Infraorbital nerve schwannoma

Schwannome du nerf infra-orbitaire

A 45-year-old woman visited our institution for explorative examination of a recent spontaneous, indolent, left suborbital mass. The patient had no prior medical history of such a finding. Clinical examination found left proptosis and a pain-free non-mobile infraorbital mass with no cutaneous signs of inflammation. The patient was not febrile, and neurological examination was normal. A non-contrast-enhanced computed tomography (CT) scan showed a tubular homogeneous, non-calcified mass in the infraorbital canal, which was enlarged because of the tumor (Fig. 1). Cortical bone erosion of the infraorbital canal and orbital floor was also present, and the left ocular globe and inferior rectus muscle were displaced upwards by the mass.

The patient also underwent magnetic resonance imaging (MRI; 1.5 T). On T1-weighted images (T1-WI), the mass presented as a homogeneous isointense signal (not shown here), and as a homogeneous hyperintense signal on T2-WI (Fig. 2A), with clear and homogeneous enhancement after gadolinium injection (Fig. 2B).

The location of the mass in the infraorbital canal and the MRI signal abnormalities were strongly evocative of an infraorbital nerve schwannoma. The patient subsequently underwent surgery to remove the mass (Fig. 3), which was excised by sacrificing the infraorbital nerve. Reconstruction of the orbital floor was performed, using a bone graft taken from the skull.

Histopathological examination (hematoxylin and eosin staining; original magnification × 200) showed spindle cells with irregular and wavy nuclei (Fig. 4A). On immunohistochemistry, the tumor was strongly positive for S-100 protein antigen (Fig. 4B). These findings pointed to a benign schwannoma.

Schwannomas are well-differentiated, encapsulated, benign lesions that are considered class I tumors by the World Health Organization (WHO) and arise from the Schwann cells of peripheral neural sheaths [3]. Orbital schwannomas represent 1 to 4% of orbital tumors [4] and usually arise from the supraorbital or supratrochlear nerves. However, infraorbital nerve involvement is unusual, with only 10 cases previously reported in the literature [2,5,6]. The main differential diagnoses for a mass at such a location are neurofibroma, perineural tumor infiltration and solitary fibrous tumor.

In the present case, the enlargement of the infraorbital canal with erosion of the cortical edges, but no bony effraction or extension into soft tissue, was suggestive of a slow-growing benign process. Also, the signal abnormalities seen on MR pulse sequences were strongly evocative of a schwannoma, according to the MRI characteristics found at other locations such as isointense signals on T1-WI, homogeneous hyperintense signals on T2-WI and marked homogeneous enhancement [1].

Despite its rarity, infraorbital nerve schwannoma may be considered a possible diagnosis in the case of an indolent infraorbital mass. Multislice CT and MRI are useful for making a positive diagnosis at this unusual location for schwannoma, and can also serve as a guide during surgical removal of the mass.
Figure 2  Magnetic resonance imaging (1.5 T) scans of the facial area. A. Coronal T2 spin-echo (SE)-weighted image (WI) shows a hyperintense, well-delineated tissue lesion that is causing the walls of the left infraorbital canal to bulge outwards (black arrows). In addition, the left rectus inferior muscle and orbital globe are displaced upwards by the mass (black arrowhead). B. Coronal T1-WI with fat saturation after intravenous gadolinium injection shows intense, homogeneous enhancement of the mass lesion, but with no extension into soft tissue.

Figure 3  Perioperative view shows a well-circumscribed, encapsulated, pink mass (black arrow) after surgical exposure of the orbital floor. The eye is visible to the right on the photograph (black arrowhead).

Figure 4  Histopathological findings. A. Hematoxylin and eosin staining, original magnification x 200, shows dense spindle-cell proliferation (arrow) with collagen bundles and nuclei exhibiting a wavy pattern. B. Intense and diffuse reactivity with anti-S-100 protein antibody.
A 5-month-old patient presented with drug-resistant epilepsy in which changes in medication failed to control seizures. On being referred for pre-surgical evaluation, the patient received an intravenous injection of 80.17 MBq of $^{18}$F-fluorodeoxyglucose (FDG), 30 min after which integrated positron emission tomography (PET) and computed tomography (CT) was acquired, using a Biograph 16 (Siemens; Erlangen, Germany). Following CT without contrast enhancement (2-mm slices) for anatomical co-registration, attenuation and scatter correction, magnetic resonance imaging (MRI) was performed, using a 3.0-T Magnetom Trio (Siemens).

Arterial spin-labeling (ASL) was also performed with a PASL sequence, using the QUIPSII perfusion mode and the following parameters: 16 slices; voxel size: $3.4 \times 3.4 \times 6 \text{ mm}$; $\text{TA}=5:55 \text{ min}$; $\lambda=0.9 \text{ mL/g}$; $\alpha=95\%$; $\text{TE}/\text{TR}/T1/T2/T12(\text{blood}3\text{T})$ (ms) = 15/5000/700/1800/1496.19. Relative cerebral blood flow (ReCBF) maps for ASL were calculated online by the MRI scanner and offline for CEPWI using Syngo Perfusion (MRI) software. Susceptibility-weighted imaging (SWI) was performed using 3D acquisition with an in-plane resolution of $1 \times 1 \times 1 \text{ mm}$. Diffusion-weighted imaging (DWI) with a 30-directional scan was acquired as well.

MRI demonstrated asymmetry between both hemispheres, with polymicrogyria on the left (Fig. 1). ASL perfusion showed hyperperfusion in the left parieto-occipital regions (Figs. 2 and 3 a, b), while PET/CT showed enhanced glucose utilization in the left parieto-occipital lobes (Fig. 3 c–f).

Epilepsy is a common disorder that is generally managed pharmacologically. However, in a number of cases, simple pharmacological treatment is not sufficient and surgical treatment becomes necessary. Focal lesions can be detected electrophysiologically and clinically, but may be localized with greater precision using high-resolution 3D MRI acquisitions for delineation of anatomical and pathological structures [1]. In addition, other functional methods, such as diffusion and perfusion MRI, have been advocated [2,3]. However, in the very young, it may be necessary to use alternative methods to contrast-enhanced perfusion techniques [4]. One such method is ASL [5], which has been applied in a number of pathological conditions, especially cerebral ischemia [6].

In the case of drug-resistant epilepsy, ictal perfusion changes can be detected by methods such as EEG-triggered PET, using perfusion or metabolic tracers, and single-photon emission CT (SPECT), all of which require radiolabeled compounds. In terms of radiation exposure, MRI perfusion techniques are less invasive, but also require the use of contrast. Indeed, recently, there has been concern over the occurrence of systemic nephrogenic fibrosis with the use of contrast media. Moreover, due to their inherent working properties, these conventional contrast-based MRI perfusion techniques can only reliably demonstrate cerebral blood flow loss, and not increases.

ASL is a promising non contrast-based brain perfusion technique that offers the possibility to CBF mapping of the whole of the brain. While this does not precisely reflect what we see on PET/CT examination—which explores metabolism and not perfusion, as does ASL—the data do correlate surprisingly well, which may also be because of injection of the radiotracer close to the occurrence of an epileptic crisis. This is in agreement with the literature on ASL.

References


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Arterial spin-labeling demonstrates ictal cortical hyperperfusion in epilepsy secondary to hemimegalencephaly

IRM de perfusion par marquage des spins montrant une hyperperfusion corticale dans un cas d’épilepsie secondaire à une hémimegalencéphalie

Key words

Arterial spin-labeling; Epilepsy; MRI; Perfusion

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