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ORIGINAL ARTICLE

Intracranial germ cell tumours: A 21-year review☆

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KEYWORDS
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Abstract
Introduction: Intracranial germ cell tumours are rare in children. They are a heterogeneous group of neoplasms that show different clinical manifestations despite having a common origin.
Patients and methods: A retrospective analysis was carried out on the epidemiological and histological characteristics, clinical manifestations, and outcomes of 20 patients diagnosed with intracranial germ cell tumours in the Niño Jesús Children's Hospital of Madrid from 1994 to 2014.
Results: A total of 20 patients were identified: 14 boys and 6 girls. The mean age was 11.1 years (range 2–18 years). Histological confirmation of the diagnosis was obtained in 95% of the patients. Of the 20 patients, 14 were pure germinoma (70%) and 6 non-seminomatous germ cell tumours (30%). The most frequent locations were pineal (45%) and suprasellar (45%). The most frequent clinical symptoms in pineal tumours at diagnosis were headache and vomiting (77.77%), followed by visual disturbances (44.4%). In suprasellar tumours it was polydipsia and polyuria (100%). At diagnosis, 90% of the patients received radiotherapy, and 55% received chemotherapy combined with radiotherapy. There was a relapse in 4 patients (20%), and 3 of them died. Overall survival was 80%; 85.7% for pure germinomas and 60% for non-seminomatous germ cell tumours.
Conclusions: The most common histological subtype was pure germinoma. Germ cell tumours include heterogeneous disease entities that have a variable prognosis. Thus, an accurate diagnosis is vital for patient counselling and treatment planning.

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PALABRAS CLAVE
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Tumores germinales intracraneales: revisión de 21 años

Resumen
Introducción: Los tumores de células germinales intracraneales son un grupo poco frecuente de tumores en niños. Comprenden un grupo heterogéneo de neoplasias, que aunque comparten un origen común, presentan comportamientos clínicos muy diferentes.
Pacientes y métodos: Análisis retrospectivo de las características epidemiológicas e histológicas, las manifestaciones clínicas y la evolución de 20 pacientes diagnosticados de tumor de células germinales intracraneal en el Hospital Infantil Universitario Niño Jesús de Madrid durante los años 1994-2014.
Resultados: Se obtuvieron 20 pacientes: 14 niños y 6 niñas. La edad media fue de 11,1 años (rango 2-18 años). Se realizó confirmación histológica en el 95% de los pacientes. De los 20 pacientes, 14 fueron germinomas puros (70%) y 6 tumores de células germinales no germinomas (30%). Las localizaciones más frecuentes fueron pineal (45%) y supraselar (45%). Los síntomas más frecuentes en el momento del diagnóstico en los tumores de localización pineal fueron cefalea y vómitos (77,77%), seguido de alteraciones visuales (44,4%), y en los tumores de localización supraselar, polidipsia y poliuria (100%). En el momento del diagnóstico recibieron radioterapia el 90% de los pacientes y quimioterapia asociada a la radioterapia el 55%. Presentaron recaída tumoral 4 pacientes (20%), de los cuales 3 fallecieron. La supervivencia global fue del 80%, siendo un 85,7% para los germinomas y un 60% para los no germinomas.
Conclusión: El tipo histológico más frecuente fue el germinoma. Los tumores de células germinales son un grupo heterogéneo de tumores que conllevan un pronóstico diferente, por lo que un adecuado diagnóstico y estadificación es fundamental para planear el tratamiento.

Introduction

Intracranial (IC) germ cell tumours (GCTs) are a rare and heterogeneous group of tumours found in adolescents and young adults. Their incidence varies widely between geographical regions, and they amount to 2–4% of brain tumours in individuals aged 0–19 years in Europe.[1,2]

Germ cell tumours arise from primordial germ cells that lodge ectopically in the central nervous system (CSN) during the migration from the yolk sac to the gonadal ridge and may undergo malignant transformation. Different tumours develop based on the degree of differentiation of these germ cells at this point.

Germ cell tumours that arise in the CNS share histological, genetic and clinical characteristics with extracranial GCTs. The World Health Organization (WHO) has divided GCTs into two broad groups: germinomatous and nongerminomatous GCTs (NGGCTs)[3,4] based on histology and the presence of tumour markers (Tables 1 and 2). Other classification schemes have divided GCTs into secreting and nonsecreting tumours. Secreting tumours have alpha-fetoprotein (AFP) positivity or beta-human chorionic gonadotropin (BHCG) elevations greater than 50 IU/L in the cerebrospinal fluid (CSF) or any serum BHCG or AFP positivity[5,6] (Table 1).

The clinical presentation varies based on the location and size of the tumour. The most frequent location is the pineal region, followed by the suprasellar region.[7] The most common presenting symptoms are endocrine abnormalities, signs of raised intracranial pressure and visual disturbances.

Table 1 Classification of intracranial germ cell tumours proposed by the WHO.

Germ cell tumours
Germinoma
Nongerminomatous germ cell tumours
Embryonal carcinoma
Yolk sac tumour
Choriocarcinoma
Teratoma
Benign teratoma
Immature
Mature
Teratoma with malignant transformation
Mixed germ cell tumour

From: Louis et al., editors.[3]

Accurate staging and histological examination are important factors for the classification of patients into prognostic groups.[4,8] Imaging studies cannot differentiate GCTs from other tumours, so diagnosis requires histological confirmation except in cases with elevated levels of tumour markers such as AFP and BHCG.[3] Surgical management options are determined by tumour location. Traditionally, these tumours were treated with craniospinal irradiation. In the past two decades, advances in imaging techniques, radiotherapy (RTX) and surgery, and the addition of chemotherapy to treatment protocols have led to a significant improvement in the prognosis of malignant GCTs, and especially