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ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии

საქართველოს სამედიცინო სიახლენი

GEORGIAN MEDICAL NEWS

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GMN: Georgian Medical News is peer-reviewed, published monthly journal committed to promoting the science and art of medicine and the betterment of public health, published by the GMN Editorial Board since 1994. GMN carries original scientific articles on medicine, biology and pharmacy, which are of experimental, theoretical and practical character; publishes original research, reviews, commentaries, editorials, essays, medical news, and correspondence in English and Russian.

GMN is indexed in MEDLINE, SCOPUS, PubMed and VINITI Russian Academy of Sciences. The full text content is available through EBSCO databases.

GMN: Медицинские новости Грузии - ежемесячный рецензируемый научный журнал, издаётся Редакционной коллегией с 1994 года на русском и английском языках в целях поддержки медицинской науки и улучшения здравоохранения. В журнале публикуются оригинальные научные статьи в области медицины, биологии и фармации, статьи обзорного характера, научные сообщения, новости медицины и здравоохранения. Журнал индексируется в MEDLINE, отражён в базе данных SCOPUS, PubMed и ВИНИТИ РАН. Полнотекстовые статьи журнала доступны через БД EBSCO.

GMN: Georgian Medical News – საქართველოს სამედიცინო სიახლენი – არის ყოველთვიური სამეცნიერო სამედიცინო რეცენზირებადი ჟურნალი, გამოიცემა 1994 წლიდან, წარმოადგენს სარედაქციო კოლეგიისა და აშშ-ის მეცნიერების, განათლების, ინდუსტრიის, ხელოვნებისა და ბუნებისმეტყველების საერთაშორისო აკადემიის ერთობლივ გამოცემას. GMN-ში რუსულ და ინგლისურ ენებზე ქვეყნდება ექსპერიმენტული, თეორიული და პრაქტიკული ხასიათის ორიგინალური სამეცნიერო სტატიები მედიცინის, ბიოლოგიისა და ფარმაციის სფეროში, მიმოხილვითი ხასიათის სტატიები.

ჟურნალი ინდექსირებულია MEDLINE-ის საერთაშორისო სისტემაში, ასახულია SCOPUS-ის, PubMed-ის და ВИНИТИ РАН-ის მონაცემთა ბაზებში. სტატიების სრული ტექსტი ხელმისაწვდომია EBSCO-ს მონაცემთა ბაზებიდან.

WEBSITE

www.geomednews.com

К СВЕДЕНИЮ АВТОРОВ!

При направлении статьи в редакцию необходимо соблюдать следующие правила:

1. Статья должна быть представлена в двух экземплярах, на русском или английском языках, напечатанная через **полтора интервала на одной стороне стандартного листа с шириной левого поля в три сантиметра**. Используемый компьютерный шрифт для текста на русском и английском языках - **Times New Roman** (**Кириллица**), для текста на грузинском языке следует использовать **AcadNusx.** Размер шрифта - **12**. К рукописи, напечатанной на компьютере, должен быть приложен CD со статьей.

2. Размер статьи должен быть не менее десяти и не более двадцати страниц машинописи, включая указатель литературы и резюме на английском, русском и грузинском языках.

3. В статье должны быть освещены актуальность данного материала, методы и результаты исследования и их обсуждение.

При представлении в печать научных экспериментальных работ авторы должны указывать вид и количество экспериментальных животных, применявшиеся методы обезболивания и усыпления (в ходе острых опытов).

4. К статье должны быть приложены краткое (на полстраницы) резюме на английском, русском и грузинском языках (включающее следующие разделы: цель исследования, материал и методы, результаты и заключение) и список ключевых слов (key words).

5. Таблицы необходимо представлять в печатной форме. Фотокопии не принимаются. **Все цифровые, итоговые и процентные данные в таблицах должны соответствовать таковым в тексте статьи**. Таблицы и графики должны быть озаглавлены.

6. Фотографии должны быть контрастными, фотокопии с рентгенограмм - в позитивном изображении. Рисунки, чертежи и диаграммы следует озаглавить, пронумеровать и вставить в соответствующее место текста **в tiff формате**.

В подписях к микрофотографиям следует указывать степень увеличения через окуляр или объектив и метод окраски или импрегнации срезов.

7. Фамилии отечественных авторов приводятся в оригинальной транскрипции.

8. При оформлении и направлении статей в журнал МНГ просим авторов соблюдать правила, изложенные в «Единых требованиях к рукописям, представляемым в биомедицинские журналы», принятых Международным комитетом редакторов медицинских журналов - http://www.spinesurgery.ru/files/publish.pdf и http://www.nlm.nih.gov/bsd/uniform_requirements.html
В конце каждой оригинальной статьи приводится библиографический список. В список литературы включаются все материалы, на которые имеются ссылки в тексте. Список составляется в алфавитном порядке и нумеруется. Литературный источник приводится на языке оригинала. В списке литературы сначала приводятся работы, написанные знаками грузинского алфавита, затем кириллицей и латиницей. Ссылки на цитируемые работы в тексте статьи даются в квадратных скобках в виде номера, соответствующего номеру данной работы в списке литературы. Большинство цитированных источников должны быть за последние 5-7 лет.

9. Для получения права на публикацию статья должна иметь от руководителя работы или учреждения визу и сопроводительное отношение, написанные или напечатанные на бланке и заверенные подписью и печатью.

10. В конце статьи должны быть подписи всех авторов, полностью приведены их фамилии, имена и отчества, указаны служебный и домашний номера телефонов и адреса или иные координаты. Количество авторов (соавторов) не должно превышать пяти человек.

11. Редакция оставляет за собой право сокращать и исправлять статьи. Корректура авторам не высылается, вся работа и сверка проводится по авторскому оригиналу.

12. Недопустимо направление в редакцию работ, представленных к печати в иных издательствах или опубликованных в других изданиях.

При нарушении указанных правил статьи не рассматриваются.

REQUIREMENTS

Please note, materials submitted to the Editorial Office Staff are supposed to meet the following requirements:

1. Articles must be provided with a double copy, in English or Russian languages and typed or compu-ter-printed on a single side of standard typing paper, with the left margin of **3** centimeters width, and **1.5** spacing between the lines, typeface - **Times New Roman** (**Cyrillic**), print size - **12** (referring to Georgian and Russian materials). With computer-printed texts please enclose a CD carrying the same file titled with Latin symbols.

