: Cytological diagnosis, Neoplasms, Salivary gland tumours

CASE REPORT

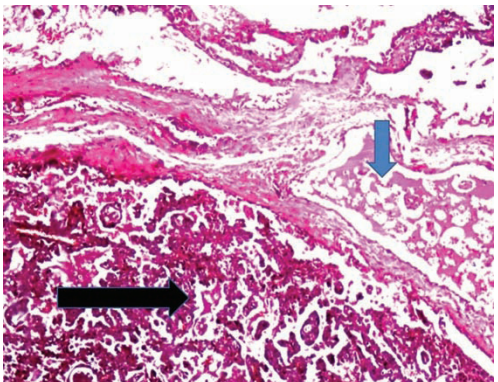
A 20-year-old female presented with a painful swelling on left cheek for the last four months. It was progressively increasing in size. There was no similar complaint in the past. On physical examination, patient was conscious, oriented and thin built, pulse- 78/min, BP-120/80mmHg and was afebrile. There was no pallor, icterus, cyanosis or pedal oedema. On left side, Level IB and Level II cervical lymph nodes were enlarged. On local examination, there was a firm swelling on left cheek extending horizontally from anterior border of ramus of mandible to 2 cm lateral to the nasal alae and vertically 4 cm from angle of mouth to the left ear lobule. It was 3 x 3 cm size, non-tender, had irregular surface, indistinct margins, firm to hard in consistency and was fixed to underlying structures. Overlying skin was not fixed. There was no swelling on the right side. A provisional diagnosis of malignant tumour with lymph node metastasis was rendered. FNA performed with a 23 gauge needle attached on a 10 ml syringe yielded 0.5 ml blood tinged fluid. Two smears were prepared directly and four smears were prepared from the button formed after centrifugation of remaining fluid at 3500 rpm for five minutes. The smears were stained with Giemsa and Papanicolaou stain. Microscopic examination revealed moderately cellular smear with numerous dissociated round and polygonal cells along with few small cell clusters. These cells had well defined cell borders and abundant amount of vacuolated cytoplasm [Table/Fig-1]. Nuclei were eccentrically placed, round to oval, with fine chromatin, inconspicuous nucleoli and mild anisokaryosis. Some of the cells showed pink staining cytoplasmic inclusions [Table/Fig-2]. Few cells with oncocytic change and occasional intermediate cells also noted. Background was haemorrhagic and no necrosis or mucin was noted. Due to the presence of large number of vacuolated cells, and intermediate cells in a haemorrhagic background, a diagnosis of intermediate grade mucoepidermoid carcinoma was rendered and histopathological confirmation was advised.

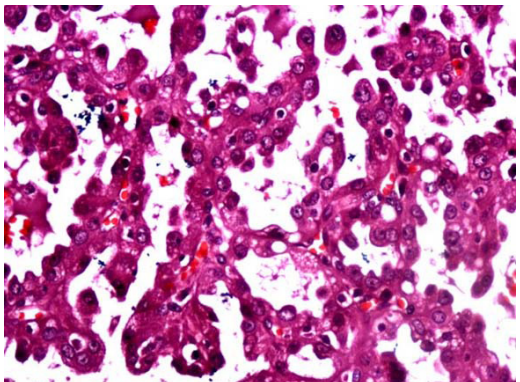


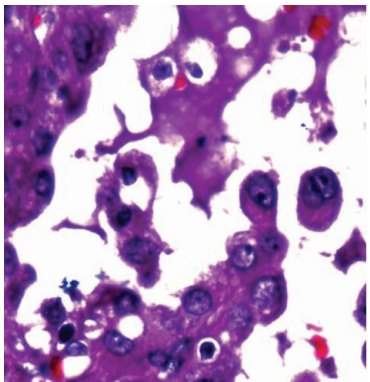


[Table/Fig-1]: Cytology smear depicts moderate cellularity with predominantly singly dispersed cells with vacuolated cytoplasm (Papanicolaou 10X).
[Table/Fig-2]: Cytology smear showing cells with well-defined borders and vacuolated cytoplasm (Papanicolaou 40X), inset shows intracytoplasmic eosinophilic inclusion (Papanicolaou 40X).

Surgical specimen sent for histopathology examination included a soft tissue mass measuring 6 x 3 x 2 cm with attached bone and teeth and tagged anterior and superior margin. Cut surface showed greyish brown tumour with solid and tiny cystic areas. Two lymph nodes were identified.







[Table/Fig-3]: Histopathology section showing tumour with papillary (black arrow) and cystic areas (Blue arrow) (H&E, 4X). **[Table/Fig-4]:** Histopathology section showing papillary projections with tombstone row appearance (H&E, 40X). **[Table/Fig-5]:** Histopathology section showing cells with round to oval cells with eccentric nuclei and small nucleoli (H&E 40X).

