JBR–BTR, 2010, 93: 196-197.

INTRACRANIAL GERM CELL TUMOR

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Germ cell tumours represent about 3 to 8% of pediatric brain tumours. Occurrence of diabetes insipidus is common in the case of suprasellar germ cell tumors. The diagnosis may be advanced by MRI owing to the location and relatively univocal characteristics of the lesion signal. The existence of a bifocal mass developed in both suprasellar region and pineal zone is highly suggestive of a germinoma. The most important notion is to recognize that at the time of diabetes insipidus diagnosis in a child, the cerebral mass might be too small to be identified by MRI. In such patients, repeating imaging study should be obtained.

Key-word: Brain neoplasms, in infants and children.

Case report

A 12-year-old-girl was admitted at the emergency department because of areactive mydriasis associated with oculomotor paresis of the left eye. Six month earlier, she was explored by brain MR for hypotrophy and occurrence of diabetes insipidus. At this time, a suprasellar mass was evidenced and a biopsy was undertaken which led to the diagnosis of lymphocytic hypophysitis (Fig. 1). Actually, a new brain MR demonstrated a multifocal insult reaching both the suprasellar zone and the pineal gland (Fig. 2). On such an imaging basis, the previous diagnosis was revisited to finally conclude to the existence of an intracranial germ cell tumour.

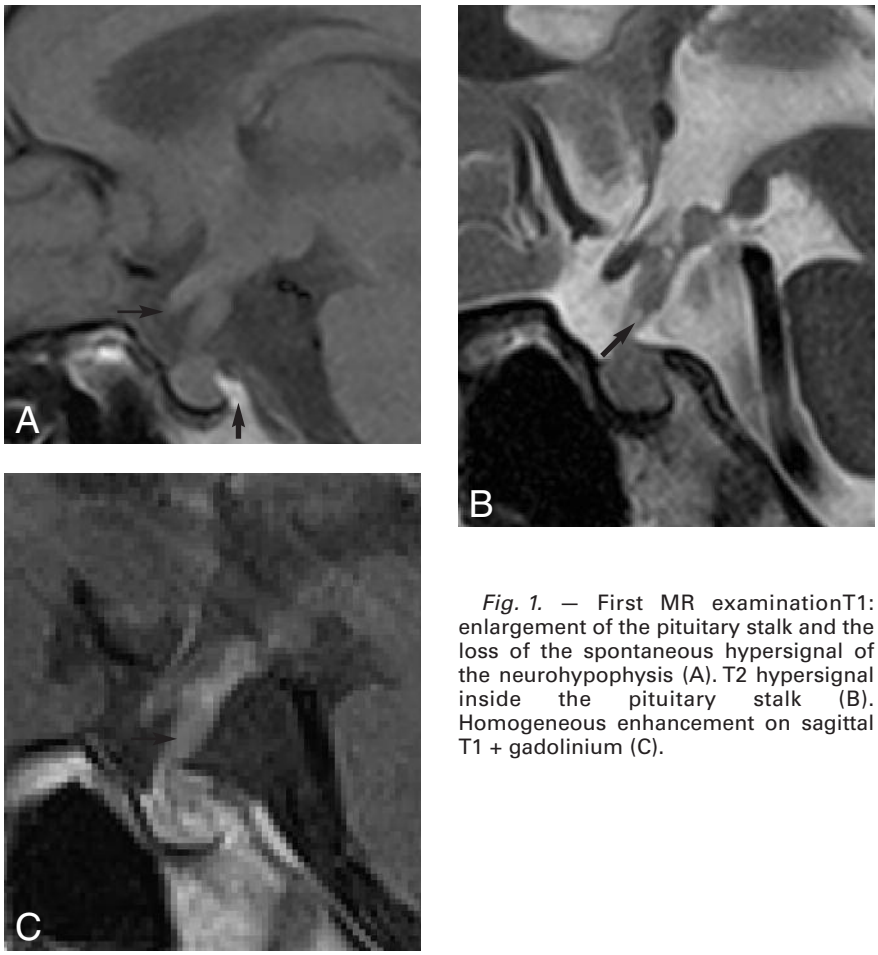

Fig. 1. — First MR examinationT1: enlargement of the pituitary stalk and the loss of the spontaneous hypersignal of the neurohypophysis (A). T2 hypersignal inside the pituitary stalk (B). Homogeneous enhancement on sagittal T1 + gadolinium (C).

Discussion

Differential diagnosis of the suprasellar masses in a child with diabetes insipidus includes several hypothesis including craniopahryngioma, germ cell tumour, Langerhans' cell histiocytosis, lymphocytic hypophysitis and granuloma (sarcoidosis or tuberculosis) (1). However, our findings of an insult interesting simultaneously the suprasellar zone and the pineal region, reduce the field of pathological hypothesis. Morphology and localization of the multifocal process rather suggest the diagnosis of brain *bifocal germinal tumour*. On this basis, the biopsy specimen was revisited and confirmed the diagnosis of germinal tumour in spite of absence of blood tumoral markers.

