Mucoepidermoid carcinoma - unknown primary and late distant metastasis: an unusual course of the disease

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Abstract

The authors report the case to understand this unusual presentation and prognosis of mucoepidermoid carcinoma following treatment. We present a case of mucoepidermoid carcinoma in a 67-year-old man. The cancer was diagnosed in the right side of his neck but the primary tumor remained unknown despite attempts at staging. The neck was treated successfully and followed up for 2 years. Metastasis of the primary lesion to the left triceps was diagnosed following a swelling noticed by the patient during the 2-year follow up period. Mucoepidermoid carcinoma can be a low, intermediate or high-grade malignancy and can metastasize to different parts of the body. However, this is the first case report of a metastasis to the triceps muscle.

Introduction

Mucoepidermoid carcinoma is the most common salivary gland malignancy[1] and most of these tumor arise from the minor salivary glands especially in the palate. It is typically composed of varying amounts of epidermoid cells, intermediate cells and mucous producing cells. Mucoepidermoid carcinoma is believed to arise from the reserve cells of excretory ducts. It displays a variety of biological behaviour depending on the differentiation of the tumor cells. High grade tumors are generally considered to be highly aggressive while low grade ones behave more like a benign tumor.[2]

Case Report

A 67-year-old man presented with a painless right sided neck swelling which had gradually increased in size over the preceding 12 months. The patient had not sought medical advice sooner as the swelling did not bother him. He had no aerodigestive or systemic symptoms. The patient had smoked 20 cigarettes a day since the age of 16.

On initial examination there was a 6 cm firm mass occupying most of Level II on the right side of the neck. There were no skin changes over the mass and no other palpable lymph nodes. There was no abnormality on examination of the upper aerodigestive tract. Blood results showed a raised Ca 19-9 of 899 and a mildly raised CEA of 6. A computed tomography (CT) scan showed a number of enlarged right sided Level V lymph nodes. FNA cytology was non – diagnostic and the patient was listed for panendoscopy and biopsy of right neck mass, as per the diagnostic protocol in place at the time.

Samples obtained from the panendoscopy showed no dysplasia or malignancy. However, the biopsy of the right neck mass showed fibrous areas containing scanty invasive fragments of low grade adenocarcinoma composed of fairly uniform intermediate cells and scattered mucin containing cells, suggestive of mucoepidermoid carcinoma (Figures 1 and 2).

After discussion at the Multi Disciplinary Clinic, the patient underwent a right Radical Neck Dissection for extensive Mucoepidermoid Carcinoma lymphadenopathy of unknown primary. Post- operatively he received radiotherapy of 60 Gray over six weeks to the right neck from the skull-base to the clavicle. Following this the patient was reviewed monthly for 12 months and then 2 monthly. He continued to smoke 20 cigarettes daily.

Twenty-three months following his original referral the patient was urgently re-referred to the clinic by his general practitioner (GP) with a suspicious mass in his contralateral left arm. There was no history of trauma. On examination the patient had an approximately 3.5×2.5 cm lesion in his left arm in the region of the triceps. The mass was smooth and non – tender. A magnetic resonance imaging (MRI) scan was reported as showing the lesion was suspicious of malignancy. A CT scan of the neck at this time demonstrated two small areas suspicious of recurrence within the neck, the larger of which measured 2 cm in diameter (Figure 3). The CT also noted partial collapse of both T11 and T12. FNA cytology of the neck lesions confirmed recurrent Mucoepidermoid Carcinoma and biopsy of the triceps lesion demonstrated metastatic spread (Figure 4). An isotope bone scan showed changes in the right sacro-iliac joint which were then confirmed on an MRI scan. This patient had developed multiple recurrences involving the triceps, sacro-iliac joint and thoracic spine, as well as the neck. The patient continues to have palliative care.

Discussion

Mucoepidermoid carcinoma (MEC) is the commonest malignancy of the salivary glands, with approximately 50% of primary tumors affecting the minor salivary glands.[1] The tumor displays a variety of biological behaviour. The differentiation of the tumor can be low, intermediate or high grade. Low-grade tumors gen-

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Key words: mucoepidermoid, metastasis.

Received for publication: 1 October 2011.
Accepted for publication: 14 October 2011.

