CASE REPORT

Papillary cystic variant of the acinic cell adenocarcinoma

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Summary The papillary cystic variant of the acinic cell adenocarcinoma is a rare salivary gland tumour, which may behave in an unpredictable manner and exhibit non-specific histological features. Its diagnosis is important as it may prognosticate for a poorer outcome and it has been reported to universally fatal. We present a case of this unusual tumour and discuss the diagnostic and management difficulties.

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KEYWORDS
Acinic cell carcinoma; Papillary cystic variant; Histology; Prognosis

Introduction

Acinic cell adenocarcinoma is an uncommon tumour of salivary glands with unusual clinical and histopathological features. The papillary cystic variant is especially uncommon and its diagnosis has therapeutic and prognostic significance.

Case report

A 31 year old Filipino patient was referred with a lump at the right angle of mandible. There was recent painless enlargement. She had undergone an operation for a parotid lump in the Philippines six years previously and developed a recurrent lump three months after surgery. She was not aware of the nature of the procedure or the diagnosis, and had not been followed up.

Clinical examination revealed a 3 cm well circumscribed, semi-mobile, painless bilobed mass at the right angle of the mandible. Facial nerve function was intact and there was no lymphadenopathy or pharyngeal extension.

Her medical records from the Philippines were obtained and showed she had undergone a lumpectomy for a pleomorphic salivary adenoma (PSA). The clinical diagnosis of a recurrent PSA was presumed, and an FNAC and CTS were arranged prior to surgery.

Cytology showed cells with prominent nucleoli and nuclear pleomorphism and the possibility of a second unrelated primary of a mucoepidermoid...
carcinoma or papillary adenocarcinoma was suggested.

CTS demonstrated a bilobulated enhancing mass inferior to the parotid gland residue with no evidence of lymphadenopathy (Fig. 1).

At surgery an encapsulated mass was found and a completion superficial parotidectomy with preservation of the facial nerve was performed. Her post-operative recovery was uneventful.

Histologically, the tumour was generally well circumscribed and surrounded by compressed fibrous tissue (Fig. 2). There was infiltration and lymphatic invasion. The parenchyma had a mixed pattern, and nuclear pleomorphism and mitoses

Figure 1  CTS illustrating the bilobed mass in the right parotid gland.

Figure 2  Illustrating the tumour at low power.
were present. In places there was prominent follicular arrangement comprising multiple cystic lumina filled with a eosinophilic proteinaceous material and lined by intercalated duct like cells. Elsewhere there were large cysts containing papillary projections with tombstoning of the epithelial lining (Fig. 3). The differential diagnosis of a secondary papillary or follicular thyroid cancer was excluded by immunostaining for thyroglobulin.

The histological findings were unexpected and the original specimen slides were obtained for comparison. This showed a tumour with pleomorphic morphology but with features similar to the second specimen. The original diagnosis was therefore revised and a final diagnosis of papillary cystic variant of acinic cell adenocarcinoma with recurrence was concluded.

The patient received post-operative radiotherapy.

Discussion

Acinic cell adenocarcinomas comprise 2.5% of all salivary gland tumours and 12–17% of all salivary gland cancers, of which more than 92% occur in the parotid gland. There is with the exception of the latter series, a female predilection and the median age is in the fifth decade. The tumour was originally regarded as benign but Buxton et al. were first to recognise its malignant potential. It frequently exhibits malignant histological features and has been reclassified on this basis. This is consistent with its clinical behaviour.

It commonly presents as a painless slow growing rubbery mass, with fixation and facial nerve involvement developing in advanced cases. An unusual feature is the occurrence of synchronous or metachronous tumours in the contralateral gland and when this is considered with its other clinical features it can be misdiagnosed for the commoner Warthins tumour or PSA.

Histologically it may exhibit cellular pleomorphism and differentiation from benign tumours is essential. The tumours are encapsulated and features of capsular invasion are not uncommon. Nuclear pleomorphism, cellular atypia and mitoses may be present, and predict a poorer outcome. The tumours may be solid or cystic and distinct morphological growth patterns are seen. These are described as solid, microcystic, follicular and papillary cystic.

Cells resembling serous acinar and intercalated ducts are typical. The papillary cystic variant maybe differentiated by the presence of prominent follicular arrangement, papillary projections, multiple cystic luminae and tombstoning.

The five year survival rate for acinic cell carcinomas is 90% but according to Spiro et al. the papillary cystic variant should be conferred significance because it has a poorer prognosis. In this series there were no survivors at 10 years.

Prolonged disease progression, local recurrence (35%), nodal (16%), distant and haematogenous spread have been reported and adjuvant radiotherapy should be considered. Survival in acinic cell carcinoma is generally better in younger patients but the outcome is less favourable when patients present with pain or facial nerve dysfunction.
The diagnosis of the papillary cystic variant of the acinic cell adenocarcinoma has significant implications. It is uncommon. It may not be considered, or may be misdiagnosed clinically on the basis of relatively benign characteristics, or when present bilaterally. It may be misdiagnosed cytologically and may thus be accorded less significance and managed conservatively as Warthin’s and PSA tumours sometimes are, or excised by less radical methods with narrower margins. Lumpectomy or extra-capsular dissection procedures may not be suitable methods. Histologically they may be misdiagnosed for a benign tumour and this will clearly have both therapeutic and prognostic consequences. Its diagnosis will also have implications for the pattern of follow up as very late recurrences up to 30 years on have been reported, and the possible development of a further tumour in contralateral gland should always be considered. When local recurrences do occur additional local surgery can provide good control.

In summary the acinic cell carcinoma is a rare malignant tumour which may exhibit unusual and prolonged disease progression and whose prognosis is dependent on clinical stage and the completion of excision. Diagnosis maybe difficult as clinical, cytological and histological features maybe ambivalent. Prognosis of the papillary-cystic variant has been reported to be universally fatal within 10 years.

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References


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