2. Size of the article, including index and resume in English, Russian and Georgian languages must be at least 10 pages and not exceed the limit of 20 pages of typed or computer-printed text.

3. Submitted material must include a coverage of a topical subject, research methods, results, and review.

Authors of the scientific-research works must indicate the number of experimental biological species drawn in, list the employed methods of anesthetization and soporific means used during acute tests.

4. Articles must have a short (half page) abstract in English, Russian and Georgian (including the following sections: aim of study, material and methods, results and conclusions) and a list of key words.

5. Tables must be presented in an original typed or computer-printed form, instead of a photocopied version. **Numbers, totals, percentile data on the tables must coincide with those in the texts of the articles.** Tables and graphs must be headed.

6. Photographs are required to be contrasted and must be submitted with doubles. Please number each photograph with a pencil on its back, indicate author's name, title of the article (short version), and mark out its top and bottom parts. Drawings must be accurate, drafts and diagrams drawn in Indian ink (or black ink). Photocopies of the X-ray photographs must be presented in a positive image in **tiff format**.

Accurately numbered subtitles for each illustration must be listed on a separate sheet of paper. In the subtitles for the microphotographs please indicate the ocular and objective lens magnification power, method of coloring or impregnation of the microscopic sections (preparations).

7. Please indicate last names, first and middle initials of the native authors, present names and initials of the foreign authors in the transcription of the original language, enclose in parenthesis corresponding number under which the author is listed in the reference materials.

8. Please follow guidance offered to authors by The International Committee of Medical Journal Editors guidance in its Uniform Requirements for Manuscripts Submitted to Biomedical Journals publication available online at: http://www.nlm.nih.gov/bsd/uniform_requirements.html
http://www.icmje.org/urm_full.pdf
In GMN style for each work cited in the text, a bibliographic reference is given, and this is located at the end of the article under the title "References". All references cited in the text must be listed. The list of references should be arranged alphabetically and then numbered. References are numbered in the text [numbers in square brackets] and in the reference list and numbers are repeated throughout the text as needed. The bibliographic description is given in the language of publication (citations in Georgian script are followed by Cyrillic and Latin).

9. To obtain the rights of publication articles must be accompanied by a visa from the project instructor or the establishment, where the work has been performed, and a reference letter, both written or typed on a special signed form, certified by a stamp or a seal.

10. Articles must be signed by all of the authors at the end, and they must be provided with a list of full names, office and home phone numbers and addresses or other non-office locations where the authors could be reached. The number of the authors (co-authors) must not exceed the limit of 5 people.

11. Editorial Staff reserves the rights to cut down in size and correct the articles. Proof-sheets are not sent out to the authors. The entire editorial and collation work is performed according to the author's original text.

12. Sending in the works that have already been assigned to the press by other Editorial Staffs or have been printed by other publishers is not permissible.

Articles that Fail to Meet the Aforementioned Requirements are not Assigned to be Reviewed.

ავტორთა საყურადღებოდ!

რედაქციაში სტატიის წარმოდგენისას საჭიროა დავიცვათ შემდეგი წესები:

1. სტატია უნდა წარმოადგინოთ 2 ცალად, რუსულ ან ინგლისურ ენებზე,დაბეჭდილი სტანდარტული ფურცლის 1 გვერდზე, 3 სმ სიგანის მარცხენა ველისა და სტრიქონებს შორის 1,5 ინტერვალის დაცვით. გამოყენებული კომპიუტერული შრიფტი რუსულ და ინგლისურენოვან ტექსტებში - **Times New Roman (Кириллица)**, ხოლო ქართულენოვან ტექსტში საჭიროა გამოვიყენოთ **AcadNusx.** შრიფტის ზომა – 12. სტატიას თან უნდა ახლდეს CD სტატიით.

2. სტატიის მოცულობა არ უნდა შეადგენდეს 10 გვერდზე ნაკლებს და 20 გვერდზე მეტს ლიტერატურის სიის და რეზიუმეების (ინგლისურ, რუსულ და ქართულ ენებზე) ჩათვლით.

3. სტატიაში საჭიროა გაშუქდეს: საკითხის აქტუალობა; კვლევის მიზანი; საკვლევი მასალა და გამოყენებული მეთოდები; მიღებული შედეგები და მათი განსჯა. ექსპერიმენტული ხასიათის სტატიების წარმოდგენისას ავტორებმა უნდა მიუთითონ საექსპერიმენტო ცხოველების სახეობა და რაოდენობა; გაუტკივარებისა და დაძინების მეთოდები (მწვავე ცდების პირობებში).

4. სტატიას თან უნდა ახლდეს რეზიუმე ინგლისურ, რუსულ და ქართულ ენებზე არანაკლებ ნახევარი გვერდის მოცულობისა (სათაურის, ავტორების, დაწესებულების მითითებით და უნდა შეიცავდეს შემდეგ განყოფილებებს: მიზანი, მასალა და მეთოდები, შედეგები და დასკვნები; ტექსტუალური ნაწილი არ უნდა იყოს 15 სტრიქონზე ნაკლები) და საკვანძო სიტყვების ჩამონათვალი (key words).

5. ცხრილები საჭიროა წარმოადგინოთ ნაბეჭდი სახით. ყველა ციფრული, შემაჯამებელი და პროცენტული მონაცემები უნდა შეესაბამებოდეს ტექსტში მოყვანილს.

6. ფოტოსურათები უნდა იყოს კონტრასტული; სურათები, ნახაზები, დიაგრამები - დასათაურებული, დანომრილი და სათანადო ადგილას ჩასმული. რენტგენოგრამების ფოტოასლები წარმოადგინეთ პოზიტიური გამოსახულებით **tiff** ფორმატში. მიკროფოტოსურათების წარწერებში საჭიროა მიუთითოთ ოკულარის ან ობიექტივის საშუალებით გადიდების ხარისხი, ანათალების შეღებვის ან იმპრეგნაციის მეთოდი და აღნიშნოთ სურათის ზედა და ქვედა ნაწილები.

7. სამამულო ავტორების გვარები სტატიაში აღინიშნება ინიციალების თანდართვით, უცხოურისა – უცხოური ტრანსკრიპციით.

