Transient regression of an intracranial germ cell tumour after intravenous steroid administration: a case report

Mario Mascalchi, Federico Roncaroli, Fabrizio Salvi, Giorgio Frank

Abstract
Magnetic resonance imaging showed transient regression of the lesion after intravenous steroid administration in a patient with intracranial multifocal germ cell tumour. Prominent lymphocyte infiltration of the tumour was seen at histological examination and presumably accounts for the regression. Germ cell tumour must be included in the differential diagnosis of intracranial mass lesions sensitive to steroids. (J Neurol Neurosurg Psychiatry 1998;64:670–672)

Keywords: germ cell tumour; steroid therapy

Intracranial germ cell tumours are neoplasms of children and young adults, histologically identical to those seen in the gonads.1 They occur in pineal and suprasellar regions and, rarely, in basal ganglia and thalami.2–5 Imaging features of germ cell tumours are indistinguishable from those of CNS lymphomas and neurosarcoidosis,6 and biopsy is necessary for differential diagnosis. However, it has been suggested7–9 that regression after intravenous steroid administration of a contrast enhanced mass abutting the CSF spaces is an indirect clue to the possibility of CNS lymphoma or sarcoidosis. Germ cell tumours are known to be very sensitive to radiation and chemotherapy.2–3,9 We report a patient with a multifocal intracranial germ cell tumour in whom transient regression of the lesion after intravenous steroids deferred stereotaxic biopsy and appropriate treatment.

Case report
An 18 year old immunocompetent woman was admitted to hospital in June 1993 because of persistent polydipsia and galactorrhea. Laboratory investigations established a diagnosis of diabetes insipidus, hyperprolactinaemia (40 ng/ml, normal<20 ng/ml), and subclinical hypothyroidism due to low TSH secretion. Brain MRI showed two lesions, localised in the pituitary stalk and below the head of the left caudate nucleus, which enhanced after contrast administration (fig 1 A). The lesions were iso-intense to the cerebral grey matter and surrounded by an irregular hyperintense rim in proton density and T2 weighted images. Chest radiography and gallium scanning were normal, excluding pulmonary sarcoidosis. No cutaneous lesions were present. Serum angiotensin converting enzyme (ACE) concentration was 143 U/l (normal<40 U/l) and the patient received a tentative diagnosis of neurosarcoidosis for which 20 mg/day prednisone was started. Brain MRI three months later showed substantial regression of the two lesions (fig 1 B). Steroids were interrupted in September 1994. She did well until September 1995 when she was readmitted because of confusion, headache, and vomiting. Laboratory examinations disclosed diabetes insipidus, global hypopituitarism, and increased serum ACE (221 U/l). Brain MRI in October 1995 disclosed that the enhanced lesions had considerably increased in size, obliterated the third ventricle and left foramen of Monro, and extended along the walls of the lateral ventricles (fig 1 C). Histological examination of a stereotaxic biopsy of the larger mass showed large polygonal cells with eosinophilic cytoplasm and atypical nuclei of different size with coarse chromatin and prominent nucleoli. These cells were intermingled with a diffuse infiltrate of mature lymphocytes (fig 2). No granulomas were seen. Staining with PAS-diastase did not show any cytoplasmic globules within the polygonal cells. The neoplastic cells were intensely reactive to anticytokeratin antisemur, whereas other reactions, including antiplacental alkaline phosphatase (anti-PLAP), anti-human β-chorionic gonadotrophin, and anti-α-fetoprotein were negative. Lymphocytes showed immunophenotype ‘T’ as they reacted to anti-UCHL1 (CD45RO) antisemur. Overall, the histological and the immunocytochemical features were consistent with those of an embryonal carcinoma. The patient was treated with chemotherapy and radiation therapy, and MRI four, eight, and 16 months later showed considerable size reduction of the lesions without contrast enhancement.
Intracranial germ cell tumours are multiple in up to 45% of cases. Their MRI features are similar to those of CNS lymphoma and neurosarcoidosis, and include lesions of intermediate or high signal intensity in T2 weighted images which usually abut the CSF spaces and densely enhance after contrast administration.

Clinical and MRI features at presentation in our patient narrowed the differential diagnosis to neurosarcoidosis, CNS lymphoma, or germ cell tumour. Because of increased serum ACE, a tentative diagnosis of neurosarcoidosis was initially favoured, and this seemed corroborated by lesion regression after intravenous administration of steroid. This is a well known phenomenon in neurosarcoidosis, but has never been described in germ cell tumour. Recurrence of the lesions after interruption of steroids in our patient prompted biopsy of the mass, which disclosed a germ cell tumour. Germ cell tumours are classified according to histological and immunocytochemical findings as two fundamental types—germinoma and non-germinoma—which have different prognoses and require different treatment protocols. The non-germinoma types include embryonal carcinoma, yolk sac tumour, choriocarcinoma, and immature teratoma. Histological features in our patient were consistent with the two cell (large polygonal cells and lymphocytes) pattern typical of germinoma, and we submit that the diffuse lymphocyte infiltration may account for the transient lesion regression after treatment.
with steroid. Moreover, as the intense lymphocytic reaction which is often seen in germinomas is thought to be protective, we speculate that destruction of lymphocytes by steroids may actually have facilitated tumour growth in our patient. A diagnosis of embryonal carcinoma was made based on the high variation in size and shape of the atypical nuclei, the positivity of all the neoplastic cells to anticytokeratin antiserum, and the negative reaction to anti-PLAP antiserum.\textsuperscript{11–14} Moreover, a reactive infiltrate of T-lymphocytes may be present in embryonal carcinoma, but is absent in other less differentiated variants of germ cell tumour including yolk sac tumour and chorioncarcinoma.\textsuperscript{15} Due to the diagnosis of embryonal carcinoma and extension of the lesion along the ventricular system, a combined chemotherapy and radiation therapy approach was preferred in our patient to radiation therapy alone.\textsuperscript{13–19}

In conclusion, our finding indicates that germ cell tumour has to be added to CNS lymphoma and neurosarcoidosis in the list of intracranial mass lesions responsive to steroids and reinforces the view that biopsy of these lesions is mandatory for appropriate therapy.

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