CASE REPORT

Primary Cutaneous Mucoepidermoid Carcinoma of Thigh

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ABSTRACT

Mucoepidermoid carcinoma (MEC) of the skin is an exceedingly rare but distinct neoplasm with respect to its histopathological features. It is similar, if not identical to MEC of the salivary gland, a neoplasm whose prognosis is correlated with the pathologic grade. We present two cases of primary cutaneous mucoepidermoid carcinomas, presenting as a cystic lesion of the thigh. No other tumors were found in the thoroughly examined patients. Histologically both these cases had the characteristics of Primary cutaneous MEC. This is a very rare presentation of a non salivary gland MEC.

Keyword: Mucoepidermoid carcinoma, cystic lesion, thigh

INTRODUCTION

Mucoepidermoid carcinoma is the most common primary malignant salivary gland tumor. It accounts for about 15% of all *salivary* gland tumors[1]. It was first described in 1945 and was thought to occur only in salivary glands[2].Later studies showed that these tumors can also occur in the lacrimal sac[3], bronchus[4], pleura[5], jaw[6] and skin[7].The tumor is divided into low and high grade depending on its histopathological grading. Although distant metastasis is rare but reports of metastasis to regional lymph nodes, lungs and distant organs have been documented in literature. We report 2 cases of primary cutaneous Mucoepidermoid Carcinoma, both presenting as a subcutaneous cystic lesion of thigh.

Case 1

A 55 year old female was referred to the medical oncology department with a large cystic lesion of the right thigh. She noted the swelling 8 months prior to presentation. It was initially small in size but progressively increased over the past few months. The swelling was not associated with pain or fever, but occasionally caused some discomfort which was relieved by oral analgesics. There were no other local or systemic symptoms. She had a medical history of hypertension which was controlled with anti-hypertensives. Her social history showed that she was a nonsmoker and a teetotaler. She was postmenopausal for 10 years with 5 healthy children. Further she had no family history of cancer.

On examination, there was a painless cystic swelling on the lateral aspect of the right thigh which measured 6x5 cm. It was well circumscribed, soft and cystic in consistency and was adherent to the overlying skin. However it was found to be freely mobile over the underlying structures. There were no palpable lymph nodes or organomegaly. Examination of head and neck was unremarkable. No X-Rays or Scans were performed prior to presentation.

This patient underwent excision biopsy of the lesion. Microscopy revealed a thick fibrotic cyst wall. Predominantly the cyst lining was hemorrhagic with several foci of inflammation. Focally there were foci of low grade mucoepidermoid carcinoma comprising of cribriform and papillary patterns. Tumor cells were squamoid with abundant cytoplasm and mild pleomorphism. Some tumor cells showed cytoplasmic and luminal mucin, which tested positive with mucicarmine stain. All these features were consistent with low grade MEC.

Case 2

A 21 year old primigravida female presented with a swelling on the left thigh which she noticed to be rapidly increasing in size along with grey-white discoloration of the overlying skin. The swelling was not associated with any pain, fever or any history of trauma. The patient was pregnant for 16 weeks. She had no co- morbidities and did not have any family history of cancer.

On examination, there was a cystic swelling on the anterior aspect of left thigh measuring 7cm x 5 cm. The overlying skin was adherent to the mass and showed grey-white discoloration. The mass was non-tender, cystic in consistency and had ill-defined margins. No local lymphadenopathy or organomegaly was found. Her systemic survey and physical examination were unremarkable.

Her excision biopsy was performed. Microscopic examination revealed a cystic neoplasm present in the dermis of the skin. It was composed of squamoid cells with abundant eosinophilic to clear cytoplasm with focal areas of mucinous differentiation. PASD and mucicarmine stain was positive for mucin. Few

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mitotic figures were also noted. The tumor had infiltrative margins. These findings were consistent with a Cutaneous Mucoepidermoid Carcinoma with intermediate to high grade features.

Her follow-up re-excision was performed and metastatic workup was also done. The lesion was excised and histological confirmation of tumor free margins was achieved. She was not subjected to any post-op chemo/radiotherapy.