8. სტატიას თან უნდა ახლდეს ავტორის მიერ გამოყენებული სამამულო და უცხოური შრომების ბიბლიოგრაფიული სია (ბოლო 5-8 წლის სიღრმით). ანბანური წყობით წარმოდგენილ ბიბლიოგრაფიულ სიაში მიუთითეთ ჯერ სამამულო, შემდეგ უცხოელი ავტორები (გვარი, ინიციალები, სტატიის სათაური, ჟურნალის დასახელება, გამოცემის ადგილი, წელი, ჟურნალის №, პირველი და ბოლო გვერდები). მონოგრაფიის შემთხვევაში მიუთითეთ გამოცემის წელი, ადგილი და გვერდების საერთო რაოდენობა. ტექსტში კვადრატულ ფჩხილებში უნდა მიუთითოთ ავტორის შესაბამისი N ლიტერატურის სიის მიხედვით. მიზანშეწონილია, რომ ციტირებული წყაროების უმეტესი ნაწილი იყოს 5-6 წლის სიღრმის.

9. სტატიას თან უნდა ახლდეს: ა) დაწესებულების ან სამეცნიერო ხელმძღვანელის წარდგინება, დამოწმებული ხელმოწერითა და ბეჭდით; ბ) დარგის სპეციალისტის დამოწმებული რეცენზია, რომელშიც მითითებული იქნება საკითხის აქტუალობა, მასალის საკმაობა, მეთოდის სანდოობა, შედეგების სამეცნიერო-პრაქტიკული მნიშვნელობა.

10. სტატიის ბოლოს საჭიროა ყველა ავტორის ხელმოწერა, რომელთა რაოდენობა არ უნდა აღემატებოდეს 5-ს.

11. რედაქცია იტოვებს უფლებას შეასწოროს სტატია. ტექსტზე მუშაობა და შეჯერება ხდება საავტორო ორიგინალის მიხედვით.

12. დაუშვებელია რედაქციაში ისეთი სტატიის წარდგენა, რომელიც დასაბეჭდად წარდგენილი იყო სხვა რედაქციაში ან გამოქვეყნებული იყო სხვა გამოცემებში.

აღნიშნული წესების დარღვევის შემთხვევაში სტატიები არ განიხილება.

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EPILEPTIC SEIZURES REVEALING STURGE WEBER'S DISEASE IN A TROPICAL ENVIRONMENT: STUDY OF EIGHT CASES

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Abstract.

Introduction: In sub-Saharan Africa (SSA), the etiological factors of epilepsy are multiple and phacomatoses, in particular Sturge weber's disease, are rarely reported due to under-medicalization and insufficient multidisciplinary care.

Patient and Methods: We carried out a retrospective study of 216 patients hospitalized for recurrent epileptic seizures between 2015 and 2022 in the neurology and pediatrics department of the University Hospital Center of Conakry, among whom eight (8) patients were identified for Sturge Weber's disease in order to reassess this pathology from a clinical and paraclinical point of view in a tropical environment.

Results: Sturge Weber's disease was retained in eight (8) on the presence of symptomatic partial epileptic seizures (age 6 months to 14 years) with frequency of status epilepticus, homonymous lateral hemiparesis linked to occipital involvement, piriform calcifications on imaging and ocular disorders.

The delay in consultation and medical care revealed severe mental deterioration in our patients.

Conclusion: this study shows a stereotyped clinical picture in a context of aggravation of signs related to a delay in multidisciplinary management.

These results are important for the diagnostic, therapeutic and prognostic discussion.

Key words. Sturge weber disease, Conakry-Guinea.

Introduction.

In Guinea, the existence of a study project on epilepsy in collaboration with Harvard Medical school [1-5] focused on the clinical and paraclinical evaluation and the management of epileptics allows the identification of the various clinical and therapeutic aspects of epilepsy. Thus, provider of recurrent epileptic seizures with more or less severe mental deterioration, neuro-ectodermoses, in particular Sturge Weber Krabe disease and Bonneville tuberous sclerosis, are classified among the most neglected and misunderstood orphan diseases in the tropics.

The existence of neurological manifestations in the form of partial motor epilepsies involving the hemi body opposite to the facial angioma, sometimes secondarily generalized, associated with progressive mental deterioration, expression of Sturge Weber's disease, is now a well-established fact since the initial publications. from [6-8] and recent ones [9-13] and by many revisited series [14,15].

The recent introduction of cerebral computed tomography and magnetic resonance imaging, the development of electroencephalography techniques have enabled a better diagnostic approach to phacomatosis, in particular Sturge Weber's disease, which occurs in populations of all ethnic and geographical origins with an incidence of 1 in 10,000 to 20,000 births in the general population [16]. The interest of this work lies in the fact that these observations clearly illustrate Sturge Weber's disease, a little-known condition in Sub-Saharan Africa, as evidenced by the scarcity of publications in the literature and the diagnostic difficulty it entails with the conditions, neurodegenerative disorders with epileptic connotations, in particular Klippel Trenaunay syndrome, Parkes Weber syndrome.

Material and Methods.

The eight (8) patients (5 boys and 3 girls) were observed between January 1, 2015, and December 31, 2022, in the neurology and pediatrics departments, the only centers in the country equipped to treat patients suffering from neurological pathologies such as epilepsies, infirmities of cerebral origin, neurodegenerative diseases of the nervous system, demyelinating diseases, neuro-infections and cerebrovascular accidents.

All patients underwent a systematic neurological examination by a neurologist and a specialized psychiatrist: history of the disease with precise description of the seizures, family history in the history with research of the relatives or those suffering from illnesses associated with the comitiality, medical history of the patient, gestational and neonatal history, and neonatal central nervous system trauma.

The inclusion criteria were based on those dictated by Bachur et al. [17], Bodensteiner et al. [18] on the basis of focal seizures and neurological deficit: EEG (EEG, NIPPON Neurofax, Japan) with slow wave spikes in one hemisphere with unilateral onset of epilepsy which may be secondarily generalized MRI/CT (CT Scanners 16-slice-spinal Toshiba United States) pear-shaped calcifications in the parieto-occipital cortex, Angioinsent calcification in T1, Hyper signal in T2 flair MRI (MRI scanners ,1.5T SIGNA- UNITED STATES) Progressive cortical parieto-occipital hemiatrophy.

All the patients benefited from a paraclinical examination and according to the semiological Oto-Rhino-Laryngological presentation (Laryngoscope FNL-10 RP3, PENTAX France),

exploration with an audiogram (Audiogram AD629 b, French interacoustic) and ophthalmological (ophthalmoscope IECLR6, 3000 HEINE mini, Germany) examination with fundus and visual acuity.