Clinics and Practice 2011; 1:e97
doi:10.4081/cp.2011.e97

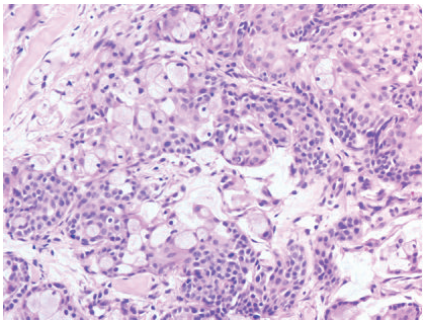

Figure 1. Low grade mucoepidermoid carcinoma showing cystic areas with mucous, intermediate and squamous cells. Well-differentiated mucinous cells predominate and mitoses are sparse.

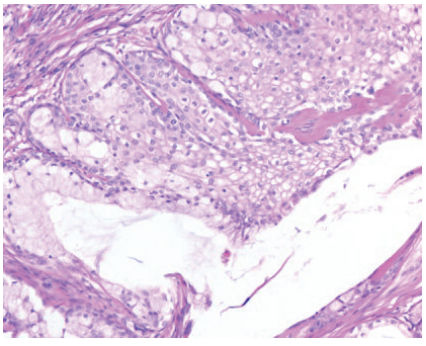

Figure 2. Right neck core biopsy showing recurrent/metastatic mucoepidermoid carcinoma with similar appearances to that seen in original resection specimen. Mucinous cells, intermediate cells and squamous cells are present.

erally behave like a benign tumor while the high grade tumors are aggressive in nature.[2]

Standard treatment for Mucoepidermoid carcinoma is surgical excision.[2] MEC has been considered by some to be a radio-resistant tumor, although postoperative radiation has been effective in some circumstances. MEC is often difficult to diagnose and grade from fine needle cytology or open biopsy.[3,4] Hence it is often misdiagnosed as a benign tumor preoperatively and is often removed with an insufficient margin. This results in a close or positive margin which is associated with poor prognosis.[5-7]

Although MEC has been considered a radio-resistant tumor, treating cases with a positive surgical margin with post – operative radiotherapy has been reported to decrease local failure.[8,9]

This case was managed by surgical excision and post-operative radiotherapy. Although adequate local control of the neck disease was achieved initially, a metastatic lesion was diagnosed in the triceps muscle, as well as the neck and skeleton. The presentation was unusual both because no primary lesion was found and also because to our knowledge this is the first reported case of MEC metastasis to the triceps muscle.

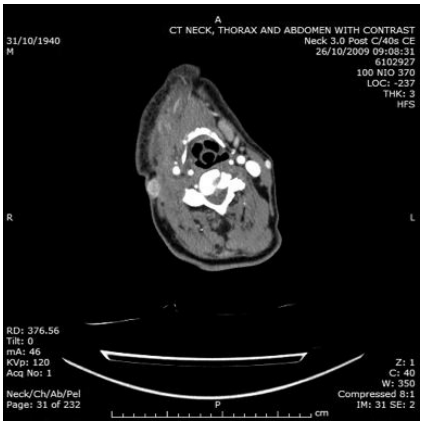

Figure 3. Loss of the tissue planes on the right side of the neck consistent with previous surgery and an enhancing subcutaneous nodule in the right side of the neck consistent with local tumor recurrence.

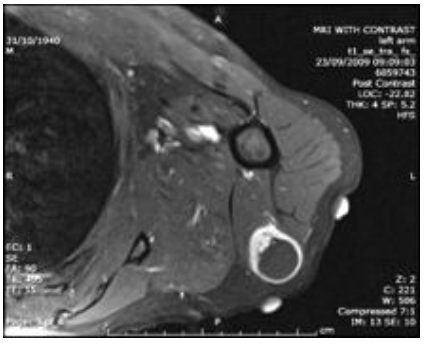

Figure 4. The lump correlates to a 3.5 cm oval soft tissue mass related to the posterior deltoid and the posterior triceps muscle. The appearances suggested this actually involves and invades the triceps muscle. There is enhancement peripherally with a central tumor nodule. The appearances are most likely due to a metastatic lesion.

In a retrospective clinical analysis of MEC from the Japanese Journal of Clinical Oncology; 28 of the 43 patients had a parotid primary and 3 had a primary in the submandibular gland.[2] Of the remainder there were 5 oral, 4 oropharyngeal and there were single cases of primaries in the larynx, maxillary sinus and epipharynx respectively. There were no cases in which a primary tumor was not found, unlike in the patient discussed in this case report. In the same study distant metastases were observed in the lung, liver and brain.[10] There were no cases reported in the study where metastasis to skeletal muscle was observed.

