Malignant peripheral nerve sheath tumour in the submandibular space

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ABSTRACT

Malignant peripheral nerve sheath tumours (MPNSTs) are a rare but aggressive form of soft tissue sarcoma with few reported cases in the anatomic location seen with the case presented here. This case involves a 23-year-old man presenting with a soft tissue mass under the mandible found to be a MPNST, which was investigated and fully excised. Adjuvant radiotherapy was used postoperatively. Although uncommon, these tumours have a poor prognosis and suitable management therefore needs to be put in place as soon as possible. Complete excision with negative margins and adjuvant radiotherapy are the current methods used for treatment. This case is unusual and highlights the management of an aggressive tumour in a difficult anatomical area, prompting the consideration of MPNST as a differential diagnosis for a soft tissue mass.

KEYWORDS

Malignant peripheral nerve sheath tumour – Head and neck neoplasms – Surgical oncology – Maxillofacial surgery – Plastic surgery

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Malignant peripheral nerve sheath tumours (MPNSTs) are an uncommon heterogeneous group of tumours arising from (or differentiating to) constituents of peripheral nerve sheaths (eg Schwann cells or perineural fibroblasts). This is typically a poorly differentiated, aggressive soft tissue tumour with a poor prognosis. It has a history of confusing nomenclature, known previously as malignant schwannoma, neurofibrosarcoma and neurogenic sarcoma. This case report details a case of MPNST in an unusual location.

Case history

A 23-year-old man presented with a 2-month history of a rapidly growing, painful mass under his right mandible, with a neurofibroma having been excised in that location six years previously. This was a tender, fluctuant, soft tissue mass. Urgent magnetic resonance imaging showed a 5.4cm × 4.7cm × 5.7cm tumour in the right submandibular space. A core biopsy showed it to be a MPNST. Following positron emission tomography and computed tomography, the lesion was surgically excised. A radical segmental mandibulectomy (Fig 1), wide local excision and local node clearance were performed, leaving a large defect requiring free fibula and anterolateral thigh flaps to cover the area. The tumour was fully excised and confirmed as a MPNST. Staining was positive for S100 protein. An additional separate neurofibroma adjacent to the tumour was also excised.

The patient recovered well postoperatively with good cosmesis and function. There was no additional local

spread found at the time of operation and the patient received postoperative radiotherapy.

Discussion

MPNSTs are rare, accounting for 5% of soft tissue sarcomas with an incidence of only 0.001% in the general population.¹ They are the malignant sequelae of neurofibromas and schwannomas. MPNSTs are generally found arising from large nerves (eg sciatic nerve, brachial plexus and sacral plexus). Typical onset is between 20 and 50 years of age, and there is an equal sex distribution. There is an association with neurofibromatosis type 1 (NF1); 5–42% of cases have NF1 although more than half arise spontaneously.² There is a higher prevalence of multiple MPNSTs in patients who have NF1; such MPNSTs are typically more aggressive and have a worse prognosis.⁵ The pathogenesis is thought to involve malignant change from pre-existing plexiform neurofibromas, with an increased risk of malignant change in patients exposed to radiation.

Typical locations for these tumours are in the trunk (~50%) and extremities (~30%); only a small minority (10–20%) is found in the head and neck region.¹ Panigrahi *et al* reported an interesting case of an orbital MPNST and reviewed other case studies of MPNSTs in unusual locations, finding only a few cases in the head and neck region.² The mainstay of published cases of MPNSTs in the head and neck region comes from Minovi *et al*, who reported a series of ten cases.⁴ Of these, eight tumours

Figure 1 Lateral and lateral views of resected specimen

were located at the lateral skull base and two lesions were in the sinonasal tract. There appear to be no case studies in the literature that describe a MPNST in the submandibular/mandibular area, such as in this case. The common presenting features of these tumours were pain, deformity, local pressure symptoms, neuralgia and nerve dysfunction (through both pressure and infiltration).

The basis of treatment is wide local excision, margins of at least 3–5cm having the most evidence of benefit. Many authors advocate as complete an excision as possible and consideration of amputation if necessary. It has been shown that full excision with negative margins has better outcomes than biopsy or partial excision, with increased survival and reduced rates of recurrence.⁶ Results have been disappointing for chemotherapy, with examples even worsening prognosis.⁴ However, many studies have demonstrated some benefit of adjuvant radiotherapy, which typically reduces recurrence and improves survival. The inclusion of brachytherapy and intraoperative electron irradiation has shown added benefits to survival and local control of disease.⁶

The outlook for patients with MPNSTs is unfavourable but is improving with better surgical options and use of concomitant treatments. The average five-year survival rate for all patients with MPNSTs is between 49% and 60% but this can be as low as 15–20% in anatomical sites such as the head and neck.^{1,7} The principal factor affecting survival is the degree of surgical treatment. Hicks *et al*

assessed 485 cases and the mean survival for those without surgical treatment was 12 months compared with 52–62 months for wide local excision.⁷ Wong *et al* reported three and five-year survival rates of 84% and 72% respectively with adjuvant radiotherapy versus 61% and 50% without radiotherapy.⁶

Conclusions

MPNSTs are one of the most aggressive forms of soft tissue tumours in the head and neck region, with a high rate of recurrence and metastasis. Management requires immediate investigation of suspicious masses and a multimodal approach to treatment. Complete excision with negative margins and postoperative radiotherapy is the current gold standard.

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