Malignant peripheral nerve sheath tumour metastasising to the parotid gland

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Summary Two rare cases of metastases from malignant peripheral nerve sheath tumours (MPNST) of the head and neck are described. The initial lesions were superficial, but despite early diagnosis and complete excision, they metastasised to the ipsilateral parotid gland. Both underwent further complete excision and neither metastatic lesion showed progression in grade. One patient is alive, however, the other died of widespread metastases. There are no other reported cases of these sarcomas in the head and neck metastasising to the parotid gland.

Malignant sarcomas of the peripheral nerves, which include neurofibrosarcoma, malignant schwannoma and malignant peripheral nerve sheath tumours (MPNST) are rare, the estimated incidence of MPNST being 0.001%, however, they still represent 5–15% of soft tissue sarcomas in the head and neck region.[1] These tumours are generally regarded as deep soft tissue lesions[2] and are often associated with neurofibromatosis[3,4] (Von Recklinghausen's disease) and previous irradiation. A superficial form has been described,[2,5] which is locally aggressive, but very rarely metastasises, and the mainstay of treatment is based on a general principle of management of sarcomas, where wide local excision is advised with adjuvant radiotherapy for incompletely excised and high grade disease. We describe the treatment of two cases of superficial MPNST of the head and neck, which metastasised and we can find no similar cases, in the literature.

Case 1

In 1995, a 31-year-old labourer presented to his GP with a 10 month history of a painless, slowly growing lesion on his left temple, which on incisional biopsy showed low grade MPNST. Staging procedures were negative and he underwent wide excision and reconstruction with a full thickness skin graft.

Follow-up was uneventful, until 1997 when he noticed a swelling in his left parotid gland but no facial palsy. A CT scan showed a 1.6×1.9 cm mass in the superficial part of the gland (Fig. 1) and fine needle aspiration cytology (FNAC) was unhelpful. A superficial parotidectomy was performed, which confirmed a completely excised metastatic MPNST with extracapsular spread (Fig. 2).

Three years later, he developed subcutaneous, pulmonary and then bony metastases and died of disease after palliative chemo and radiotherapy in 2001.

Case 2

In July 2000, a 19-year-old man underwent excision

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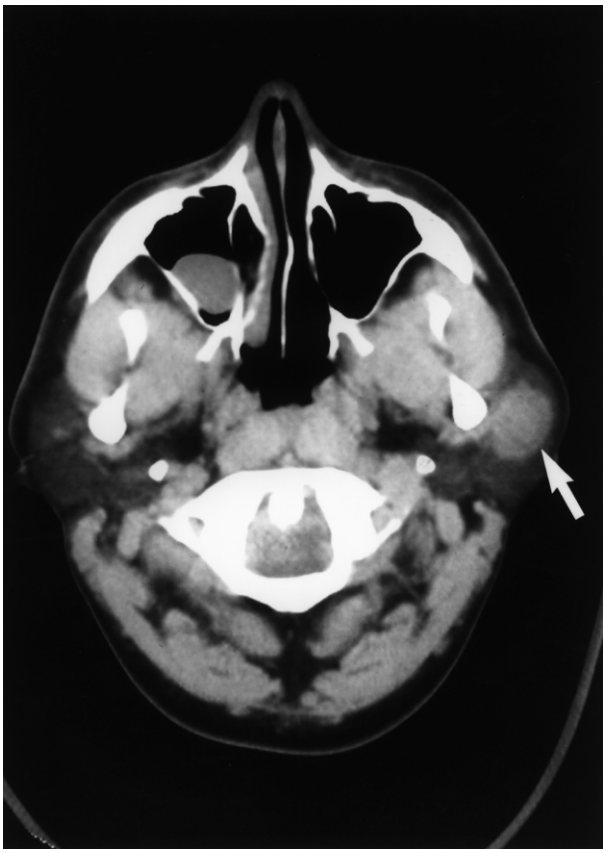

Fig. 1 CT head showing the metastatic lesion in the left parotid gland.

of a skin lesion in the right preauricular region, which was diagnosed as a low grade sarcoma and widely excised at a second operation. Nine months later he developed an FNAC positive metastatic lymph node in the ipsilateral parotid gland. CT demonstrated no other foci and review of the original biopsy together with the metastatic deposit established a diagnosis of MPNST, Trojani grade 2. He underwent a right modified radical neck dissection with incontinuity superficial parotidectomy and histology confirmed a single metastatic deposit in an intra-parotid lymph node with no progression of grade. He received 54 Gy in 27 fractions and is currently disease free 16 months later.

