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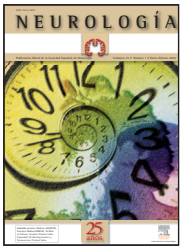

ORIGINAL ARTICLE

# Subependymal giant cell astrocytoma in tuberous sclerosis complex. A presentation of eight paediatric patients

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**KEYWORDS**
Subependymal giant-cell astrocytoma;
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Blindness;
Headache;
Hydrocephalus

**Abstract**
*Objective:* Presentation of 8 patients with subependymal giant-cell astrocytomas (SGCA) associated with tuberous sclerosis complex (TSC).
*Material and methods:* There are 8 patients, 6 males and 2 females with TSC, who presented with the tumour between the neonatal period and 24 years.
*Results:* All patients showed bilateral hypersignalised areas in zones close to the foramen of Monro. Three of the patients were admitted urgently due to blindness and increased intracranial pressure. Incomplete removal of the tumour has always been bad solution as it resulted in the death of the patient (in one case) or further surgery operation in the short term. Only one patient developed the tumour suddenly from pre-existing subependymal nodules from the childhood and they had to be removed at 24 years of age. By contrast, 32 patients with TSC and images of subependymal nodules whose CT or MR progress was followed up for between 10 and 30 years did not develop a tumour. One patient had to be operated four times over 20 years.
*Conclusions:* SGCA associated with TSC is a severe complication which as likely to develop and careful monitoring is required from neonatal age with periodic-clinical and imaging studies in order to avoid its irreversible complications. Hydrocephaly, blindness and even the death can be the main consequences. Reintervention of the recurrent tumour is often necessary.


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**PALABRAS CLAVE**
Astrocitoma subependimario de células gigantes;
Complejo de esclerosis tuberosa;
Ceguera;
Cefalea;
Hidrocefalia

**Astrocitoma subependimario de células gigantes en el complejo de esclerosis tuberosa. Presentación de ocho pacientes infantiles**

**Resumen**
*Objetivo:* Presentar 8 pacientes con astrocitomas subependimarios de células gigantes (ASGC) en relación con el complejo de esclerosis tuberosa (CET).
*Material y métodos:* Ocho pacientes, 6 varones y 2 mujeres, con CET, que desarrollaron el tumor entre la etapa neonatal y los 24 años.
*Resultados:* Todos mostraban áreas localizadas bilaterales de hiperseñal, en zonas próximas a los *foramina* de Monro. Tres ingresaron urgentemente con ceguera e hipertensión intracraneal. La extirpación parcial del tumor fue siempre una mala solución ya que acabó en reintervenciones a corto, medio o largo plazo o en la muerte de un paciente. Sólo en un caso vimos desarrollarse el tumor desde las zonas de hiperseñal subependimaria a partir de la preadolescencia para acabar en extirpación a los 24 años, mientras que 32 pacientes a los que se siguió la evolución de estas zonas de hiperseñal entre 10 y 30 años no desarrollaron tumor. Un paciente tuvo que ser operado cuatro veces a lo largo de 20 años por recidiva del tumor; se extirpó otro ASGC en el lado contralateral al mismo tiempo de la cuarta intervención en el lado del tumor primitivo. Otros 2 pacientes también mostraron recidiva y tuvieron que ser reintervenidos del tumor.
*Conclusiones:* El ASGC en relación con CET es una complicación grave cuya posibilidad de desarrollo hay que controlar cuidadosamente desde la época neonatal, con estudios periódicos clínicos y de imagen, para evitar sus complicaciones irreversibles. La hidrocefalia, la ceguera e incluso la muerte pueden ser sus consecuencias. La reintervención de tumores recidivados a menudo es necesaria.


## Introduction

The term "Subependymal giant cell astrocytoma" (SGCA) was coined by Russell et al[1] to differentiate it from other types of intracranial neoplasms, as it had previously received numerous names, such as astrocytoma, ependymoma, spongioblastoma and possible ganglioglioma. It is a benign tumour that has thick fibres and large round or oval nuclei in its structure. Most patients who present this tumour show clinical and pathological symptoms between 8 and 19 years of age.[2,3] However, there are published cases that took place at later,[4] and especially at earlier[5] ages, many of them diagnosed in the prenatal[7] or neonatal[6] age and even as early as 19 weeks of gestation. It can take place with no relation to family history of tuberous sclerosis complex (TSC),[8] as these would be sporadic cases. It is considered that the criteria to be met by subependymal giant cell astrocytoma associated to TSC for preoperative diagnosis are: injury in the area of one or both foramina of Monro, size > 5 mm and incomplete calcification.[9-11] Once these criteria have been established, excision should take place as soon as possible.[11]

Although this is a histologically benign tumour, it may have behaviour and evolution that are not so benign, carrying severe problems and even death. In this study, we show the experience obtained with this tumour in a neurology-neurosurgery paediatric unit over 39 years (1965-2004).

## Material and methods

We conducted a retrospective study in a series of 160 paediatric patients (aged under 16 years) who attended the Paediatric Neurology Service at La Paz Teaching Hospital in Madrid, between August 1965 and June 2004, and who underwent surgery when it was required. In addition to the clinical neurological and genetic history, especially seeking signs consistent with TSC diagnostic criteria, we performed a cranial radiological study: computed tomography (CT) and/or magnetic resonance imaging (MRI) with and without contrast on all patients. The study was repeated several times in all cases, with a frequency ranging from once every 6 months to once every 3 years, depending on the size of the hyperintense areas or intracranial hypersignal, especially those located in the heads of the caudate nuclei, close to the foramina of Monro. When growth was observed in these areas, the image was enhanced with intravenous contrast (gadolinium); when changes in size and density or signal were observed (thus making the tumour evident), usually MRI spectroscopy was performed to analyse if there were signs of histological malignancy. An imaging study was performed in all cases, specifically MRI in recent years. The ages of the patients were between newborn and 30 years (the latter had hyperintense areas of considerable size and confirmation that they had not changed since the previous year was necessary through annual controls).