In a separate retrospective case series, it was found that a parotid primary was commonest, being found in 9 out of 12 cases with one each in the sublingual gland, hard palate and neck.[11] Of 6 cases with follow up information, 3 had distant metastases to skin and bone. As in the Japanese study there were no cases of metastasis to the skeletal muscle and no reported cases with an unidentified primary tumor.

In the literature the lungs, liver and brain[11] are reported as common site of metastases and there are a handful of reported cases of metastases to the skin[11] and skin primaries.[10] Solitary skull metastasis form mucoepidermoid tumor has been reported by Sousa *et al.* in 2001.[12]

Grade I tumors are considered low grade and have a very low recurrence rate and almost 100% survival. Grade II and III do considerably worse, with recurrence rates around 30% and 70% respectively. Survival rate decreases to 70% and 40% for grades II and III respectively[13]. Although these are the published numbers regarding prognosis of these cases, patients need to be warned about unusual presentation of the tumor.

Conclusions

i) Mucoepidermoid carcinoma of unknown origin is a rare clinical presentation; ii) Metastasis of this lesion to triceps has not been reported in the literature; iii) Patient advice regarding the prognosis of these tumors is not easy!

References

1. Goode RK, El-Naggar AK. Mucoepidermoid carcinoma (Head and Neck). In: Barnes L, Eveson JW, Reichart P, Sidransky D, eds. Pathology and genetics of Head and Neck Tumours. World Health Organization Classification of Tumours. Lyon: IARC press; 2005. pp. 219-220.
2. Ozawa H, Tomita T, Sakamoto K, et al. Mucoepidermoid Carcinoma of the Head and Neck: Clinical Analysis of 43 patients. Jpn J Clin Oncol 2008;38:414-8.
3. Brandwein MS, Ivanov K, Wallace DI, et al. Mucoepidermoid carcinoma: a clinicopathologic study of 80 patients with special reference to histological grading. Am J Surg Pathol 2001;25:835-45.
4. Kumar N, Kapila K, Verma K. Fine needle aspiration cytology of mucoepidermoid carcinoma. A diagnostic problem. Acta Cytol 1991;35:357-9.
5. Guzzo M, Andreola S, Sirizzotti G, Cantu G. Mucoepidermoid carcinoma of the salivary glands: clinicopathologic review of 108 patients treated at the National Cancer Institute of Milan. Ann Surg Oncol 2002;9: 688-95.
6. Carrillo JF, Vázquez R, Ramírez-Ortega MC, et al. Multivariate prediction of the probability of recurrence in patients with carcinoma of the parotid gland. Cancer 2007; 109:2043-51.
7. Healey WV, Perzin KH, Smith L. Mucoepider-moid carcinoma of salivary gland origin. Classification, clinical-pathologic correlation, and results of treatment. Cancer 1970;26:368-88.
8. Rapidis AD, Givalos N, Gakiopoulou H, et al. Mucoepidermoid carcinoma of the salivary glands. Review of the literature and clinicopathological analysis of 18 patients. Oral Oncol 2007;43:130-6.
9. Hosokawa Y, Shirato H, Kagei K, et al. Role of radiotherapy for mucoepidermoid carcinoma of salivary gland. Oral Oncol 1999; 35:105-11.
10. Suárez-Peñaranda JM, Vieites B, Valeiras E, Varela-Duran J. Primary Mucoepidermoid of the Skin expressing p63. Am J Dermatopathol 2010;32:61-4.
11. Weinreb I, Seethala RR, Perez-Ordoñez, et al. Oncocytic Mucoepidermoid Carcinoma Clinicopathologic Description in a Series of 12 Cases. Am J Surg Pathol 2009;33:409-16.
12. Sousa J, Sharma RR, Pawar SJ, et al. Solitary skull metastasis from mucoepidermoid mimicking a parotid tumour. Neurol India 2001;49:194-6.
13. Emerick KS, Fabian RL, Deschler DG. Clinical presentation, management, and outcome of high-grade mucoepidermoid carcinoma of the parotid gland. Otolaryngol Head Neck Surg 2007;136:783-7.