Short Communication

Papillary cystadenocarcinoma of the sublingual gland

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ABSTRACT

A 68-year-old Caucasian gentleman presented with a 6-month history of a left sided Level I/II neck swelling involving the floor of mouth. MRI revealed a large cystic lesion and histology confirmed a diagnosis of primary papillary cystadenocarcinoma of the sublingual gland.

Papillary cystadenocarcinoma was first described in 1991 by the World Health Organisation [WHO], and is a rare malignant neoplasm characterised by cysts and papillary endo-cystic projections.

Papillary cystadenocarcinoma arising from the sublingual glands is extremely rare and has the potential to metastasise to cervical lymph nodes. This patient we report was therefore treated with surgical excision and post-operative radiotherapy.

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1. Introduction

Papillary cystadenocarcinoma was first described in 1991 by WHO, it is an extremely rare tumour, and has been described as a low-grade malignant neoplasm characterised by cysts and papillary endo-cystic projections.¹ It has previously been classified as malignant papillary cystadenoma, low grade papillary adenocarcinoma or mucus producing adenopapillary carcinoma² but in 2005 was classified as cystadenocarcinoma in the WHO classification of head and neck tumours.³

Papillary cystadenocarcinoma has been seen in the gallbladder, bile duct, pancreas, mammary gland, thyroid gland, and upper respiratory tract but is rarely seen in salivary gland parenchyma and accounts for less than 0.2% of salivary gland tumours.⁴

2. Case Report

A 68-year-old Caucasian gentleman presented with a 6-month history of a gradually enlarging neck swelling in the left submandibular region. He was otherwise locally asymptomatic with a normal swallow, good appetite and stable weight. He did however complain of excessive sweating and unexplained lethargy.

He had previously undergone a tonsillectomy and PMH included hypertension and severe osteoarthritis resulting in

wheel-chair mobility. He was a non-smoker with minimal alcohol consumption.

Examination revealed a cystic swelling of the left floor of mouth but with normal mouth opening and tongue projection. Neck examination revealed an isolated large submental/submandibular cystic mass only in communication with the oral findings, there was no other discrete lymphadenopathy palpable and remaining ENT examination including flexible nasendoscopy was unremarkable.

FNAC was performed on presentation with cytological findings consistent of a cystic lesion. MRI imaging revealed a large cystic mass enhancing on STIR sequencing [Fig. 1], features were initially suggestive of a lymphatic malformation. Considering the radiological findings and the cystic nature of this mass, a lymphangioma was considered the most likely diagnosis pre-operatively. Whilst haemangiomas and venous malformations may have similar radiological appearances these were discounted on cytological findings.

Given the uncertainty of definitive diagnosis and the size of the tumour, surgery was offered to the patient after multi-disciplinary team discussion.

Surgical excision was performed through the neck; tumour, submandibular gland and floor of mouth were removed to ensure complete removal. The floor of mouth defect was repaired with primary closure and naso-gastric tube insertion was performed peri-operatively. Post-operatively the patient was kept nil-by-mouth for five days before reinstating diet. This was well tolerated and the patient made an uncomplicated recovery. Given pre-operative findings with the expectation of a benign lesion and no

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Fig. 1. STIR weighted MRI demonstrating a multi-cystic mass in the left sublingual region.

Fig. 2. Histological slide consistent with papillary cystadenocarcinoma: [a] tumour extending close to salivary gland parenchyma [b] cystic area involving margin [c] complex papillary intraluminal projections.

cervical lymphadenopathy on imaging neck dissection was not performed.

Histology of the lesion [Fig. 2] showed complete excision of a $55 \times 60 \times 30$ mm infiltrative multicystic neoplasm arising from salivary parenchyma with normal submandibular parenchyma. Primary papillary cystadenocarcinoma was suspected based on the histological features of irregular shaped cysts lined with both cuboidal and tall columnar cells and the formation of complex papillary intraluminal projections. Given the rarity of this tumour, second opinion was sought and a diagnosis of primary papillary cystadenocarcinoma of the sublingual gland was confirmed.

3. Discussion

Given the low-grade nature of papillary cystadenocarcinomas, most patients will present with a slow growing and asymptomatic mass,⁵ but there have been several reports of more aggressive tumours with high-grade pathological features and the potential of cervical lymph node metastases.^{4,6}

There are however, no defined parameters for grading papillary cystadenocarcinomas due to the small number of cases reported. Conservative but complete surgical excision is thought to be the treatment of choice in papillary cystadenocarcinomas of the parotid but there is no defined consensus on the other major salivary glands.⁷

Foss et al. discuss the largest case series of salivary gland cystadenocarcinomas, with 57 reports, only two of these cases were found to be tumours arising from the sublingual glands.⁸ Macroscopically, all lesions were found to be cystic or multicystic masses and ranged in size from 0.4 to 6.0 cm. Microscopically, all tumors demonstrated an invasive, cystic growth pattern, and 75% had a conspicuous papillary component.⁸ This case therefore represents one of the rarest and also largest presentations of salivary gland cystadenocarcinoma.

4. Conclusions

Cystadenocarcinoma presents as a rare but interesting differential diagnosis for Lymphatic malformations in the Head and Neck clinic. We report an extremely rare case of papillary cystadenocarcinoma arising from the sublingual gland with exact pathophysiological prognosis unknown.

Although post-operative staging CT [Fig. 3] showed satisfactory surgical clearance with no evidence of cervical or mediastinal lymphadenopathy, the patient was discussed extensively at the head and neck MDT, and will receive post-operative radiotherapy

Fig. 3. Post-operative axial CT showing satisfactory tumour excision.

given the histological findings, large tumour size and close peripheral margins.

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