CASE REPORT

A malignant eccrine spiradenoma of the scalp

Whitney Chow, Mat Griffiths

St Andrew's Centre for Plastic Surgery and Burns, Mid Essex Hospital Services NHS Trust, Essex, UK

Correspondence to

Whitney Chow,
chowwhitney@yahoo.com

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SUMMARY

Malignant eccrine spiradenoma (MES) is an extremely rare but aggressive tumour, resulting in high mortality. We report the first case in the UK of metastatic MES on the scalp of a 37-year-old man who initially presented with a raised nodular mass which had grown rapidly over a few months. Excision biopsy of the lesion was performed and histology confirmed MES with evidence of cytological atypia and necrosis. A further wide local excision of the lesion with a 1 cm surgical margin was performed and full clearance was achieved. At 10-month follow-up another similar lesion had developed close to the previously excised area. A CT scan demonstrated metastatic lesions in the lungs, liver and pelvis, and subsequently chemotherapy was initiated. Although MES is uncommon, it has a high propensity to metastasise, therefore prompt identification, treatment and close follow-up of these tumours is essential.

BACKGROUND

Malignant eccrine spiradenoma (MES) is a very rare tumour, especially when arising from the scalp. We describe the first in the UK of a recurrent MES affecting the scalp with multiple lung, pelvis and liver metastases.

CASE PRESENTATION

A 37-year-old man presented to the plastic surgery department with a lesion on his scalp which arose a few months previously. It started initially as a raised nodule <1 cm in size but rapidly grew to 6×4 cm within a month. It did not bleed but was occasionally itchy and painful. The patient did not recall any preceding trauma to the scalp. There was no history of skin cancer or significant sun exposure. The patient's only medical history was a digital papillary adenocarcinoma which was excised from his finger 10 years previously.

Clinical examination revealed a large, non-ulcerated, well-defined and purplish nodule on the right side of the parietal scalp (figure 1). The lesion appeared to be mobile over underlying bone. Lymph node examination was unremarkable.

INVESTIGATIONS

An incisional biopsy of the lesion was performed; histology initially suggested a benign eccrine spiradenoma with no atypia and necrosis but an exceptionally high-proliferation rate as evidenced by elevated expression of Ki-67. Further histological analysis revealed the diagnosis to be an MES with evidence of cytological atypia and necrosis (figure 2).

Figure 1 A large non-ulcerated well-defined purplish nodule on the right side of the parietal scalp.

TREATMENT

A multidisciplinary team (MDT) meeting recommended a wide local excision of the lesion with 1 cm surgical margins followed by close observation for any sign of recurrence. The exact length of follow-up could not be determined because of the lack of guidelines on this issue. The lesion was completely excised down to the periosteum and the defect repaired with a full-thickness skin graft. Histology from the excised specimen showed that full clearance was achieved, and as there was no clinical evidence of metastasis, staging CT scan was not performed.

OUTCOME AND FOLLOW-UP

At 10 months follow-up the patient remained well, however, another similar lesion had developed close to the previously excised area, suggesting the possibility of recurrence. On examination, no cervical lymphadenopathy or organomegaly was

Figure 2 High-power (×40 magnification) histological view showing spiradenoma cells with glandular differentiation and high-mitotic count suggesting malignant behaviour.

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noted. A CT scan was recommended by the MDT which surprisingly demonstrated metastatic lesions in his lung, liver and pelvis. The patient was subsequently referred to oncology and treated with epirubicin, cisplatin and capecitabine, in accordance with our local skin cancer chemotherapy protocols. Unfortunately, there was no regression of the disease after the five cycles of chemotherapy treatment, so second-line chemotherapy treatment with three cycles of gemcitabine 180 mg and carboplatin 400 mg was planned by oncology. Oestrogen receptor status was negative therefore hormonal treatment was not advocated.

DISCUSSION

MES is an extremely rare tumour, accounting for only 0.005% of all skin tumours,¹ but it is very aggressive in nature, resulting in high mortality.² Dabska³ reported the first case of MES in 1972, and since then fewer than 60 additional cases of MES have been described in the literature. In the majority of these cases, the tumour was found on the trunk and extremities, with only four cases reported on the scalp.^{2 4-6} The typical presentation is of a longstanding asymptomatic benign eccrine spiradenoma which undergoes a change in size, becomes more tender, ulcerated or necrotic.³ It is most common in young adulthood with equal prevalence in males and females. Metastases occur in nearly 40% of cases, occurring in (in descending order of frequency); regional lymph nodes, lungs, brain and liver. Local recurrence rates have been shown to be up to 57%.⁷

There are no current guidelines for the treatment of MES. Most of the literature on MES advocates surgical excision with 1 cm margins or Mohs micrographic surgery as the main curative treatment.^{5 8} Regional lymph node dissection should be performed if metastases are clinically suspected. Radiotherapy alone or combination with chemotherapy has no current role in the treatment of MES,⁹ because sweat gland tumours are radio-resistant and studies investigating a role for chemotherapy are limited.^{7 9} On the other hand, Sridhar *et al* reported their experience with two patients of whom one patient had a metastatic oestrogen receptor positive tumour which was treated with tamoxifen. Results after 6 weeks showed local pain relief and improvement in lymph node and bone metastases.¹⁰ It has been recommended that close follow-up should be provided for patients with MES, every 3 months in the first year, then every 6 months in the second year and annually thereafter.⁸

Four cases of MES of the scalp have previously been reported,^{2 4-6} and these cases demonstrate differences in management of these patients and clinical outcomes.

Seyhan *et al*⁴ described a case of MES of the scalp with cranial involvement that had arisen 8 months following inadequate removal and malignant transformation of a benign eccrine spiradenoma. Further excision of the lesion and skin grafting was performed. At 24-month follow-up, there was no clinical evidence of tumour recurrence or metastasis.

Multiple wide local excisions may be required to achieve histological clearance of MES of the scalp, as in the case of Beekley *et al*² who reported a <2 cm MES on the vertex of the scalp. MRI and lymphoscintigraphy demonstrated no evidence of metastasis, and the patient remained clear of tumour after

14 months. Russ *et al*⁵ described a case of MES on the scalp treated with Mohs micrographic surgery. The lesion measured <2 cm and four lymph node metastases were identified using lymphoscintigraphy. The patient was monitored closely, and had no recurrence or metastasis after 12 months. One case of metastatic MES of the scalp has been reported previously.⁶ In this case, a 72-year-old black woman developed a 12 cm MES, which had enlarged over 23 months. The lesion was treated with surgical excision and skin grafting, however, the patient subsequently developed a metastasis to the neck causing C6 vertebral body compression.

To our knowledge, no guidelines exist as to how to best treat, investigate and follow-up on patients with MES. Although it is a rare condition, it is very aggressive with high rates of metastasis, therefore adequate surgical clearance and close follow-up are required. In this case, surgery with wider local excision achieved complete histological clearance, however, this failed to prevent subsequent tumour recurrence and metastasis. Therefore, repeat biopsy and early consideration of staging investigations should be recommended when tumour recurrence or metastasis is clinically suspected.

Learning points

- ▶ Malignant eccrine spiradenoma on the scalp is rare.
- ▶ This type of tumour is very aggressive in nature and has a high metastatic rate, therefore prompt identification and surgical excision of these tumours are essential.
- ▶ Close follow-up is advised post-treatment to detect recurrence or metastasis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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