Abstract. Background: Primary extramedullary plasmocytomas (EMPs) are plasma cell tumours that arise outside the bone marrow. As these tumours are rare and can present without a typical clinical picture, correct diagnosis is difficult to confirm, particularly when the lesions occur at uncommon sites. Case Report: This is a case report of a 63-year-old patient who initially presented with an isolated lump on his left knee. Biopsy showed an anaplastic tumour which was first diagnosed as localised rhabdomyosarcoma; therefore, a combined approach with neoadjuvant radiotherapy followed by surgery was recommended. Shortly before radiotherapy started, the patient suffered a stroke with hemiplegia. CT and MRI of the head revealed a single brain lesion. A resection of this cerebral lesion was performed and surprisingly revealed a cerebral EMP. After comprehensive review of this specimen and the previous biopsy, the diagnosis was changed to anaplastic bifocal EMP. Generalised myeloma was excluded; therefore the patient was treated with definitive radiotherapy of the left knee region and postoperative partial brain irradiation. Unfortunately, the patient died several months later due to fulminant progressive disease at both treated sites despite rapidly initiated chemotherapy. Conclusion: Correct diagnosis of EMP may be difficult, particularly as this disease is rare and can present with atypical clinical picture and immunophenotype. Review of the specimen by a histopathologist with special interest in soft tissue tumours or lymphoproliferative disorders is strongly recommended.

Extramedullary plasmocytoma (EMP) is a rare entity belonging to the category of non-Hodgkin lymphoma. It was reported for the first time in 1905 and since then it has been described in numerous case reports. EMPs regularly manifest without signs of systemic spread but multicentric disease is possible. Progression to multiple myeloma has been reported in about 10 to 15 % of cases (1, 2).

EMPs are predominantly located in the head and neck region, mainly in the upper respiratory tract, but may also occur in the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, and skin (3, 4). As these tumours are rare and can present without a typical clinical picture, a correct diagnosis is difficult to obtain, especially in lesions at uncommon sites. This case report presents an unusual case of an isolated soft tissue tumour which was initially misinterpreted as a rhabdomyosarcoma. The patient developed metachronous a second lesion in the brain which was resected. Only at this time was the correct diagnosis of primary EMP as bifocal disease established.

Case Report

A 63-year-old male patient initially presented with an isolated soft tissue mass of the left knee. This patient had been treated for gonarthrosis for two years before a lump on his left knee was detected and further diagnostic investigations were initiated, including X-ray, computed tomography (CT) (Figure 1) and fine-needle aspiration. Biopsy revealed a proliferation of anaplastic, malignant cells, and the tumour was interpreted initially as an alveolar rhabdomyosarcoma (RMS). Therefore, an interdisciplinary approach with neoadjuvant radiotherapy followed by surgery was initiated. Shortly before radiotherapy was started, the patient suffered from a stroke with hemiplegia. CT and magnetic resonance imaging (MRI) of the head (Figure 2) revealed a single intracerebral lesion fronto-parietally. No further metastases were detected. Assuming a solitary brain metastasis of the RMS, a surgical resection of the single brain lesion was performed in curative intention. Surprisingly, histopathological findings exposed an anaplastic EMP. After a comprehensive review of the
cerebral lesion and the previous biopsy of the soft tissue mass close to the left knee including clonality analysis, the final diagnosis of anaplastic EMP was confirmed. Generalised disease (namely, multiple myeloma) was excluded by further laboratory tests including M-protein and bone marrow biopsy.

After establishment of the correct diagnosis of EMP, definitive local radiotherapy (RT) of the left knee region with a total dose of 50 Gy (2.0 Gy daily, five times a week) and post-operative local RT of the cerebral tumour bed with a total dose of 35 Gy (2.5 Gy daily, five times a week) were performed. At the end of the RT, the lump of the left knee showed a slow regress. As pain also resolved, surgery of the knee tumour was not performed.

Two months after completion of the RT, there was significant tumour progress in both initial tumour sites, with a rapid clinical deterioration mostly through progress at the former cerebral lesion. Rapidly initiated chemotherapy with melphalan and prednisone had no effect and the patient died five months after initial diagnosis.

Discussion

Primary EMPs are rare plasma cell tumours that occur in extramedullary organs or tissues and comprise about 4% of all plasma cell tumours. About 80% of the EMPs are located in the head and neck region, mainly in the upper respiratory tract. EMPs located outside the head and neck region are very rare and, particularly as high-grade anaplastic disease, can present with atypical clinical picture, morphology and immunophenotype (1).

In this case report, an EMP occurred first as an isolated soft tissue mass of the left knee, mimicking the clinical picture of a soft tissue sarcoma and the cytological picture of a rhabdomyosarcoma. An additional second lesion in the brain aggravated the diagnostic difficulties in this case.

The diagnosis of EMP is usually based on the morphologic and immunophenotypical finding of a localised collection of monoclonal plasma cells in the absence of plasma cell proliferation elsewhere, especially in the bone marrow, and without the presence of malignant lymphoma (5). In cases of EMP with severe cytological anaplasia, there are two major differential diagnoses, firstly immunoblastic lymphoma, and secondly poorly differentiated carcinoma or sarcoma. Table I summarizes the characteristics of these entities and the presented case.

Of all plasma cell tumours, primary EMP has the best prognosis. Nevertheless, about 8% to 36% of patients with EMP show conversion to multiple myeloma, resulting in shorter survival (1, 6). Therefore, secondary diagnostic
procedures should be carried out to exclude a generalised disease once the diagnosis of EMP is confirmed histologically (7).

Detailed evaluation of the literature revealed that after adequate local treatment of EMP including surgery, radiotherapy or their combination, approximately 65% of all patients had no recurrence or systemic involvement, 21% had a local recurrence, and 14% converted to multiple myeloma; the outcome for patients with EMP located outside the head and neck region appears to be similar to those with EMP in the upper respiratory tract (1).

Currently there are no general guidelines for the treatment of patients with EMP. However, EMPs are radiosensitive tumours, and this leads to the acceptance of radiotherapy as the treatment of choice for this disease with good local control rates. The required total radiation dose ranges from 40 Gy to 60 Gy given over a period of 4 to 6 weeks (8). Surgery or combined surgery and radiotherapy have been applied as often as radiotherapy alone in the treatment of EMP in the head and neck region. Few patients undergoing chemotherapy with or without combined local treatment are described (8).

If EMP is present in a soft tissue lump that is amenable to operation, surgical excision is usually sufficient (9). In all other cases, such as the presented case, definitive radiotherapy should be preferred. Among all patients presenting an EMP outside the head and neck region, 56% were treated with surgery alone. However, complete removal of an EMP is not always possible. Especially in the upper respiratory tract adjacent vital organ structures may preclude radical intervention. For such patients, surgery followed by radiotherapy is recommended. Patients who are diagnosed with primary EMP remain under life-long medical observation because relapses or a generalised disease (namely, multiple myeloma) may appear even years after successful initial therapy (1).

**Conclusion**

This case report emphasises the diagnostic challenges regarding high-grade anaplastic extramedullary plasmocytoma. Particularly as the disease was located outside the head and neck region, it may present as an atypical clinical picture, in regard to morphology and immunophenotype. The occurrence of two separate, uncommon sites at presentation aggravated the diagnostic difficulties. Review by a histopathologist with special interest in bone and soft tissue tumours or lymphoproliferative disorders is therefore strongly recommended.

**References**