

Unusual Clinical and Neuroimaging Findings in a Child with Subependymal Giant Cell Astrocytoma (SEGA) in Sellar Position

M. Soussou^{1*}, G. El Mghari², N. El Ansari²

¹Department of Endocrinology, Mohamed VI University Hospital of Marrakech, Morocco

²Diabetology and Metabolic Diseases, Caddi Ayyad University Faculty of Medicine Marrakech, Morocco

*Corresponding author: M. Soussou
DOI: 10.36347/sjmcr.2019.v07i03.001

| Received: 22.02.2019 | Accepted: 05.03.2019 | Published: 30.03.2019

Abstract

Case Report

Subependymal giant cell astrocytoma (SEGA) is a benign midline tumor of the central nervous system. It arises from the wall of the lateral ventricle and the foramen of Monro. It occurs in 10 to 15% of individuals with tuberous sclerosis complex (TSC), often before the second decade of age. These tumors have a slow evolutionary potential but may show aggressive local invasion with peri-lesional oedema and visual disturbances. The histological appearance is typical; It is a well-defined tumor, that morphologically associates astrocytic and ganglioneuronal elements. The double differentiation is confirmed by immunohistochemistry that shows cell expression of glial fibrillary acidic protein (GFAP) and vimentin. We report the case of a 9-year-old patient with isolated SEGA mimicking a sellar and suprasellar tumor with pituitary endocrine repercussions.

Keywords: Astrocytoma, subependymal, giant cell, brain tumor, children.

Copyright © 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

SEGA is a benign tumor that originates from the wall of lateral ventricle or in the foramen of Monro [1]. It occurs in 10-15% of patients with TSC [2]. Even if it has a low proliferative potential, it is responsible for intracranial hypertension and exposes to the risk of hydrocephalus. These tumors have an astrocytic and neuroglial double differentiation [3, 4]. SEGA is one of the major diagnosis criteria and is the main cause of morbi-mortality in TSC [5]. The histological appearance is typical. The tumor presents giant cell groups whose morphology associated astrocytic and glial elements. SEGA removal is not always complete. New therapies are emerging for this indication [6]. Gamma knife radiosurgery use is limited due to the lack of proven efficiency. We report the case of a 9 year old with SEGA extension in the sellar region, and in whom imaging has led to confusion with a sellar and suprasellar tumor.

CASE REPORT

A 9-years-old boy with no prior illness was referred to us for endocrine evaluation of a sellar tumor. He was complaining of a 3 years history of bilateral decline vision. Three months before, he presented an acute left hemibody paralysis. He has been seen by a neurologist who indicated a brain MRI that showed a

sellar, suprasellar and parasellar tumor, well-circumscribed lobulated of 42*43*47mm with cystic component slightly hypointense on T1 weighted images and hyperintense and heterogenous in T2 weighted images (Figures 1). This process arrives in contact to the optic chiasma and was responsible of a biventricular hydrocephaly. The process has a side-to-sellar extension respecting the ipsilateral cavernous sinus with an intimate contact with the interne carotid artery. No other cortical brain lesions or subcortical was identified.

On physical examination, blood pressure was at 100/80mmgh, postprandial capillary glycemia was at 145 mg/dl, he weighted 43kg with a height of 1,44m which made a BMI of 21kg/m². He had prepubertal external genitalia. Neurological examination showed a left pyramidal syndrome and a bitemporal hemianopsia. Laboratory studies showed: Testosterone: 0.025 ng / ml, FSH: 1.84 pg / ml, LH: 0.262 m IU / ml, Prolactin: 4.19 ng / ml, TSH: 1,598MUI / ml, T4: 17,39pmol / l, Cortisol: 152microg / l.

Surgery allowed partial resection. The postoperative follow were marked by the occurrence of a polyuropolydipsic syndrome put under vasopressin. Histologically, the tumor presented a glial fibrillary proliferation with large mono and multinucleate cells. The cytoplasm was abundant granular and eosinophilic.

The nucleus showed vesicular chromatin without abnormal mitosis. Immunohistochemistry was consistent with a diffuse cytoplasmic expression of GFAP, a moderate distribution of antiPS-100 and anti-neurofilament antibodies. The three months postoperative endocrine assessment was consistent with

a corticotrop, thyreotrop and vasopressin deficiency. Additional imaging exams were performed and showed no malformations supporting the diagnosis of TSC. The patient had complementary external radiotherapy treatment.



