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Ectopic intracranial germinoma

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ABSTRACT

Intracranial ectopic germinomas are often associated with synchronous midline disease. Germinomas involving the corpus callosum are exceedingly rare. The reported imaging appearance is not as varied as one might expect and a review of the literature reveals a few common imaging features amongst most ectopic lesions, including cyst formation. We report a 24-year-old man with panhypopituitarism. Neuroimaging revealed three enhancing lesions involving the pituitary infundibulum, the pineal region, and a parenchymal lesion involving the genu of the corpus callosum. The described ectopic mass, a parenchymal lesion, was associated with small peripheral cysts. Stereotactic biopsy and histopathological evaluation revealed this mass to be a germinoma. Following chemotherapy and radiation therapy, there was near-total resolution of the intracranial disease. Preoperative imaging plays an important role, not only in delineating the extent of disease, but also in assisting in generating an appropriate differential diagnosis. Germinomas in the corpus callosum are exceedingly rare but should be considered in the differential of any young patient with a characteristic cystic and solid intra-axial mass.

1. Introduction

Primary intracranial germ cell tumors are a rare heterogenous group of neoplasms that preferentially affect children and adolescents. The World Health Organization classifies these lesions into six histologic subtypes: germinoma, teratoma, embryonal carcinoma, choriocarcinoma, yolk-sac tumor and mixed tumor [1]. Of these, germinomas comprise nearly two-thirds of all intracranial germ cell tumors.

Germ cell tumors of the central nervous system are rare and usually arise from midline structures [2]. The pathogenesis is not well understood but it has been hypothesized as the result of the mismigration of primordial germ cells from the midline embryonic disk [3]. Sano suggested that of all subtypes of germ cell tumors, germinomas may be the only ones to originate from primordial germ cells [4]. The most common sites of occurrence include the pineal and neurohypophyseal regions, but when located outside of these regions, it is considered ectopic. Reported ectopic sites included the basal ganglia, thalamus, corpus callosum, septum pellucidum, lateral ventricles, temporal lobe and cerebellum [5,2]. It is reported that intracranial germinomas of the corpus callosum are exceedingly rare and the incidence is as low as 0.7% [5,2].

We report a patient with an intracranial germinoma of the corpus callosum with synchronous disease of the neurohypophysis and pineal region.

2. Case report

A 24-year-old man who was otherwise healthy developed progressive insatiable thirst, urinary frequency, impotence, muscle

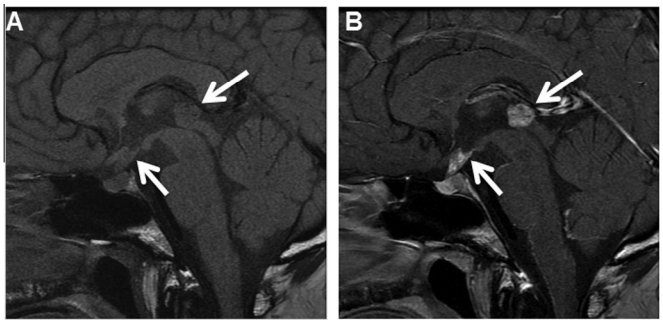

Fig. 1. Preoperative midsagittal pre-contrast (A) and post-contrast (B) T1-weighted MRI showing marked thickening and enhancement of the pituitary infundibulum. A heterogenously enhancing rounded soft tissue mass is present in the pineal region (arrow).

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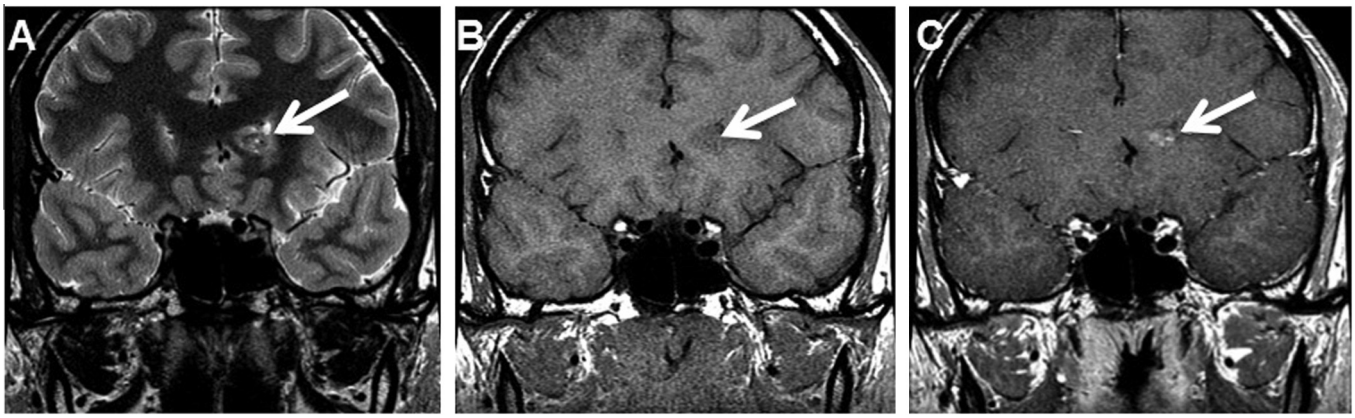

Fig. 2. Preoperative coronal T2-weighted (A) and pre-contrast (B) and post-contrast (C) T1-weighted MRI showing a focal, heterogeneous T2 hyperintense lesion in the genu of the corpus callosum on the left with small associated peripheral cysts. Heterogenous enhancement is noted of the solid component (arrow).

aches and fatigue. The patient was referred to an endocrinologist and subsequently diagnosed with panhypopituitarism.

