Painful peripheral facial palsy

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1. Case report

A 55 year-old man presented in emergency with grade IV right peripheral facial palsy on the House and Brackmann classification, and poorly systematized right semicranial pain of sudden onset 3 weeks previously, managed by corticotherapy (Prednisolone 1 mg/kg/day (i.e., 80 mg) for 5 days) as idiopathic facial palsy. There were no signs of cochleovestibular involvement. He had a history of smoking (80 pack-years). Initial pure-tone and speech audiometry found no hearing loss; impedancemetry found conserved stapedial reflex. Given the painful and non-regressive nature of the palsy, temporal bone CT was performed (Fig. 1A) and found a lytic image. Temporal bone MRI (Fig. 1B and C) was requested to specify the nature of the lesion.

2. Questions

Question 1: Interpret Fig. 1.
As part of extension assessment, contrast-enhanced cerebral, thoracic, abdominal and pelvic CT (Fig. 2) was requested.

Question 2: Interpret Fig. 2. How do you confirm the pathology diagnosis?
CT also showed bone lesions of the calvarium. A whole-body bone scan (Fig. 3) was performed.

Question 3: Interpret Fig. 3. What treatment do you propose?
Fig. 1. Right temporal bone CT, axial slices (A); axial T1-weighted cerebral MRI without gadolinium injection, through the temporal bone (B); axial T1-weighted cerebral MRI with gadolinium injection, through the temporal bone (C).

Fig. 2. Parenchymatous-window thoracic CT, axial slices, with contrast medium injection.
Fig. 3. Whole-body technetium-99 bone scan.

What is your diagnosis?
3. Replies

3.1. Reply 1

Fig. 1A is a right temporal bone axial slice CT scan without contrast injection. It shows mastoid cell involvement by a lytic lesion (arrow), with a crumbly aspect to internal table of the temporal bone. The lesion contacts the sigmoid sinus at the jugular foramen, which it enlarges, affecting the mastoid part of the facial nerve.

Fig. 1B is a cranioencephalic MRI axial T1-weighted acquisition without contrast enhancement, through the temporal bone. It shows a right temporal bone lesion (arrow) in T1 iso signal.

Fig. 1C is an axial T1-weighted fat-sat cerebral MRI acquisition with gadolinium enhancement through the temporal bone. The lesion (arrow) measures 16.23 × 26.29 mm. There is heterogeneous enhancement and a non-enhanced necrotic center. There is right sigmoid sinus compression.

These images suggest several possible diagnoses, and notably metastasis to the petrous and mastoid part of the temporal bone or endolymphatic sac tumor. Complete extension assessment is therefore necessary before undertaking surgical biopsy.

3.2. Reply 2

Fig. 2 is a contrast-enhanced parenchymatous-window thoracic CT scan, axial slice. It shows a ramified speculated tissue lesion of the apical segment of the right superior lobe (arrow), suggesting a presumably primary tumoral process.

The suggested diagnosis was metastasis of a primary lung tumor to the petrous part of the right temporal bone. Assessment required to be completed by anatomopathology. Transmastoid biopsy was considered, but entailed a risk of sigmoid sinus lesion, meningeal breach and aggravation of the facial palsy by compressive intratumoral hematoma or direct lesion. After multidisciplinary discussion with the neurosurgery and pneumology teams, it was decided to perform CT-guided biopsy of the primary pulmonary lesion, as entailing less risk. Histology found grade IV acinar adenocarcinoma.

3.3. Reply 3

Fig. 3 is a whole-body technetium-99 bone scan, showing several hyper fixation sites indicative of secondary bone lesions of the calvarium, right temporal bone, right iliac wing, sacrum and tibias.

This was a T2N2M1 pulmonary adenocarcinoma, with multiple bone metastases. A multidisciplinary oncology team meeting decided on palliative care, comprising: 3 cycles of cisplatin and pemetrexed (Alimta®) chemotherapy; analgesic right temporal radiation therapy.

CT reassessment during treatment found dissociated evolution, with a reduced pulmonary lesion but considerably increased temporal bone lesions.

Second-line chemotherapy with 3 cycles of docetaxel (Taxotère®) was initiated, but the patient died during the third cycle, 6 months after onset of facial palsy.