Fig 1. Low power magnification of specimen

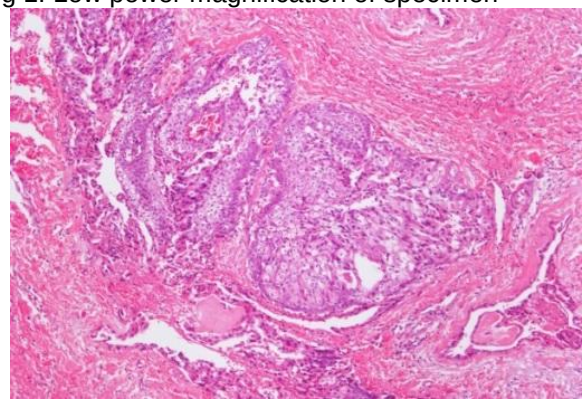
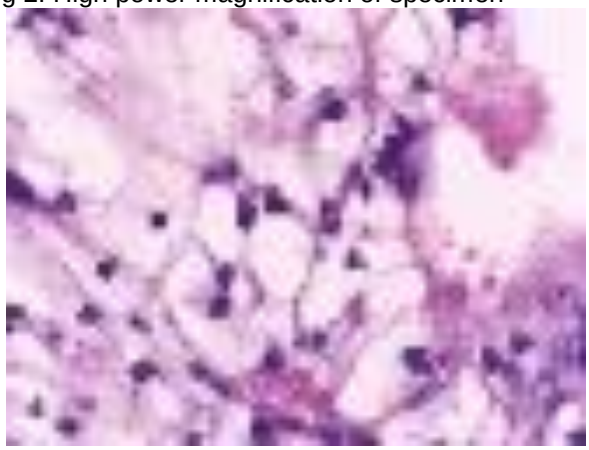


Fig 2. High power magnification of specimen



DISCUSSION

Mucoepidermoid carcinomas are malignant tumors commonly arising from the salivary glands. They make up to 15% of all salivary gland tumors[1], out of which 70% arise from the parotid gland. Other unusual sites of presentation include conjunctiva, lacrimal sac, bronchus, mandible and skin. They usually present as an asymptomatic lump but sometimes may be associated with pain which is an indication of a higher tumor grade. Both lymphatic and hematologic metastases have been demonstrated. The usual sites of metastases are the regional lymph nodes (48%), lung (25%), bone marrow (25%), distant lymph nodes (18%) and adrenal gland (18%). Few cases have been reported of metastasis to the kidney, pleura, pericardium and mediastinum[8,9,10]

Mucoepidermoid carcinomas commonly occur in adults; however, a significant number of cases in children have also been reported. They have a slight female predilection. Histologically these tumors are composed of squamoid cells, mucin producing cells or intermediate cells. They are generally divided into 3 histological grades[11].

Low Grade: When there is presence of well-formed glandular structures or micro-cysts lined by a single layer of mucinous columnar cells. Occasional papillary or cribriform structures maybe encountered.

Intermediate Grade: Presence of solid nests of squamoid cells with intermediate basaloid cells. Papillary and cystic infoldings are also present.

High Grade: The tumor predominantly comprises of solid nests and cords of tumor cells which maybe squamoid or basaloid. There is prominent nuclear pleomorphism. The cystic component is less than 20% of the entire tumor. There is presence of more mitotic figures (usually more than 4/10HPF), necrosis and perineural invasion.

The clinical course and prognosis depends upon the grade of the neoplasm. Low-grade tumors may invade locally and recur in about 15% of the cases, but only rarely do they metastasize and so yield a 5-year survival rate of more than 90%. By contrast, high grade neoplasms and, to a somewhat lesser extent, intermediate grade tumors are invasive and difficult to excise and so recur in about 25% to 30% of cases and, in 30% of cases, disseminate to distant sites, The 5-year survival rate of these high grade tumors is only 50%[12].

Both of the above mentioned cases presented to us with a cystic swelling of thigh and both were female patients. The tumor with a higher histopathological grade showed more rapid increase in size and with infiltrative margins. While the low grade tumor showed a slow growing pattern it is highly possible that both were secondary to primary salivary gland tumors; however our investigation did not reveal any salivary gland lesion or any other evidence of a primary origin. Sometimes histologically cutaneous variant of MEC can be confused with adeno-squamous carcinoma of the skin. The histological features of mucigenic cell positivity with mucin stain is helpful in distinguishing both of them.

The primary treatment of this form of cancer is an adequate surgical excision. High grade tumors are difficult to excise completely as they tend to infiltrate the surrounding tissue. Hence, the recurrent rate of such tumors is around 30% and 5 years survival rate is 50%.

CONCLUSION

Primary cutaneous mucoepidermoid carcinoma is a very rare entity. It can arise at any part of the body with or without apparent involvement of the over lying skin. Therefore, we should have a high index of suspicion when dealing with such unusual presentations of tumors having adenoid and squamoid components. Cautious histological evaluation, wide surgical excision, metastatic workup and close follow-up of the patients is essential owing to dismal prognosis of high grade mucoepidermoid carcinomas.

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