Fig-1: MRI showing an heterogeneous process in sellar, suprasellar and parasellar position

DISCUSSION

SEGA is classified as astrocytic tumors within tumors of the neuro-epithelial tissue according to the pathological classification of the central nervous system [7]. It occurs mostly in children, adolescents or young adults with TSC which is a genetic autosomal dominant phacomatosis [2]. In half of cases, it appears without a known family history because of neomutations [2]. The existence of SEGA is one of the major diagnosis criteria for the TSC [2]. The occurrence of SEGA outside a context of TSC requires the inactivation in the same tumor cell of 2 alleles [8]. This is an exceptional event and is probably the mechanism in our case. SEGA usually occurs on a subependymal nodule, often before the second decade of age [9]. Subependymal nodules have a very slow evolutionary potential but may present local invasion and peri-lesional edema [9]. The visual impact on the optic chiasma is described well before the transformation to a SEGA [9]. Histopathologically, it associates astrocytic and ganglioneural elements [8]. The giant cell astrocytoma simply corresponds to a subependymal nodule with a growth potential [9]. The predictive criteria of tumor progression are a diameter greater than 12mm, incomplete calcifications and the contrast enhancement [9].

On imaging, computed tomography is effective in detecting calcified ependymal nodules whereas magnetic resonance imaging is more appropriate to specify the number and location of brain hamartomatous, which presents with a hyperintense signal [4]. There is no imaging that is performed in differentiating a SEGA and a subependymal nodule [4]. MRI shows a subependymal image near the interventricular foramen that is usually causing hydrocephalus [4]. Typically, it is hypointense in T1

images and heterogeneous with a hyperintense center and hypointense periphery on T2 [4]. In our patient, the confusion with a sellar tumor particularly a craniopharyngioma was because of the fairly low position of the tumor and its progression toward the sella. This is to our knowledge, the first SEGA that presents a sellar extension.

The treatment of SEGA includes surgery, medical complementary treatment based on everolimus and gamma knife radiosurgery [6]. Our patient did not benefit from the new targeted therapies because it is not available in our context. Particular issues are taken into consideration in the follow up especially hypothalamic disturbances and the impact on growth, puberty and food intake regulation.

CONCLUSION

SEGA is rare tumor of the central nervous system. The diagnosis is made according to multiple clinical, radiological and histopathological features especially in a context of tuberous sclerosis. Clinicians should think about that diagnosis even in the absence of context of tuberous sclerosis and with an unusual extension to the skull base.

REFERENCES

1. Jung TY, Kim YH, Jung S, Baek HJ, Lee KH. The clinical characteristics of subependymal giant cell astrocytoma: five cases. *Brain tumor research and treatment*. 2015 Apr 1;3(1):44-7.
2. Belfquih H, Akhaddar A, Gazzaz M, Boucetta M. Les manifestations neuro-encéphaliques de la sclérose tubéreuse de Bourneville. *La Revue de médecine interne*. 2012 Aug 1;33(8):433-8.

3. Maurage CA, Sevestre H. Anatomie pathologique des tumeurs des ventricules latéraux. *Neurochirurgie*. 2011 Sep 1;57(4-6):193-8.
4. Delmaire C, Gauvrit JY, Hajj E, Ares GS, Ayachi M, Reyns N, Dubois F, Pruvo JP. Tumeurs cranio-encéphaliques de la ligne médiane. *Journal de radiologie*. 2006 Jun 1;87(6):764-78.
5. Northrup H, Krueger DA, Roberds S, Smith K, Sampson J, Korf B, Kwiatkowski DJ, Mowat D, Nellist M, Povey S, de Vries P. Tuberous sclerosis complex diagnostic criteria update: recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference. *Pediatric neurology*. 2013 Oct 1;49(4):243-54.
6. Józwiak S, Nabbout R, Curatolo P. Management of subependymal giant cell astrocytoma (SEGA) associated with tuberous sclerosis complex (TSC): clinical recommendations. *European journal of paediatric neurology*. 2013 Jul 1;17(4):348-52.
7. Louis DN, Perry A, Reifenberger G, Von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P, Ellison DW. The 2016 World Health Organization classification of tumors of the central nervous system: a summary. *Acta neuropathologica*. 2016 Jun 1;131(6):803-20.
8. Henske EP1, Józwiak S2,3, Kingswood JC4. Tuberous sclerosis complex. *Nat Rev Dis Primers*. 2016;(26)2:1603-5
9. Chow CW, Klug GL, Lewis EA. Subependymal giant-cell astrocytoma in children: An unusual discrepancy between histological and clinical features. *Journal of neurosurgery*. 1988 Jun 1;68(6):880-3.