MRI was performed as part of the work up for the patient's pituitary insufficiency. Post-contrast T1-weighted imaging revealed three avidly enhancing intracranial lesions in the pituitary infundibulum, pineal region and left corpus callosum (Fig. 1, 2). Coronal T2-weighted imaging shows a heterogeneous, T2 hyperintense lesion in the genu of the corpus callosum on the left with small peripheral cysts (Fig. 2). The initial suggested differential diagnosis included sarcoidosis, chronic basal meningitis, and possibly germ cell tumor. Follow-up imaging approximately 6 weeks later revealed slight interval enlargement.

Stereotactic biopsy was performed of the left corpus callosal lesion and histological examination demonstrated a pure germinoma, which is a cellular biphasic lesion composed of two different cell types. One cell type comprises large, undifferentiated cells with round vesicular nuclei, prominent nucleoli, discrete cell membranes and relatively abundant cytoplasm. These large germinoma cells represent the main tumor cells, form monomorphous sheets and are immunoreactive for octamer-binding transcription factor 4 and placental alkaline phosphatase. Intermixed are numerous mature, reactive T-lymphocytes, which are identified by staining for CD3. Additional histological evaluations and stains for alpha-fetoprotein, creatine kinase 30 and beta- human chorionic gonadotropin did not demonstrate other germ cell elements and hence excluded a mixed phenotype (Fig. 3).

The patient received chemotherapy followed by radiation therapy. Post-therapeutic MRI at 3 and 6 months revealed a normal caliber pituitary infundibulum, resolution of the pineal region soft tissue mass and resolution of the abnormal enhancement at all three sites (Fig. 4).

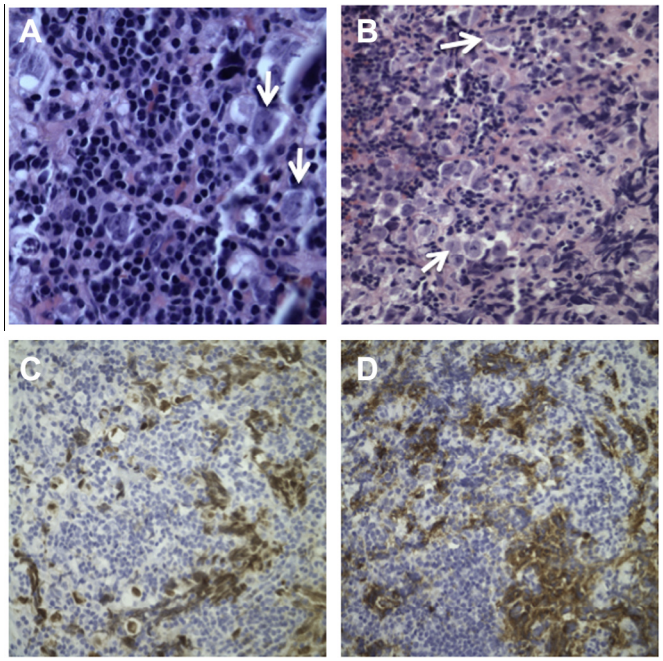

Fig. 3. Histological sections. 40× (A) and 40× (B) are hematoxylin and eosin stained sections, which show a biphasic lesion composed of large germinoma cells (arrows) and numerous small reactive T-lymphocytes. The germinoma cells are highlighted by staining for octamer-binding transcription factor 4 (C) 20× and placental alkaline phosphatase (D) 20×.

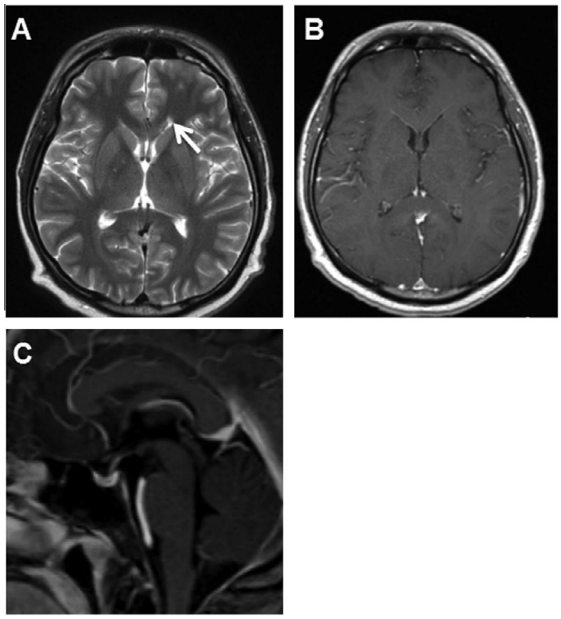

Fig. 4. Post-therapeutic axial T2-weighted (A) and axial (B) and midsagittal (C) T1-weighted post-contrast MRI showing minimal residual T2 hyperintensity and cystic change in the genu of the corpus callosum (arrow). There is complete resolution of abnormal enhancement at all three sites. Note that the pituitary infundibulum is normal in caliber and the pineal region soft tissue mass has resolved.