4. Comments

Idiopathic facial palsy is often considered in adult facial palsy, but should in fact be a diagnosis of elimination. Painful and persistent facial palsy casts doubt on such a diagnosis and should lead to temporal bone CT scan to rule out tumoral etiology. The physiopathology of this pain is complex: neural involvement by invasion or compression of the sensory contingent of the facial nerve innervating the Ramsey Hunt region, or of the trigeminal nerve in case of large lesion. The bone involvement itself may be implicated, with release of inflammation mediators by metastatic cells.

In the petrous part of the temporal bone, tumors liable to induce facial palsy are: cholesteatoma, paraganglioma, Langerhans cell histiocytosis, lymphoma, sarcoma, multiple myeloma in the elderly, endolymphatic sac tumor and metastasis of solid malignant tumor [1]. In the present case, the location, lytic CT aspect and heterogeneous MRI aspect with enhancement on injection might suggest malignant endolymphatic sac tumor with pulmonary metastasis. The symptomatology, however, usually includes cochlear and vestibular signs; moreover, although cases of adenocarcinoma have been reported, they had a papillary rather than acinar architecture; and finally, only one case of remote metastasis is to be found in the literature [2].

The malignant solid tumors most liable to metastasize in this region, according to Gloria-Cruz et al. [3], are mammary adenocarcinomas and, secondly, lung cancers. Adenocarcinoma is the most frequent histologic form of lung cancer found in petrous bone metastasis; others are undifferentiated carcinoma and small-cell neuroendocrine carcinoma [4].

The petrous apex is the region of the temporal bone most frequently involved by metastasis. Bone invasion is initially hematogenic, from the bone marrow of the petrous apex [3], spreading contiguously toward the mastoid. Isolated mastoid involvement, as in the present case, is rare: only 1.3% of temporal bone metastases in the series reported by Gloria-Cruz et al. [3].

The clinical expression of intrapetrous metastasis is polymorphic. Hearing loss seems to be the most frequent sign [3], followed by peripheral facial palsy, which is rarely isolated as in the present case. Bakhos et al. [1] reported the following symptoms in 2 patients: hearing loss, tinnitus, headache, dysphonia and retro-auricular tumefaction. Other revelatory signs mentioned in the literature are vertigo, otalgia, ototrauma and external auditory canal tumefaction [3].

Tumoral involvement of the facial nerve, by compression, inflammation or invasion, may occur at any point along the trajectory: in the brainstem, within the internal auditory canal, in the labyrinthine, tympanic or mastoid intrapetrosal segment, or at the exit from the stylomastoid foramen [4]. Facial canal invasion does not necessarily entail peripheral facial palsy: only 6 of the 14 patients with facial canal invasion in Gloria-Cruz et al.’s series [3] showed peripheral facial palsy (42.8%).

A lytic lesion on temporal bone CT is suggestive of a secondary lesion, and should be followed by MRI with T1 and T2-weighted sequences and T1 acquisition with gadolinium injection, to specify the lesion and its limits and explore for endocranial and meningeal complications. In case of early clinical signs in a non-advanced tumor, absence of bone erosion on CT and MRI may lead to misdiagnosis [3].

Histologic diagnosis is essential to treatment strategy. When a secondary lesion is suspected, however, biopsy should be preceded by complete extension assessment. Lesions are less easily accessible to surgery when situated in the petrous apex than in the mastoid: in the present case, given the location, transmastoid biopsy was considered, but was not carried out because the extension assessment revealed the much more accessible primary lesion [1].

The reference examination for bone metastasis is technetium-99 bone scan. Its specificity is, however, low as it detects osteoblastic activity whether of malignant or benign etiology. In case of malignancy, moreover, this activity may persist despite a good treatment response, making bone scan unsuited to assessing the latter. Positron emission tomography coupled to CT (PET-CT) is at present a great focus of interest for the detection of secondary bone lesions [5], and seems to be dependent on primary lesion
In bronchopulmonary cancer, the two examinations show comparable performance, with easier lesion location on the skeleton using bone scan and better specificity with PET-CT in case of doubt (which was not the case with the present patient). The other advantages of PET-CT are better image quality and the possibility of also detecting other non-osseous metastases.

Intrapetrous metastasis from a solid tumor indicates an advanced stage [1]. When a primary tumor with a single metastasis is operable, curative surgery at both locations is feasible. Treatment, however, is often in fact palliative, associating platinum-based chemotherapy and radiation therapy focusing on the metastases [1,4].

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References