CASE REPORT

Shared decision-making in treatment of Merkel cell carcinoma

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SUMMARY

An 82-year-old woman presented with an asymptomatic mass, rapidly growing on her left cheek for the previous 3 months. Punch biopsy of the tumour was performed, and the pathology was compatible with Merkel cell carcinoma. A resection margin of more than 1 cm would involve left oral commissura, potentially damaging speech, eating and drinking ability. The patient had a strong wish of keeping surgery simple in order to maintain quality of life. Tumour excision was performed with 1 cm resection margin, and postoperatively the patient was referred to adjuvant radiation therapy. Sensibility of upper and lower lip remained unaffected, while motor innervation of left upper lip was impaired. Despite this, the patient's ability to talk and eat was unaffected. Surgery, with adjunctive radiation therapy, is the first-line of treatment for the primary tumour. The option for a more conservative treatment is not first choice, but can be considered upon individual assessment.

BACKGROUND

Rapid growing masses are highly suspicious of malignancy. Merkel cell carcinoma is a rare form of skin cancer, often treated with wide excision margins. When located in the face near significant facial features, reconstruction can be challenging. Involving the patient in the decision-making is mandatory.

CASE PRESENTATION

An 82-year-old woman presented with an asymptomatic, rapidly growing mass on her left cheek for the past 3 months ([figure 1](#)).

The patient's medical history revealed muscular rheumatism and temporal arthritis, with good symptom control achieved by Prednisolone. On physical examination, a 3 cm red nodular tumour with palpable broader underlying extension (4 cm total) was present on the left cheek. No palpable lymphadenopathy was found on the neck, in the axillae or groins.

INVESTIGATIONS

Punch biopsy of the tumour was performed, and the histological findings were compatible with Merkel cell carcinoma. Tumour cells expressed positive reaction for the neuroendocrine marker synaptophysin together with a positive reaction for the epithelial markers cytokeratin 20 (dotlike) and pancytokeratin. Tumour cells were negative for thyroid transcription factor (TTF-1) excluding metastasis from a small cell carcinoma of the lung.

Figure 1 Rapidly growing, asymptomatic mass on her left cheek. Well defined, non-adherent.

Chest X-ray was performed, showing no infiltrations.

TREATMENT

The patient had a strong wish for keeping surgery simple in order to maintain quality of life. Resection margin of more than 1 cm would involve left oral commissura, potentially damaging speech, eating and drinking ability. Several reconstructive possibilities were considered, including primary radiation therapy, and the patient had the different surgical reconstructive scenarios depending on excision margin explained.

Ultimately, tumour excision was performed with 1 cm resection margin, reconstructed with a distal V-Y advancement flap with fish-mouth closure combined with cranial reverse-V-shaped excision of excess skin and subcutaneous fat ([figure 2](#)). Postoperatively, the patient was referred to adjuvant radiation therapy.

OUTCOME AND FOLLOW-UP

Microscopic examination of surgical specimens declared clear margins of resection. The flap presented fully vital immediately after surgery and 1 week postoperative. Sensibility of upper and lower lip remained unaffected, while motor innervation of left upper lip was impaired.

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Figure 2 Distal V-Y advancement flap with fish-mouth closure combined with cranial reverse-V-shaped excision of excess skin.

(figure 3). Despite this, the patient's ability to talk and eat was unaffected.

DISCUSSION

Merkel cell carcinoma is an aggressive and relatively uncommon form of skin cancer, considered to be a type of neuroendocrine tumour. Merkel cell polyomavirus (MCV) likely contributes to the development of the majority of these tumours. They predominantly occur in people over 65 years of age with sun-exposed skin and have a high propensity for local recurrence and regional lymph node metastases.^{1 2}

Surgery is the first-line treatment for the primary tumour. A wide, clear margin of resection of 2–3 cm is advised.^{3–7} Staging is based on TNM classification, and prognosis strongly depends on the disease stage. Sentinel node biopsy (SNB) should be performed in cases where lymph nodes are not clinically detectable, equivalent to cN0 stage. It remains controversial whether SNB prolongs survival or just prolongs disease-free survival and improves local control.^{8–11} In addition, radical lymph node dissection has not shown to increase overall survival, but is advised in case of positive sentinel node. Radiation therapy as primary treatment is generally not advised but adjunctive radiation therapy to the tumour site has shown to reduce local recurrence. The role of adjunctive radiation therapy to regional nodes in case of positive sentinel node is controversial, and both advocating and opposing studies can be found in the literature.^{5–7} Systemic chemotherapy, akin to regimens for small cell carcinoma of the lung, may be considered as an adjuvant following surgery in case of metastatic disease.^{5 12}

The option for a more conservative treatment is not first choice, but can be considered upon individual assessment. In

Figure 3 Fully vital flap 1 week postoperative.

this case, radiation therapy as primary treatment was deviated due to tumour size and estimated prolonged duration of treatment. The patient had clinically no enlarged lymph nodes at the time of diagnosis and treatment. The procedure of SNB, as well as the possibility of further resection depending on the histological findings, was carefully explained to the patient and relatives. The patient clearly expressed no intention of proceeding with total resection of the lymph nodes if indicated by SNB, due to her age and general health. So given the potential complication, risk and questionable gain on SNB, we refrained from performing this procedure due to the patient's wish for limited surgery. No further diagnostic imaging was performed besides chest X-ray, as it would only be of academical interest and not beneficial to the patient.

Learning points

- Multidisciplinary approach is advised in the treatment of Merkel cell carcinoma.
- A combination of surgical excision with wide margins and adjuvant radiation therapy is preferred.
- Staging is based on TNM classification, and prognosis strongly depends on the disease stage.
- The patient's thoughts and wishes should be taken into consideration when planning the course of treatment.

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Competing interests None.

Patient consent Obtained.

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