Subependymal giant cell astrocytoma with atypical clinical and pathological features: a diagnostic pitfall

Marián Švajdler jr.[1], Ladislav Deák[2], Boris Rychlý[3], Peter Talarčík[3], Lucia Fröhlichová[1]

[1]Department of pathology, L. Pasteur's University Hospital, Košice, Slovakia
[2]Department of pediatric oncology and haematology, Children's University Hospital, Košice, Slovakia
[3]Cytopathos s.r.o., Bratislava, Slovakia

SUMMARY
Subependymal giant cell astrocytoma (SEGA) is benign, slowly growing tumor linked to the tuberous sclerosis complex. It almost always occurs near the foramen of Monro. Parenchymal extension and worrisome histological features, such as necrosis, mitoses, microvascular proliferation and pleomorphism are unusual in these tumors, but can occur rarely. A case of SEGA is presented, in a patient with no signs of tuberous sclerosis so far, with atypical imaging findings and areas of necrosis found microscopically. These worrisome features initially led to the false diagnosis of glioblastoma. The differential diagnosis of SEGA is discussed.

Keywords: subependymal giant cell astrocytoma – atypical – necrosis

Subependymálny obrovskobunkový astrocytóm s atypickými klinickými a patologickými črtami: diagnostická pasca

SÚHRN
Subependymálny obrovskobunkový astrocytóm (SEGA) je benígny pomaly rastúci tumor asociovaný so syndrómom tuberóznej sklerózy. Vyskytuje sa takmer výlučne v oblasti foramen Monro. Šírene do parenchýmu hemisféry a znepokojujúce histologické črty ako sú nekrózy, mitózy, mikrovaskulárna proliferácia a pleomorfia sú nezvyčajné, ale vzácne môžu byť prítomné. Prezentujeme prípad SEGA u pacienta, u ktorého zatiaľ nie sú prítomné ďalšie známky syndrómu tuberóznej sklerózy, s atypickým radiologickým nálezom a mikroskopicky prítomnými nekrózami. Tieto znepokojujúce črty iniciálne viedli k nesprávnej diagnóze glioblastómu. Je diskutovaná diferenciálna diagnóza SEGA.

Kľúčové slová: subependymálny obrovskobunkový astrocytóm – atypický – nekróza

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Subependymal giant cell astrocytoma (SEGA) is benign, WHO grade 1, slowly growing tumor linked to the tuberous sclerosis complex (TSC), which forms an expansive mass in the wall of the lateral or third ventricle, almost always near the foramen of Monro (1–3). Parenchymal extension and worrisome histological features, such as necrosis, mitoses, microvascular proliferation or pleomorphism, are unusual in these tumors, but can occur (4). We present an unusual case of SEGA in a patient with no signs of TSC so far, which formed a large solid and cystic parenchymal mass, with a shift of the midline structures and microscopically showed areas of necrosis. These worrisome features led to the false diagnosis of glioblastoma.

CASE REPORT

1.5-year-old girl was admitted to the hospital in June 2008 because of walking difficulties and febrility. At admission, signs of intracranial hypertension were found. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a large expansive tumor in the left fronto-parieto-temporal region, with a shift of the midline structures and hydrocephalus. Gadolinium enhanced MRI showed non-homogenous, predominantly peripheral ("ring") contrast enhancement of the tumor (Fig. 1). Partial resection of the tumor was performed (Fig. 2).

Microscopically, the tumor was composed of large gemistocyte-like cells with prominent excentric nuclei, some with prominent nucleoli. Binucleated cells resembling dysplastic ganglion cells could also be seen. The cytoplasm was fibrillary to glassy and the growth of the tumor was solid and expansive. Mitoses were hard to find and microvascular proliferation was not present. However, large areas of geographic necrosis without pseudopalisading were found (Fig. 3).

By immunohistochemistry, most of the neoplastic cells were GFAP positive (clone 6F2, DiagnosticBioSystems), with patchy neurofilament protein expression (clone 2F11, DiagnosticBioSystems) (Fig. 4). Neural filaments were not stained in the background, confirming the non-infiltrative growth pattern. The Ki-67 labeling index (clone MIB-1, DAKO) was very low, approximately 1 %. P53 (clone DO-7, Neomarkers) focally and faintly stained some of the nuclei (< 2 %, *not shown*).

At the time of sign-out, no further clinical data, including the results of imaging were available to the pathologist. Despite the presence of necrosis, the case was signed-out as SEGA, WHO grade 1.

After the sign-out, clinicians asked for a second look opinion, because of atypical and worrisome imaging characteristics of the tumor and the presence of the necrosis, not quite consistent with

✉ ***Correspondence address:***
Marián Švajdler jr., M.D.
Department of Pathology, L. Pasteur's University Hospital
Trieda SNP 1, 041 90 Košice, Slovakia
Tel/Fax: +421 55 640 2945
e-mail: svajdler@yahoo.com