CHAPTER 6

Malignant sellar, parasellar and skull base tumors

Clinicians

R. Grob, J.-V. Chantelard, E.-Ch. Antoine, J. de Recondo, B. Charbonnel, D. Khayat

Radiologists

B. Dupas, J.-M. Mussini, M. Gayet-Delacroix, D. Buthiau

Accurate imaging by CT and particularly MRI is indicated in cases where the endocrinological and/or secondary neurological signs point to a tumor involving the sellar, parasellar regions or the skull base.

Endocrine signs

Hypersecretion of the anterior pituitary hormones causes the clinical features which necessitate imaging of the hypothalamo-pituitary region to determine the etiology. Hyperprolactinemia is the commonest, but acromegaly due to growth hormone hypersecretion, Cushing’s disease and exceptionally gonadotrophin and thyrotrphin secreting adenomas are also encountered. Insufficiency of the anterior and posterior pituitary is frequently associated with tumors and is an indication for imaging. Diabetes insipidus and polydipsia are often associated with a hypothalamic lesion which first excludes simple causes such as diabetes mellitus or hypercalceemia.

Neurological signs

Involvement of cranial nerves

Cranial nerve involvement may be isolated or include several contiguous or even topographically remote nerves. In the latter case, it is useful to describe the associated syndromes according to the localization (table 6.1): syndrome of the sphenoidal fissure, jugular foramen, petrous apex... These syndromes comprising of various cranial nerve deficits should always immediately raise the suspicion of an underlying tumor. Clinical examination at this stage is a useful guide to the radiological investigations.

Similarly, syndromes of the anterior, midportion or posterior wall of the cavernous sinuses are defined by the associated cranial nerve deficits and have the same valuable feature of localizing signs. These are very similar or even identical to the so-called parasellar syndromes. They are likely to be due to arterial or venous abnormalities, either acute or chronic, or local inflammatory lesions as tumors.

Isolated cranial nerve involvement is much less specific both for localization and etiology. For instance, oculomotor nerve paresis, pure hemifacial spasm,

Table 6.1. The principal clinical syndromes of multiple cranial nerve involvement

<table>
<thead>
<tr>
<th>Semiology</th>
<th>Name of syndrome</th>
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<tbody>
<tr>
<td>II, III, IV, V, VI</td>
<td>Syndrome of the orbital apex</td>
</tr>
<tr>
<td>III, IV, V, VI</td>
<td>Syndrome of the sphenoidal fissure</td>
</tr>
<tr>
<td>III, IV, VI, ± 2 or 3</td>
<td>Syndrome of the cavernous sinus</td>
</tr>
<tr>
<td>V, VI</td>
<td>Syndrome of the petrous apex, called Gradenigo’s syndrome</td>
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<td>V, sympathetic</td>
<td>Syndrome of the parastrigeminal space, called Rader’s syndrome</td>
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<tr>
<td>VII, VIII, ± other nerves</td>
<td>Syndrome of the cerebello-pontine angle</td>
</tr>
<tr>
<td>IX, X, XI</td>
<td>Syndrome of the jugular foramen</td>
</tr>
<tr>
<td>IX, X, XI, XII</td>
<td>Syndrome of the anterior condyle</td>
</tr>
<tr>
<td>I ± II ± III ± IV...XII</td>
<td>Garcia’s syndrome</td>
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regardless of the speed of onset, can be due to a multitude of etiologies. Neoplastic disease is not always the commonest. The most extreme example is palsy of the V1th cranial nerve which is the least specific of all the lesions in the CNS (the "Cinderella of Neurology" according to J. Lapresle). This explains the necessity and value of imaging over a large area if there are no preexisting localizing clinical signs.

Painful syndromes indicating a tumor of the skull base or parasellar region result from the sensory or very rich sympathetic innervation of certain areas of the meninges and should be considered separately. Such is the case for retro-orbital and parasellar lesions simulating painful ophthalmoplegia (Tolosa-Hunt) or ophthalmoplegic migraine; this also applies to certain lesions of the petrous bone invading the tentorium cerebelli and readily causing headaches suggestive of migraine or facial pain of vascular origin.

The sequelae of parenchymal involvement

These have become rare as presenting features and are avoided by the early use of modern imaging techniques in the majority of cases. Nevertheless, focal signs may occur during the clinical progression of such a tumor: epileptic fit, usually focal and especially "temporal", uncinate or hallucinatory, pyramidal motor deficit, signs of compression or distortion of the metencephalon or mesencephalon... the mechanisms are variable. A meningioma of the lesser wing of the sphenoid elevates the temporal cortex, whereas the same tumor in a more medial location, e.g. parasellar, may encase the internal carotid artery causing thrombosis and subsequent infarction.

CSF flow disturbances

Intracranial hypertension is rare in these tumors. Headaches are most commonly due to bone pain or related to invasion of sensory nerves such as the trigeminal nerve. Tumor expansion is rarely sufficient to cause pure intracranial hypertension. True hydrocephalus remains the exception:
- tumor of the skull base associated with an inflammatory reaction or tumor invasion of the arachnoid space which produces a communicating hydrocephalus;
- large lesion of the clivus and foramen magnum, complete obstruction to the circulation of CSF.

In these rare situations, the signs of intracranial hypertension may dominate the clinical picture at presentation.

Diagnosis of an anterior pituitary tumor

**Pituitary adenomas**

These represent 10% of intracranial tumors. They either present with hormonal hypersecretion (most common in young patients) or are non-functioning and are clinically silent until the onset of a complication (patients more than 50 years old) due to compression of the chiasm, hypopituitarism or hemorrhagic complications and/or acute necrosis [17, 27]. Rarely, adenomas arise in the context of a familial multiple endocrine neoplasia syndrome (MENS). The radiological classification is based on size criteria: a microadenoma is less than 10 mm and a macroadenoma greater than 10 mm. It is sometimes difficult to identify a microadenoma within a healthy pituitary and conversely to differentiate the normal pituitary parenchyma compressed by a macroadenoma.

**Diagnosis of microadenomas** (figs 6.1 & 6.2)

These benign lesions will not be described here.

**Diagnosis of macroadenomas**

Imaging is able to assess the extent of an intra- and/or suprasellar tumor of greater than 10 mm in diameter, but the differentiation between an expanding and invasive macroadenoma may sometimes be difficult. Pretherapeutic investigation is essential, examination in three planes is possible with MRI, the post-gadolinium T1-weighted sequence is indispensable to appreciate the limits and extent of the tumor and particularly its relationship to the cavernous sinus. CT is still useful for the diagnosis of calcifications. MRI would appear from now on to be superior to CT in the diagnosis, preoperative assessment and follow-up, either during medical treatment or post-operatively.

**Findings**

**CT**

Calcifications are visible in the sellar region on the pre-contrast examination; the enhancement following contrast injection is heterogeneous and depends on the consistency of the tumor, i.e. solid, partially necrotic or hemorrhagic. Following the injection, the contours of the tumor are generally best defined relative to the adjacent structures; but the extension of the