All patients underwent a series of complementary examinations (Auto Hematology Analyzer 5, BK-6310 Biotase Germany), including CBC, ESR, fasting blood glucose, 24-hour proteinuria, ionogram, serum calcium, serum phosphorus, transaminase SGPT and SGOT, CPK, CRP. Analysis of cerebrospinal fluid by lumbar puncture performed in all patients allowed cytological and biochemical evaluation: proteinorachia, glycorachia and chlorurachia (Compact immunoanalysis machine Mimi VIDAS, BIOMERIEUX, France).

Viral studies were carried out using PCR tests: HSV1/2, VZB, EBV, CMV and enteroviruses to exclude viral meningoencephalitis frequent in tropical environments.

Neuroradiological examinations, in particular MRI, were carried out in six patients with sequences in T1 and T2 in the axial and frontal planes and cerebral computed tomography in two patients.

During hospitalization, two electroencephalography tracings were performed in all patients on admission and on discharge. The electroencephalogram was performed in all patients and the graphs were classified into 3 types:

Type I:

- EEG with dominance of topography alpha rhythms parieto-occipital whose amplitude is greater than 40 μvolts without pathological rhythm.
- EEG with predominance of alpha rhythms of small amplitudes up to 25 μ volts with a tendency to flattening.

Type II:

- EEG without dominance proper with the existence of irregular alpha rhythms without the presence of pathological waves.
- EEG with theta rhythms of 4 to 6 cycles/second especially of low amplitude temporo-parietal topography of 30 to 40 μ volts, isolated or sometimes grouped in the form of paroxysmal bursts.

Type III:

- EEG with theta and delta rhythms showing abnormal figures.
- EEG with slowing of alpha rhythms associated with bursts of theta and delta waves.

Figure 1. *Left proptosis due to dilation of the ophthalmic vein and peri palpebral skin angioma.*

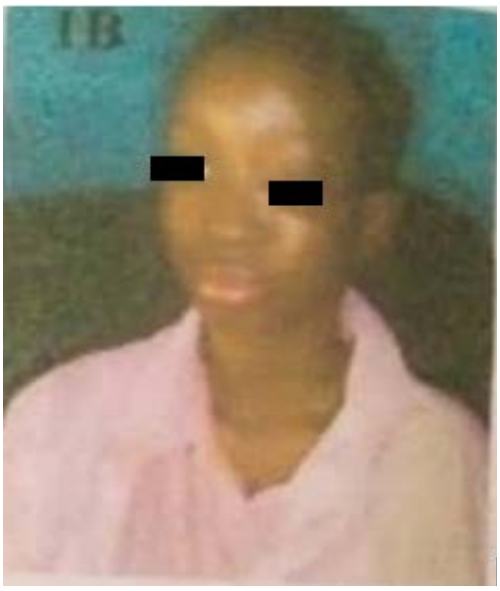

Figure 2. *Facial angioma associated with left glaucoma.*

Results.

The study population includes 8 patients (5 boys and 3 girls aged 2 to 11 years) (table I). The duration of the first visit to the hospital varied from 2 to 4 years and all the patients came from rural areas and had all consulted in traditional medicine and the failure of this treatment and the appearance of status epilepticus signals in 5 patients were the reason first consultations in neurology because of worsening cognitive functions. In 3 cases, a notion of epileptic affection was noted in the parents. The analysis of the results of this study focused on the clinical (Table I), biological, neuroradiological and electrophysiological (Table II) data.

This table highlights under the seniority heading that the onset of seizures does not correspond at all to the first consultations in neurology due essentially to socio-cultural conceptions of epilepsy in Africa. The analysis of the complaints noted the presence in his patients of chronic pseudo-migraine headaches with sometimes cephalalgic paroxysms in 4 patients Table I (patient: 3,4,7,8).

Image Data.

Imaging ccerebral examination showed abnormalities homolateral to the facial angioma with the presence of calcifications appearing hypo intense in T1 and hyper intense in T2. Figures (4,5,6,7,8,9).

Electro-Encephalographic Data.

The analysis of the bioelectric activity of the brain in this patient highlighted encephalographic tracings of type II in two cases and 6 in three cases considered almost all epileptogenic.

In 2 cases aedelirium observed, stage fright they showed a Dissymmetric hypersynchronous load on all derivates.

Discussion.

This study reports eight cases of Sturge weber's disease revealed by epileptic seizures, diagnosed at the University Hospital of Conakry.

These are all presumed certain cases whose diagnosis has been made in the neurology and neuropediatric departments of the University Hospital of Conakry.

Table 1. *Clinical characteristics of the eight (8) patients observed.*

No.	Gender/ Age	AF/L	Type of seizures	Seniority	Exam Ophthalmologist	Det. Is lying	Background Neurological Manifestations
1	M/2 years	AFD	Left partial motor seizures	4 months	-	Incessant crying	-
2	M/6yrs	AFG	Tonic seizures	Four years	Retinal detachment	Mental deterioration	Right hemiparesis
3	F/12 years old	AFG	Secondarily generalized Jacksonian Bravais seizures	8 years	Enlargement eye-ball	epileptic dementia stigma	Right lateral homonymous hemianopsia Hemiparesis
4	M/7yrs	AFD	Generalized tonic-clonic seizures	3 years	Glaucoma	Mental retardation	Hemiparesis Pseudo migraine headache
5	F/6yrs	AFD	Left partial motor seizures	4 years	-	Recurrent psychomotor agitation	Left hemiparesis
6	M/5yrs	AFG	Right hemigeneralized seizures - frequent MAL state	3 years	-	Incessant fears	
7	F/8 years	AFD	Right partial motor seizures STATE OF MAL	5 years	Glaucoma	Severe mental retardation	Left hemiparesis and Right pseudo migraine headache
8	M/11 years old	AFD	Secondarily generalized left partial seizures	7 years	Choroidal angioma	Mental retardation	Lateral homonymous hemianopsia Right pseudo migraine headache

M= male, F= female, AFD/AFG= right/left facial angioma. Ex oph: ophthalmology examination, Ment Det : Mental Deterioration.

