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HYPOPITUITARISM DUE TO HYPOTHALAMIC B-CELL LYMPHOMA

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Key-word: Lymphoma

Background: A 41-year-old male was referred with symptoms highly suggestive of insidious onset global pituitary failure. The patient showed evidence of a developing anterior and posterior pituitary hypofunction (hypopituitarism) that followed a characteristic sequence and which is frequently associated with expanding, clinically non-functioning pituitary adenoma or sellar or parasellar masses. Brain MRI and surgery were performed.

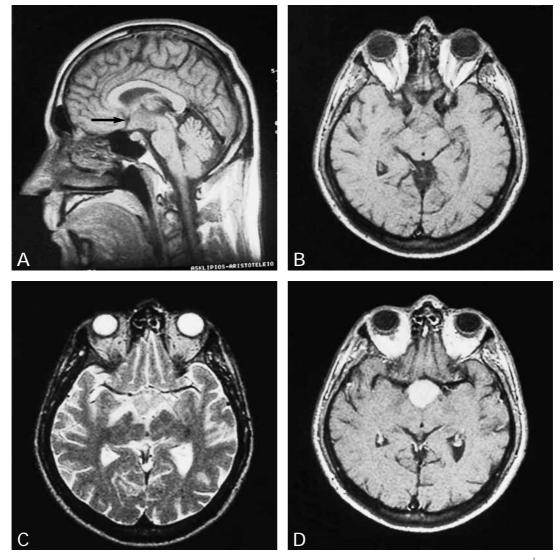


Fig. 1A 1B 1C 1D

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Work-up

MRI of the brain (Fig. 1) shows on axial (A) (arrow) and sagittal (B) T1-weighted MR-images (TR/TE: 700/15) a 3 cm lobulated suprasellarhypothalamic mass with low signal intensity, compressing the optic chiasm. On fast-spin echo T2-weighted MR-images (C) (TR/TE: 5000/100) the mass shows an intermediate signal intensity. The optic tract posteriorly to the mass shows a high signal intensity suggesting edema. Axial and coronal post contrast T1-weighted MR-images (D) show intense almost homogeneous contrast enhancement of the mass.

Radiological diagnosis

Based on the radiological findings, surgery was decided on and pathological examination showed a *B-cell lymphoma* with rounded neoplastic cells in a diffuse pattern.

Discussion

Primary central nervous system (CNS) lymphoma refers to an isolated involvement of the craniospinal axis in the absence of a primary tumor elsewhere in the body. Primary lymphomatous disease of the CNS is now encountered frequently, in both immunocompetent and immunocompromised patients. AIDS is the leading risk factor.

Primary CNS lymphoma may arise from different parts of the brain, with deep hemispheric periventricular white matter being the most common; but the corpus callosum, the cerebellum, the orbits, and the cranial nerves may also be involved. Atypical locations of primary lymphoma include the pineal gland, the pituitary gland, the cavernous sinus, and the hypothalamus. In general, primary CNS lymphoma in an unusual location is more common in patients with AIDS. Our patient had no particular medical history. Clinical evaluation and CT of the thorax and abdomen were normal. The typical MR- findings in primary CNS lymphoma are mass lesions that are iso- to hypointense on T1- and T2-weighted images. The absence of T2 prolongation results from the dense cellularity and high nucleus-to-cytoplasm ratio of lymphoma, and it may help in the differentiation of primary CNS lymphoma from other brain tumors.

Typically, contrast enhancement is intense and homogeneous in immunocompetent patients, but often is inhomogeneous or ringlike in immunocompromised patients.

The differential diganosis of a hypothalamic mass includes hypothalamic glioma, germinoma, hypotha-lamic hamartoma, germ cell tumor, lymphocytic infundibulo-neurohypophysitis, craniopharyngioma, sarcoidosis, Langerhans cell histiocytosis, meningioma of the diaphragma sellae and pituitary adenoma.

Hypothalamic glioma tends to be solid with microcyst formation and is iso- or hypointense on T1-weighted images, hyperintense on T2-weighted images and demonstrates enhancement with contrast. Hypothalamic hamartoma presents as precocious puberty in a young child with a nodular mass in the suprasellar cistern. The mass is iso-intense with normal brain on T1-weighted images and isointense or mild hyperintense on T2-weighted images. These lesions usually do not enhance after contrast administration.

Sellar and suprasellar germinoma appears as a well-marginated lobulated homogeneous tumor with prolonged T1 and T2 relaxation times which strongly enhances after gadolinium administration. The presence of these imaging findings along with the presence of diabetes insipidus and a suprasellar mass is a clue to the diagnosis of germinoma.

When a lesion of the sellar and juxtasellar region is depicted which is iso- to hypointense on T1- and T2-weighted images and enhances strongly with contrast, lymphoma should be considered in the differential diagnosis in both immunocompetent and immunocompromised patients.

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