Haematoxylin and Eosin stained sections showed partly circumscribed tumour with cystic spaces associated with papillary projections supported by thin fibrovascular core [Table/Fig-3]. Neoplastic epithelial cells were bulging into the lumen in an uneven manner ("tombstone row" appearance) [Table/Fig-4]. The cells were polygonal to round with eccentric nucleus, small to inconspicuous nucleoli and abundant eosinophillic to vacuolated cytoplasm. Few cells showed abundant PAS positive, diastase resistant granular cytoplasm [Table/Fig-5]. Mitotic activity was minimal. Intraglandular secretions were present and stroma was hyalinized at places with chronic inflammatory infiltrate and focal haemorrhagic areas. Vascular invasion and tumour infiltration in the skeletal muscle was seen. Anterior and superior margins were also infiltrated by tumour. Normal salivary gland was seen at the periphery. Two lymph nodes were showing metastases. A histological diagnosis of papillary cystic variant of acinic cell carcinoma was made. Postoperatively the healing was uneventful and there was no sign of recurrence after six months of follow up.

DISCUSSION

Acinic cell carcinoma is a low grade, well differentiated neoplasm comprising 1%-3% of all salivary gland tumours [1]. The most common site of involvement is parotid (86%), occasionally submaxillary gland, minor salivary glands and rarely extra-salivary sites like breast, lungs and pancreas [2]. ACC-PCV is a distinct subtype and accounts for one fourth of acinic cell carcinoma [3]. It has been mostly reported to occur in younger patients (16-40 years) as compared to the classic ACC that characteristically presents in fifth decade of life [2]. Females are more commonly involved. ACC is a slow growing tumour with excellent prognosis and has a five year survival of 90%. However, ACC-PCV has poor prognosis and is universally fatal in ten years [2,3].

FNAC features of ACC are well documented. Smears show loosely clustered and acinic group of cells with bland morphology. Background shows bare tumour nuclei that differentiate it from normal salivary gland [4]. Due to various subtypes of ACC (classic, microcystic, follicular, papillary and papillary cystic), it is very difficult to diagnose on cytology [1]. Cytological diagnosis of ACC-PCV poses a diagnostic difficulty due to its polymorphous appearance, which may overlap with a number of benign and primary malignant processes including metastatic cancer [3].

In a study conducted by Ali SZ et al., at the John Hopkins Hospital, seven cases of ACC-PCV variant, confirmed on histopathology were retrospectively reviewed with regard to their FNA diagnosis. In none of those cases, a diagnosis of ACC-PCV was either made or suggested on the FNA picture. MEC was given as the diagnosis in two of the cases as neoplastic cells displayed large vacuoles resembling intracytoplasmic mucin vacuoles, similar to our case [5].

Author suggested following points to keep in mind – tightly cohesive fragments of neoplastic epithelium seen as monolayered sheets or with the prominent papillary architecture, high N:C ratio, ductal type epithelium, cystic material and degenerated cellular debris, histiocytes, cells with squamoid and metaplastic oncocytic change, vacuolated and pigmented histiocyte-like tumour cells and lack of a predominant single cell component or naked neoplastic cell nuclei [5].

Mosunjac MB et al., suggested few red flags that should prompt a pathologist to further investigate, like presence of tight cellular clusters, distinct cytoplasmic borders, large nuclei with distinct nucleoli and binucleated cells [6]. In our case, smear was mainly composed of dyscohesive singly scattered cells, with only few epithelial cell clusters. However, when cytological slides were reviewed, occasional binucleated cell and bare nuclei in the background were noted, but there was no papillary cluster. The cells had abundant amount of vacuolated cytoplasm with few showing coarse granular cytoplasm which were misdiagnosed as mucin secreting cells. In ACC-PCV, cystic fluid and vacuolated cells can mimic mucin and mucin secreting cells, so can be misleading. A mucin stain is helpful in making a correct diagnosis, which was not done in our case. ACC-PCV may undergo metaplastic oncocytic or squamoid changes which could be focal or confluent [4].

In a study by Shet T et al., six cases of ACC-PCV were analysed and it was found that diagnosis on FNA significantly differed from the histologic diagnosis. ACC-PCV was misdiagnosed as MEC due to the presence of abundant vacuolated cells, like in our case [7]. Several studies have shown that acinic cell carcinoma is one of the major culprits when it comes to a false-negative misinterpretation of a salivary gland FNA [8]. Preoperative cytological diagnosis can help surgeons to plan management of the patient.

CONCLUSION

ACC-PCV is a rare tumour of salivary gland, with poor prognosis. Cytopathologist should be aware of the variegated appearance of the tumour. This case highlights the importance of detailed cytological features on fine needle aspiration cytology.

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PARTICULARS OF CONTRIBUTORS:
1. Specialist Pathology, Department of Pathology, Veer Savarkar Aarogya Sansthan, Karawal Nagar, Delhi, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Uma Kumar,
C-7/14, Sector-8, Rohini, Delhi-110085, India.
E-mail: dr.uma.path@gmail.com

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