Table 2. *Paraclinical characteristics in eight patients suffering from Sturge Weber.*

NOT	LCR		IMAGING			EEG/TYPE		
	Cyt	**Pro**	**CT Scanner**	**MRI**	**Skull x-ray**	**I**	**II**	**III**
1	2	0.10	Giriform calcification Oculomotor palsy Atrophy	-	-	-	III	III
2	3	0.15	-	Right occipito-parietal hyperintensities	-	-	II	-
3	3	0.12	-	Hypersignals Bilateral occipito-parieto---right temporal	Rail vermicular calcification	-	III	III
4	1	0.11	-	Right occipital hyperintensities	-	-	III	III
5	2	0.10	-	Occipito-parietal hyperintensities associated with atrophy	-	-	III	III
6	2	0.60	-	Parieto-occipital hyperintensity	-	-	III	III
7	3	0.15		-	Rail vermicular calcification	-	III	III
8	2	0.15	-	Parieto-occipital hyperintensity	-	-	II	-

Cys: Cytology; Pro: protein; EEG: electroencephalogram, LCR: cerebrospinal fluid

The existence of epileptic manifestations in the form of focal motor seizures of the hemibody opposite the facial angioma, generalized tonic-clonic seizures alternating with facial seizures is a well-established fact since the old publications of [6,18] and recent [20,21]. This frequency varies from 75 to 90% of cases and it appears most often between 3 and 6 months and rarely after 14 years according to most authors [17,19].

In our series, the first therapeutic link goes through traditional medicine because of the socio-cultural conceptions that surround epileptic seizures resorting to "talisma" and other healers using Koranic writings in their practices [1-4] explaining the delays of consultation.

In general, the clinical pictures of epilepsy observed in this study do not fundamentally differ from those described in the literature [19-24] with two severe semiological entities:

Figure 3. *Unilateral gingival hyperplasia.*

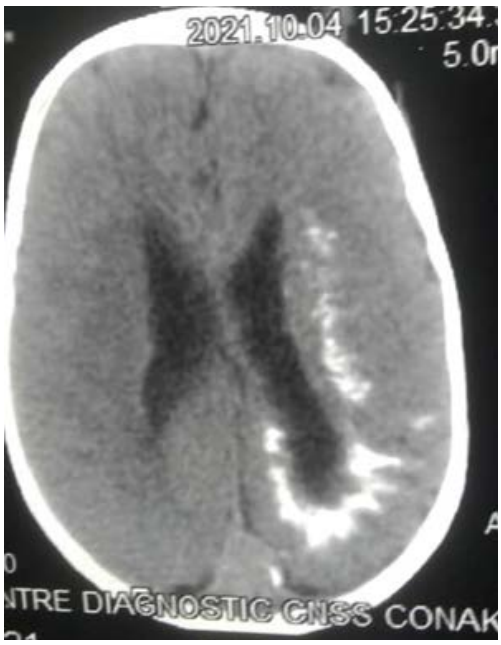

Figure 4. *Cerebral CT without injection which highlights spontaneous subcortical hyper density around the left lateral ventricle.*

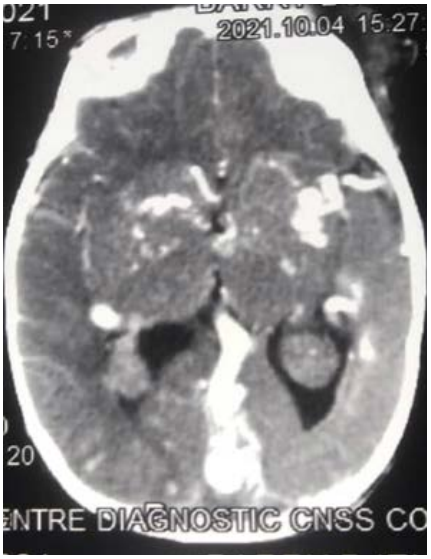

Figure 5. *CTcerebralwith injection which highlights hyper density of the choroid plexus and thickening of the tentorium of the cerebellum with bilateral calcification of the basal ganglia.*

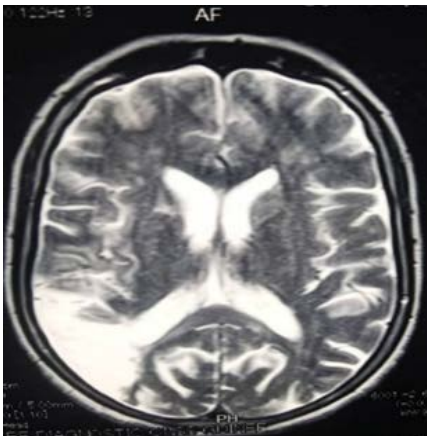

Figure 6. *MRI cerebral sequence T2 axial slice which shows a right temporo-occipital hypersignal with widening of the cortical furrows and arachnoid spaces.*

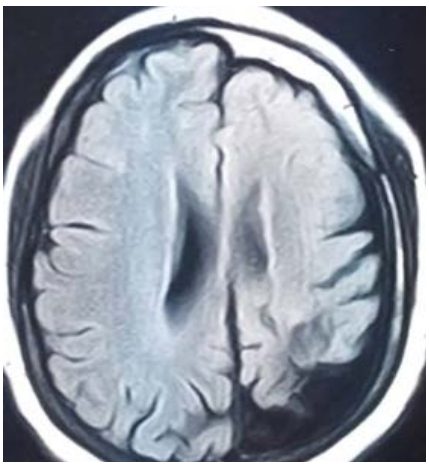

Figure 7. *Cerebral MRI sequence T2 FLAIR: Left hemi-cerebral atrophy with cortical calcifications and compensatory hypertrophy of the frontal diploe.*

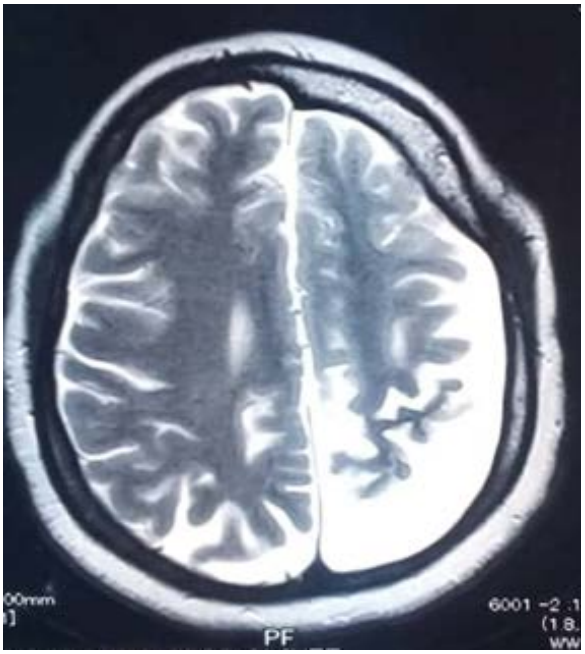

Figure 8A. *Cerebral MRI sequence T2: Hemileft cerebral atrophy with compatible cortical calcifications at the Sturge Weber's disease.*