Germ cell tumours represent about 3 to 8% of pediatric brain tumours (2). Up to 60% of the pineal proliferations are germinoma. Almost 30% of these neoplasms develop in the suprasellar-hypothalamic region. Less common locations include the basal ganglia, C-P angle, cerebellum, corpus callosum and the spinal cord. Both suprasellar and pineal gland involvement at the time of diagnosis represent 5%-10% of cases (2). Of germ cell tumours, about 65% are germinomas, 16% are teratoma, 6% are embryonal cell carcinoma, 4% are choriocarcinoma and 9% are mixed germinal tumour.

The diagnosis may be advanced by MRI owing to the location and relatively univocal caracteristics of the lesion signal. The existence of a bifocal mass developed in both suprasellar region and pineal zone is highly suggestive of a germ-cell tumour and most commonly a ger-

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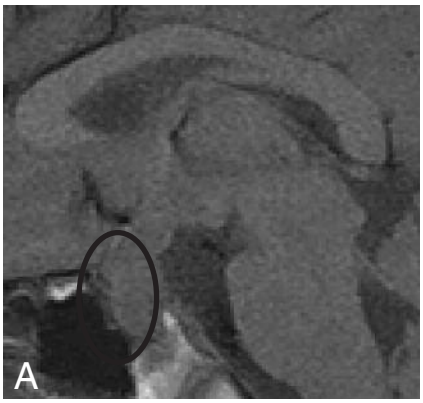

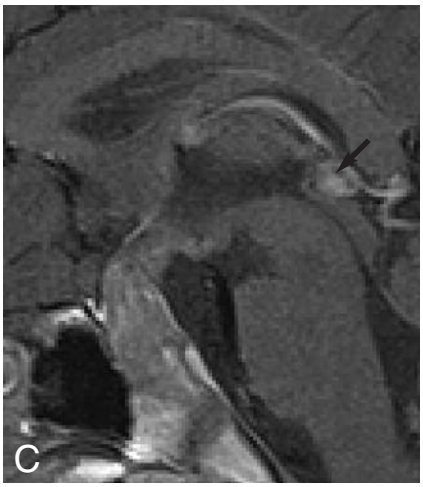

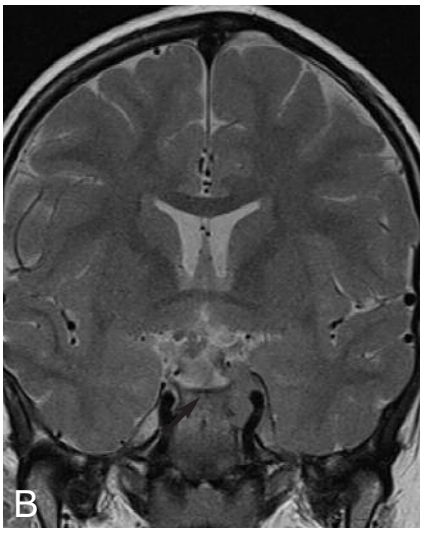

Fig. 2. — Second MR examinationT1 (A) and T2 (B): tumoral growth with extension to the supra-sellar region. T1 +gado reveals a pineal mass (C).

minoma (3). The imaging varies with histology. Germinoma are usually well-marginated tumours, either round or lobulated, that demonstrate iso to hyperintensity on T1WI and iso to hyperintensity on T2WI. A relatively hypointensity image on T2WI in a suprasellar or a pineal region in an adolescent is highly suggestive of such tumour. Enhancement is usually homogeneous but some internal heterogeneity is often seen in large germinomas (4). The lack of physiological hyperintensity of the neurohypophysis is not caracteristic of one type of lesion in particular but is correlated with the presence of a diabetes insipidus. The most important notion is to recognize that at the time of diabetes insipidus diagnosis in a child, the cerebral mass might be too small to be identified by MRI. In such patients, repeating imaging study should be obtained within 3 to 6 months and, if still negative, next exploration should be conducted after another run of 3 to 6 months (4). Stereotactic biopsy is obviously indicated even if the specimen analysis remains hard to give a definetly ruling on diagnosis (5).

Therapeutical protocol of germ cell tumours consists in chemotherapy followed by radiotherapy. Survival at 5 years of the germinoma is actually beyond 91% (2).

Accordingly, our patient was submitted to a protocol based first on VP16, Ifosfamid and Mesna and secondly on carboplatin associated with VP16. This chemotherapy was finally followed by radiotherapy focused on the suprasellar lesion. Based on this cure, the ultimate MR showed a dramatic reduction in the volume of both the pituitary stalk and the pineal mass.

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