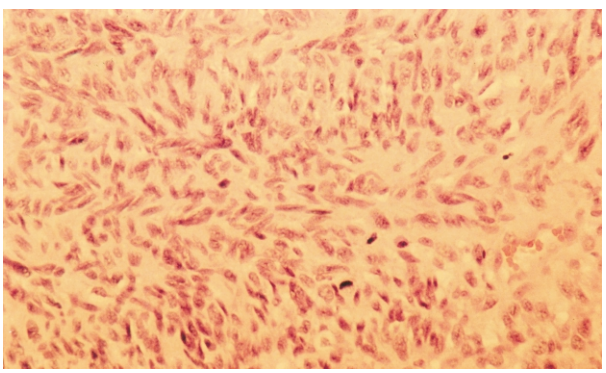

Fig. 2 Histology of the metastatic deposit in Case 1, showing a low grade spindle cell sarcoma. (Haematoxylin and Eosin × 300).

Discussion

The estimated incidence of MPNST is 0.001%, but represents 5–15% of soft tissue sarcomas of the head and neck.[1] In a current review of 56 head and neck sarcomas treated in our unit, MPNST's were the most common type, occurring in 18% of cases.[6] MPNST are generally regarded as deep soft tissue lesions,[2] arising from major nerve trunks, especially the sciatic nerve, brachial and sacral plexuses and are, therefore, commoner in the trunk, upper and lower extremities.[3] In the head and neck, they arise from all cranial nerves except the optic and olfactory, which have no nerve sheath.[7]

A superficial form has been described,[2,5] which arises in the dermis and subcutis from cutaneous nerves and is locally aggressive with a recurrence rate of 78%. The two cases in this report had wide local excision alone and unusually have had nodal metastasis rather than local recurrence, suggesting that adjuvant radiotherapy would not have helped.

Histological diagnosis has become more stringent because of similarities with other spindle cell malignancies e.g. leiomyosarcomas, malignant fibrous histiocytomas and neurotropic malignant melanoma.[8] In the past, it was necessary to demonstrate origin from a nerve,[3] however, cutaneous nerves are generally too small to identify grossly and hence electron microscopy and immunochemistry are used to show ultrastructural evidence of Schwann cell differentiation with absence of premelanosomes and absence of epidermal melanocyte proliferation. There are no specific immunohistological markers, therefore, a panel of antibodies is required and immunoreactivity with at least two of the following antibodies: S100, Leu7, myelin basic protein, glial fibrillary acid protein and PGP 9.5[8] is needed to make the diagnosis.

CT and MRI scans delineate the extent of the disease, the involvement of vital structures[9] and allow staging. MPNSTs infiltrate local tissue extensively and spread preferentially, as with other sarcomas, via the bloodstream to liver, lungs and bone rather than the lymphatics.[10] Regional lymph node involvement occurs in less than 1% of deep seated disease[3] and is even rarer in the superficial form,[5] but there are no statistics recorded. No one has previously described metastases from a

superficial head and neck primary to the parotid gland or lymph nodes. Punjabi described an MPNST in the left parotid which metastased to the contralateral parotid and auditory canal with intra-cranial extension, but it is unclear whether the right parotid was the site of the metastasis or was invaded secondarily.[10]

Poor prognostic indicators for MPNST's are a lesion greater than 5 cm diameter, incompleteness of excision and association with neurofibromatosis.[3]

Surgery is the mainstay of treatment for MPNST, requiring radical resection, and additional frozen section of proximal nerve to ensure clear margins,[1] although reviews do not suggest a margin of excision. Since lymph node involvement is very unusual, elective neck dissection is not recommended. Adjuvant high-dose radiotherapy is used, particularly after incomplete resection or where radical excision is impossible, and chemotherapy remains controversial.[11]

The superficial form of the disease after primary surgery has a local recurrence rate of 78%, metastatic spread in 22% and a 4 year survival of 66%. Unfortunately Dabski's series was small and only half the lesions were located in the head and neck region.[5]

Five year survival of all patients with MPNST's of the head and neck, ranges from 15 to 34%[1] with 50% of cases developing local recurrences and 33% metastasise, particularly to the lung.[10]

We conclude that superficial MPNST of the head and neck region should be excised as widely as possible, and elective neck dissection is not recommended. Adjuvant radiotherapy should be given when the disease is incompletely excised, the margins are close or high grade tumour.

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