CASE REPORT 1

An 85 year-old man was referred in 2001 with a left jugular node that was surgically removed. Pathologic examination showed a metastasis from a neuroendocrine tumor. There were two recurrences requiring additional surgical excisions and radiotherapy. The primary site of tumor remained unknown until 2002. At that time, appeared a painful and enlarging mass involving the soft tissues of the right temporal region.

CT and MRI showed extensive infiltration of the soft tissues of the right temporal and left frontal regions, hyperdense on CT (figure 1a), isointense on T1-W (figure 2a), hypointense on T2-W (figure 3), with marked enhancement after contrast administration (figures 1b et 2). Both temporal and frontal lesions were aggressive with local invasion of the skull and meninges underlying the scalp tumor. Bone marrow replacement underneath the lesion was well demonstrated at MRI but was no evident at CT (figure 1). The brain parenchyma was displaced by the meningeal mass without evidence of invasion.

The left frontal lesion was in close relationship with the superior sagittal sinus but there was no evidence of sinus thrombosis.

CT of the thorax and abdomen and bone scan showed no evidence of metastasis.

The patient was not a surgical candidate, and underwent chemotherapy (VP 16). Follow-up CT 15 days later showed a reduction of about 50% of the volume of the lesions.

CASE REPORT 2

A 68 year old man was referred in 2001 with a large left axillary node that was surgically excised. Pathology was consistent with a metastasis from a Merkel cell carcinoma (the primitive skin tumor was not found).

The patient presented again in July 2002 with progressive left hemiplegia and headache.

MRI showed a tumor in the right posterior parietal region. This lesion was hyperintense on T1-W images (figure 4), hyperintense on T2 and FLAIR-W images (figure 5), hypointense on diffusion-W images (figure 6) and showed ring enhancement
after contrast administration (figure 7). There was local mass effect and right lateral ventricular compression.

The tumor was surgically removed and the pathologic examination confirmed its neuroendocrine origin. A recurrence occurred later in the same cerebral location requiring additional treatment (steroids and radiotherapy).

During follow-up, the patient developed multiple intramedullary and leptomeningeal metastases associated with lymphadenopathy.

Despite treatment, the disease was fatal 17 months after the first symptoms.

DISCUSSION

MCC is an uncommon aggressive tumor of the dermis first reported by Toker in 1972 as a trabecular carcinoma of the skin [11].

Since then it is known as a primary small cell carcinoma of the skin.

Immunohistochemistry shows positive neuron specific enolase staining demonstrating the neuroendocrine nature of this tumor. Recently a specific marker (cytokeratin 20) was isolated for MCC [3].

This pathology occurs in caucasian patients with a mean age of 70 years with no sex predilection. Risk factors include sunlight exposition.

It involves predominantly head and neck regions but other locations are also described in the literature, including the nasal fossa [1, 9] and external auditory canal [8].

Skull and brain involvement are very rare and, to our knowledge, this is the second case published in the literature describing skull infiltration [5].

Because of the rarity of MCC, there is no universally accepted staging system for this type of tumor, but the imaging findings are important for treatment planning and management as well as follow-up [6].
A radiological classification based on local and distant metastasis has been proposed (stage 1: cutaneous involvement, stage 2: regional nodal invasion, stage 3: systemic metastases) [10].

Exhaustive staging necessitates further work-up: Technetium 99 lymphoscintigraphy is used for precise node sampling, and the distant metastases (present in 1/3 patients at the initial presentation) can be evaluated at Octreotide scintigraphy [4] or at PET imaging with fluoro-2-deoxy-d-glucose.

The prognosis is poor with an average survival of 8 months for stage 3 disease [10]. Poor prognostic factors include male sex, size of the primary tumor >2cm and metastatic disease [7].

Treatment must be aggressive with wide-margin surgical excision and an adjuvant radiotherapy. Chemotherapy is used if surgery is contraindicated and for stage 3 disease. However, the optimal therapeutic protocol remains controversial [2].
In summary, one should suggest a diagnosis of MCC in the presence of aggressive and extensive cutaneous lesions of the head and neck in an elderly patient.

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REFERENCES


