Radiology Quiz

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Clinical History

A 10-year-old child presenting a strabismus and an optic atrophy associated with neurofibromatosis. Magnetic resonance exam is shown.

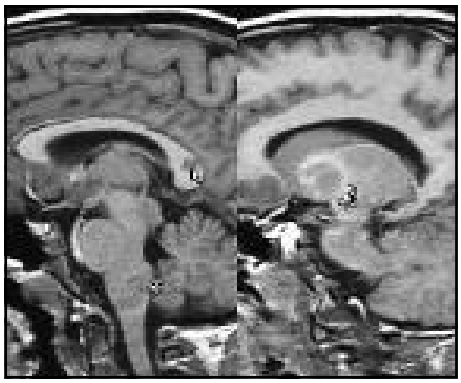

Figure 1a - Sagittal cut; GE-T1w.

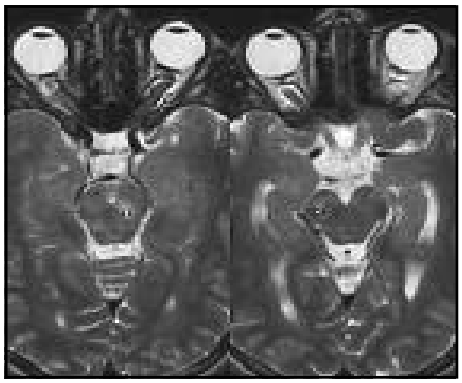

Figure 1b - Axial cut; SE-T2w.

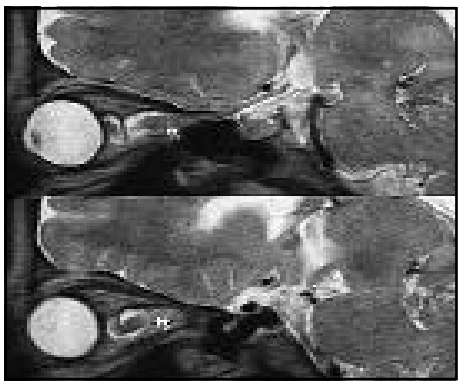

Figure 1c - Sagittal oblique cut; SE-PDw.

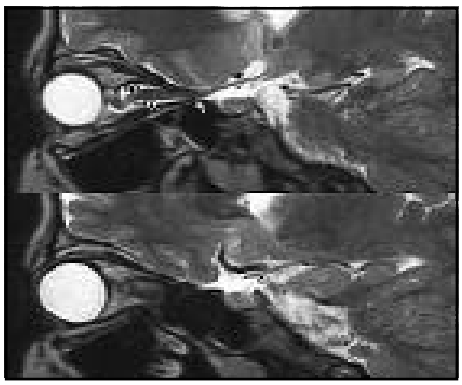

Figure 1d - Sagittal oblique cut; SE-T2w.

Questions

1. What are the radiographic abnormalities?
2. What is the diagnosis?

Answer Page

Radiological findings. Magnetic resonance imaging (MRI) shows a lesion in the globus pallidus, slightly hyperintense on T1w (**Figure 1a, right,**↑↑↑) associated with a hypointense small mass involving the splenium (**Figure 1a, left,** ↑) and medulla with mass effect on the fourth ventricle (**Figure 1a, left,** ↑↑), T2w shows high signal lesion in the brain stem (**Figure 1b,***). The orbital exploration shows enlargement of both intraorbital optic nerves, well shown in axial neuro-ocular plane (**Figure 1b**), as well as on the sagittal oblique cuts, oriented along the optic nerve axis (**Figures 1c and 1d**). The exam performed using the head coil showed that the enlargement of the optic nerve is readily distinguished from prominent perioptic cerebrospinal fluid (CSF) spaces showing very high signal intensity close to the vitreous on T2w sequences (**Figure 1b, left,** ↑↑**; Figure 1d, up, ****). The intraorbital optic nerves showing a hyposignal close brain white matter on T2w, present a somehow fusiform enlargement (**Figure 1c,** ↑↑). The sagittal oblique cuts passing through the optic canals showed that at the left side, the optic nerve tumor extends into the intracranial cavity (**Figure 1c, up,** ↑) with fusiform enlargement of the cisternal portion of the optic nerve at this level but without enlargement of the optic canal itself. The right optic nerve tumor did not involve the cisternal optic nerve (**Figure 1d, ***). Note the kinking of the intraorbital optic nerve well demonstrated on the sagittal oblique cuts.

Note that the apparent enlargement of the perioptic subarachnoid space does not always correspond to CSF trapping but could be due to peritumoral reactive arachnoidal hyperplasia which is rather characteristic of patients with neurofibromatosis. In neurofibromatosis type 1 patients, optic gliomas are generally low grade pilocytic astrocytomas, favoring long term MRI and clinical follow-up and conservative treatment.

Diagnosis. Bilateral optic nerve glioma with arachnoid hyperplasia associated to hamartomas of the globi pallidi.

Discussion. The morphological features of hamartoma are tubular enlargement of the optic nerves which may be fusiform or even eccentric, with kinking and cystic formation (CSF entrapment). Invasion of leptomeningeal spaces, intermixing glial and arachnoid cells. There may be associated arachnoid hyperplasia mimicking meningiomas. They may show frequent extension to the intracanalicular with normal optic canals. Also, cisternal optic nerves and chiasm, optic tracts and radiations, may be involved.

The characteristic MRI findings are SE-T1w: isointense to gray matter, SE-PDw: isointense to gray matter, SE-T2w: isointense to white matter, homogeneous (mainly tubular forms) and heterogeneous of T2w (large fusiform): peripheral high signal intensity due to arachnoid hyperplasia and/or cystic subarachnoid entrapped spaces. Contrast enhancement is unusual in bilateral optic nerve gliomas of the neurofibromatosis type, but it may occur peripherally due to extraneural growth of the glioma, and it helps to differentiate tumor extension from peripheral arachnoid hyperplasia in which is usually not enhanced.

References

1. Tamraz J, Outin C, Forjaz Secca M, Soussi B. MRI Principles of the Head, Skull base and Spine. A clinical approach. Hagerstone, Maryland, New York: Springer-Verlag; 2002. p. 567.