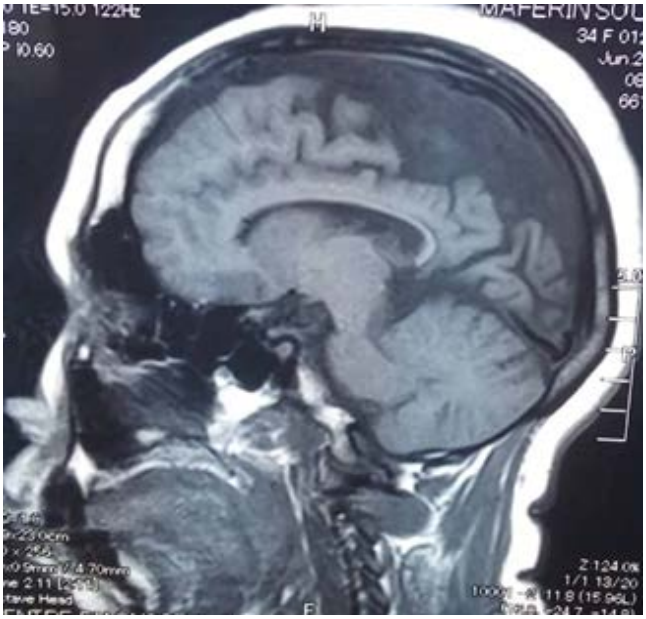

Figure 8B. *Brain MRI: sagittal section in T1Left hemi-cerebral atrophy with cortical calcifications and compensatory hypertrophy of the diploe in compatible frontal at the Sturge Weber's disease.*

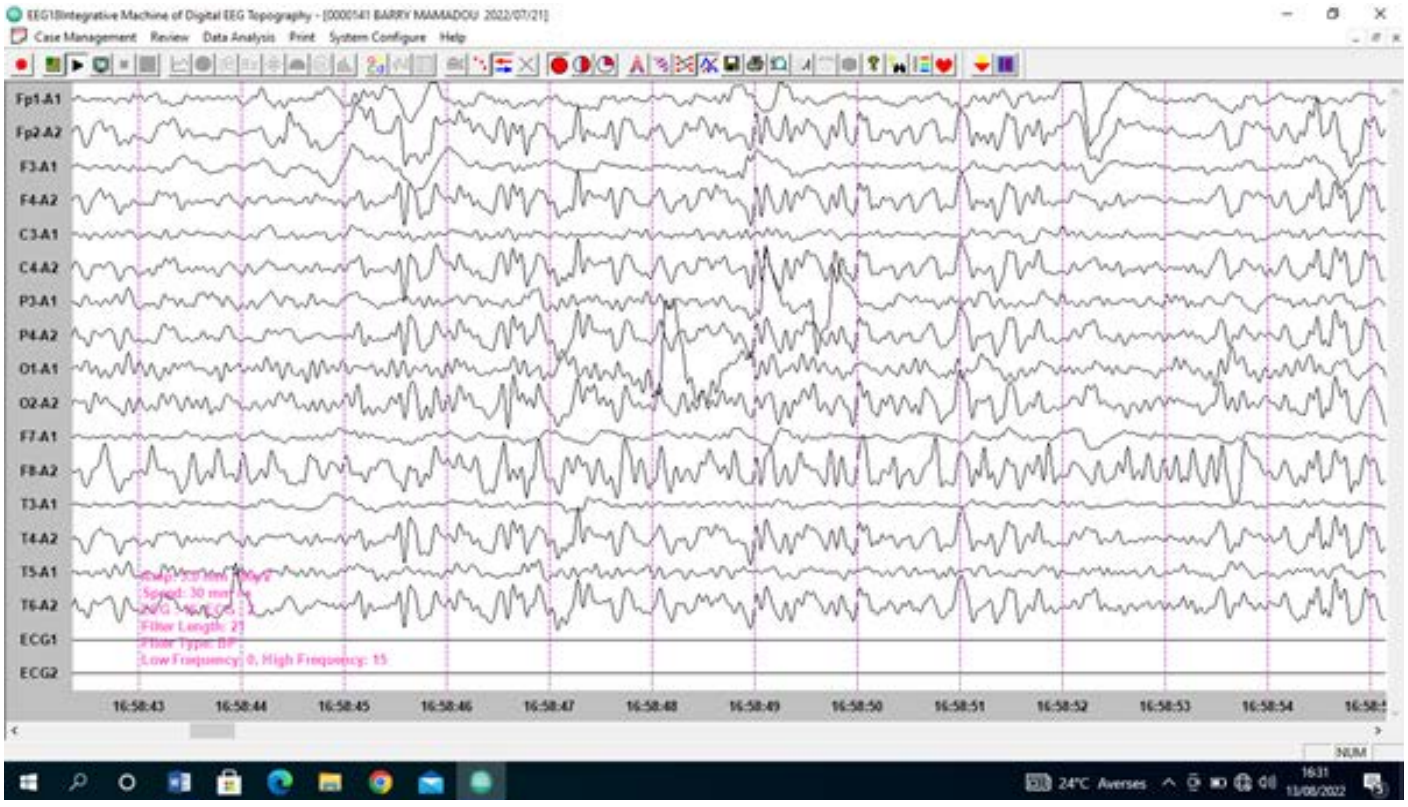

Figure 9A. *The EEG shows rare paroxysmal abnormalities in the type of slow spikes and numerous sequences of diffuse delta theta slow waves testifying to cerebral suffering compatible with type II.*

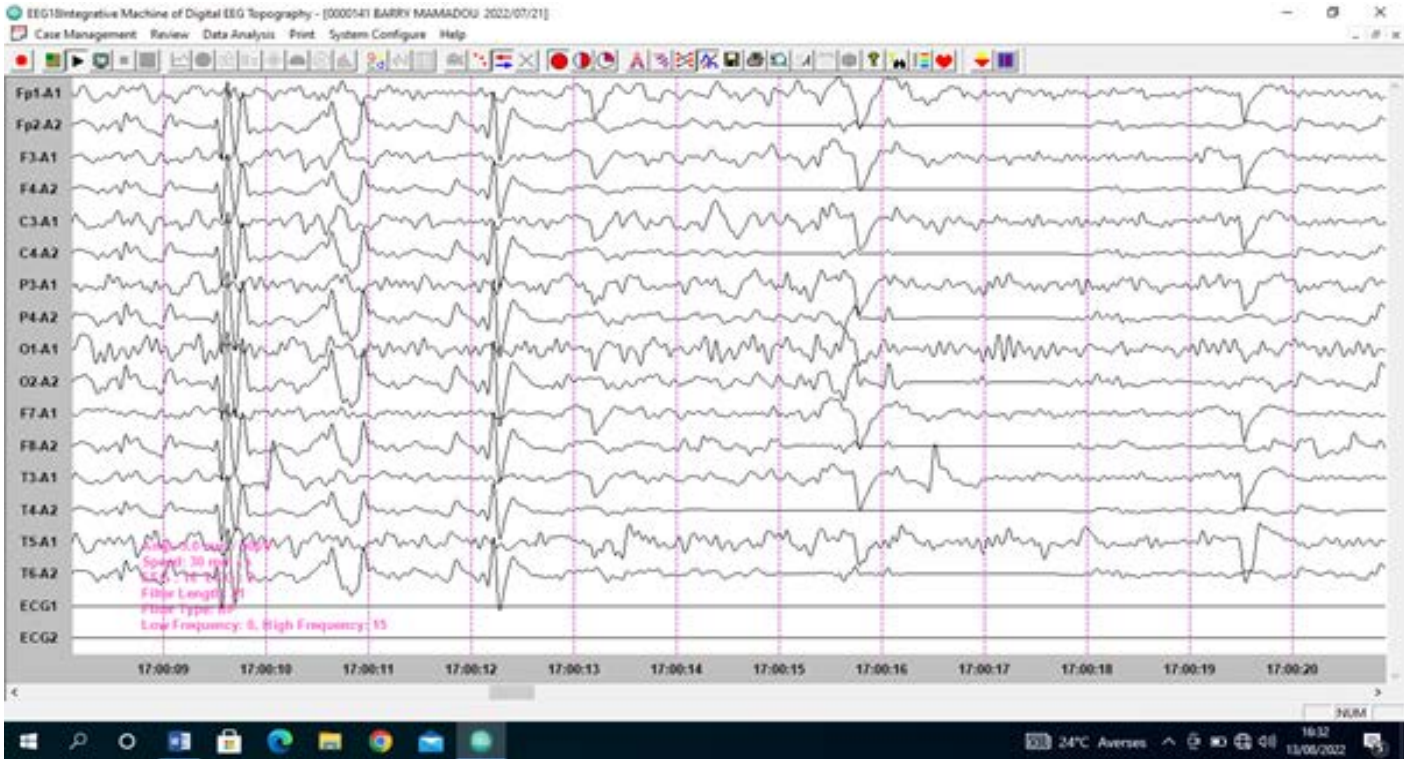

Figure 9B. *EEG shows generalized short bursts of slow spikes separated by low amplitude tracing intervals as well as slow delta theta waves suggesting diffuse cerebral pain compatible with type II.*

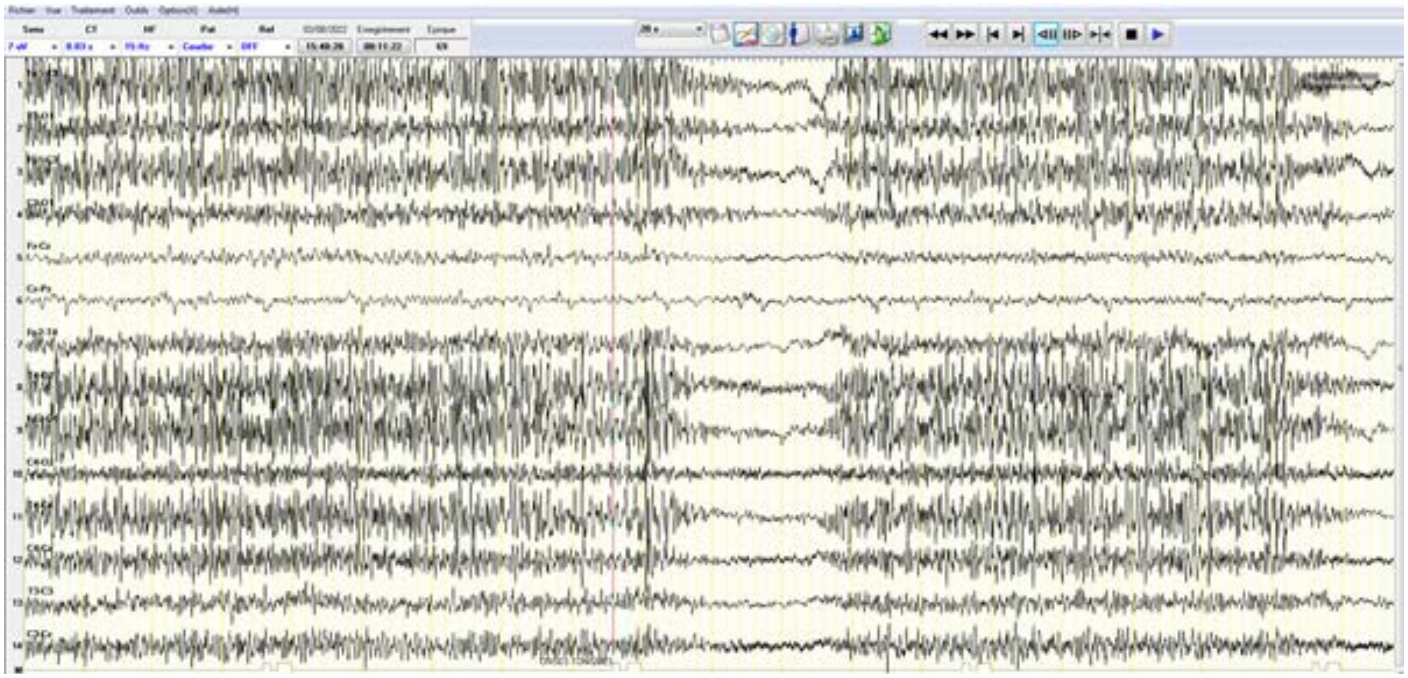

Figure 10. *Type III wake-up EEG trace performed, showing a slow monophasic, rhythmic background interspersed with bursts of generalized spike-waves compatible at an EMI.*

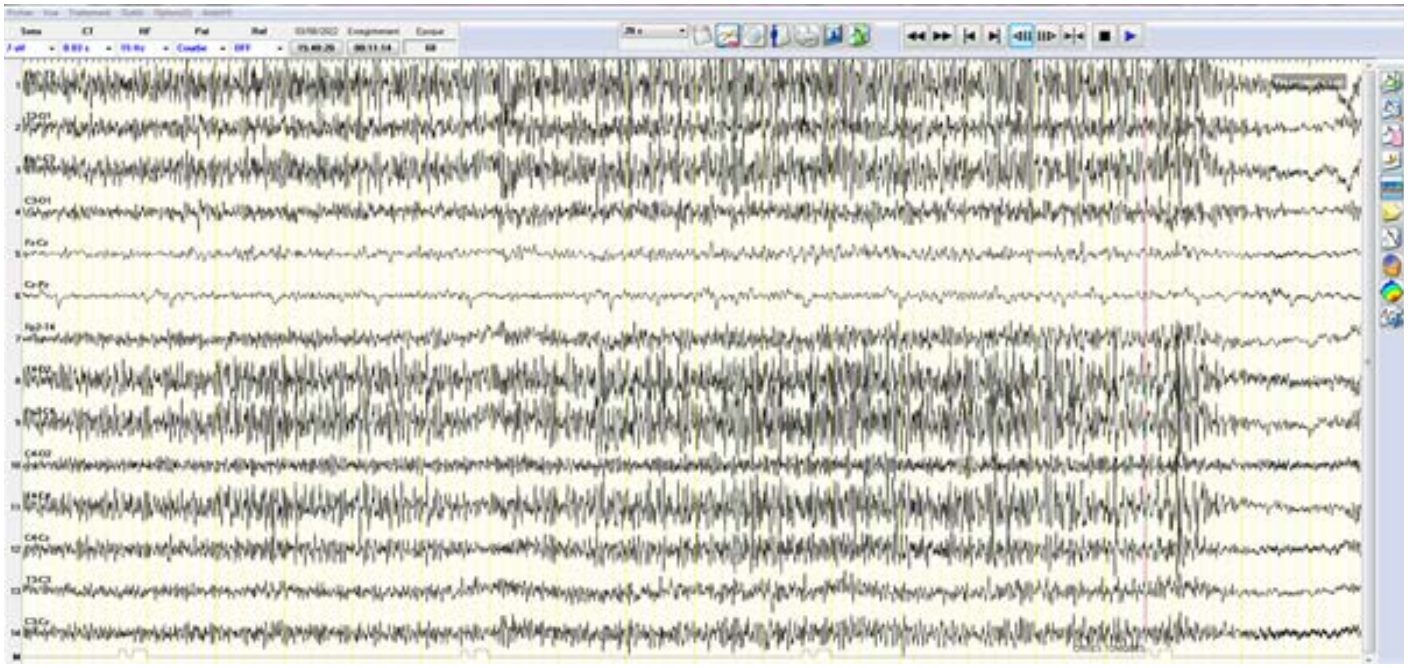

Figure 11. *Type III Awake EEG of an 8-year-old child, showing prolonged discharge of bi-frontal spikes-waves, temporally diffusing, rhythmic compatible at an EMI.*

high frequency of epilepticus and progressive severe mental deterioration due to insufficient multidisciplinary management.

This state of evil often presents itself in the form of confusional syndrome of brutal installation with alternation of phases where the patient seems dazed, haggard, automatisms with motor signs, clones of the chin of the labial commissures and extremities with tonic deviation of the head and eyes, this type of confusional status was observed in two (2) patients when they stopped treatment based on sodium valproate, phenobarbital and carbamazepine, the only antiepileptic available in Guinea.

In all cases, these paroxysmal states in this condition are associated with other abnormalities making the diagnosis of Sturge weber's disease easy: "lie de via" facial angioma and an ipsilateral angioma of fluctuant deficits such as hemiplegia, presence aphasia or hemianopsia, during which clinical migraine-like headaches were noted in half of our patients (patients 3.4, 7.8). On the pathophysiological level, there are no anatomoclic observations in the literature that allow discussion of the pathogenetic hypotheses on precise anatomical bases of these pseudo migraine headaches with the type of paroxysmal hemicrania noted in 30% in the literature and 50% in our series. Two hypotheses are usually proposed to explain the appearance of these headaches. They would be due to a neuronal hypersensitivity, expression of leptomeningeal alterations by chronic irritation and on the other hand to a bad regulation of the localized blood flow or at a distance from the angioma. Other neurological manifestations could also be the consequence of repeated epileptic seizures or arterial or venous occlusions [17,13].

Ocular manifestations expressed by glaucoma sometimes requiring surgical treatment have been reported by the authors [25,13] and oral manifestations have been listed in other series [26]. In the literature [25,13] glaucoma with a prevalence rate of 30 to 70% is linked to an anterior chamber malformation, high episcleral venous pressure (EVP) and changes in ocular hemodynamics [25,13].

Neuroradiological data, in particular computed tomography, reveals cortical calcifications early on [27,28] and on MRI the calcifications appear hypointense on T1 and hyperintense, particularly on echogradient T2 [29,30]. Cerebral bioelectric activity almost shows disturbances with abnormalities focused on the side of the unilateral or bilateral facial angioma and can appear spontaneously or during hyperpnea (Figures 1 and 2). In general, the differential diagnosis of Sturge Weber's disease is posed with Klippel Trenaunay syndrome [31], Parkes-Weber syndrome, Wyburn-Mason syndrome [32], Nova syndrome [33]. , hemangioma syndrome [34] and Rendu-Osler Weber syndrome [35]. and presence of visceral involvement. Klippel Trenaunay syndrome associates a complex congenital malformation: angioma in the limbs, venous malformations, and soft tissue hypertrophy. Diagnostic difficulties arise when Sturge weber disease is associated with klippel Trenauny syndrome as reported in the work of Kentab AY et al. [36] and J Second et al.[37].

Conclusion.

This study of 8 cases of Sturge Weber's disease revealed by polymorphic epileptic seizures shows the presence and persistence of this condition in a tropical environment. Stereotyped clinical semiology requires its inclusion in the multiple etiologies of epilepsies in Africa and raises the issue of multidisciplinary management of this condition.

Conflict of interest.

Authors have no conflicts of interest to declare.

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