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Calcification of the Central Gray Nuclei Revealing a Pseudo-Hypoparathyroidism Complicated by Rickets with Pros of a One CAS

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Abstract: ***Pseudo-hypoparathyroidism, of which there are several types, is. Relates to a disorder of parathyroid function, consisting of peripheral resistance of the target organs, kidneys and bones, to parathyroid hormone.***

Keyword: Calcification of the central gray nuclei, pseudo-hypoparathyroidism, rickets

1. Observation

We report the case of a 12-year-old adolescent with no particular history, admitted to the pediatric neurology department at Albert Royer CHUN de Fann for a motor deficit of the four limbs associated with a language disorder and convulsive seizures

We have discussed metabolic encephalopathy:

A pseudo-hypo-parathyroid type endocrinopathy
Intact parathyroid hormone: **160.1pg** / ml [**1.59 - 7.21**] **ab**normal
Vit D (25 - 0H) <8,1mg / ml Deficiency **<10mg / ml ab**normal

Treatment initiated after endocrinology advice
- Colecalciferol or vitamin D 3 100,000IU: 2 ampoules taken once per month
- Carbamazepine suspension 1 case x 2 / day
-Physiotherapy

2. Introduction

Pseudo-hypoparathyroidism, of which there are several types, is. relates to a disorder of parathyroid function, consisting of peripheral resistance of the target organs, kidneys and bones, to parathyroid hormone. As for rickets, they are linked to a metabolic disorder of vitamin D either secondary to a deficiency of intake (deficiency form), or to a disorder of hydroxylation of vitamin D in (pseudo-deficiency form), or even resistance. Peripheral terminal metabolite (resistance to 1-25 OHD), some forms may be linked to renal phosphate leakage (vitamin-resistant rickets, particularly familial X-linked). These are often familial affections for pseudo-hypoparathyroidism or vitamin-resistant rickets, or of nutritional origin for deficiency rickets. (**Rojbi et al**)

Observation
We report the case of a 12-year-old adolescent with no particular history, admitted to the pediatric neurology department at Albert Royer CHUN de Fann for a motor deficit of the four limbs associated with a language disorder and convulsive seizures.

The symptoms began on March 16 with the occurrence of repeated fall associated with difficulty in walking for which they consulted a hospital center where a medication prescribed with amendment for a week.

Follow-up of recurrence of falls associated with secondarily generalized focal convulsive seizures and regression of psychomotor acquisitions. After having gone through various primary and secondary medical structures as well as traditional therapists for about 15 months without amendment, hence the reason for consultation in pediatric neurology to Albert Royer for support.

At the exam, the patient was in good general condition, mucous membrane of normal color, anectic, flexible calf, no objectified limb edema.

Vital parameters were normal, showing no disturbance of consciousness, motor aphasia

In addition, he presented:
- A motor deficit in the four spastic limbs with a segmental muscle strength of 4/5 in the 4 limbs according to the [**Medicalresearch consul**] MRC scale
- 4-limb hypertonia with axial hypotonia
- Vivid osteotendinous reflexes to four limbs
- Amyotrophies with four members
- Cutaneo-plantar reflexes in bilateral extension
- Flexible neck, no impairment of objectified oculomotricity
- Other examinations without special features:

In front of this table associating
- A bilateral pyramidal syndrome of the cortical type
- Secondarily generalized focal convulsive seizures
- A motor aphasia type Broca

We discussed:
- Encephalopathy
- Or an intracranial expansive process

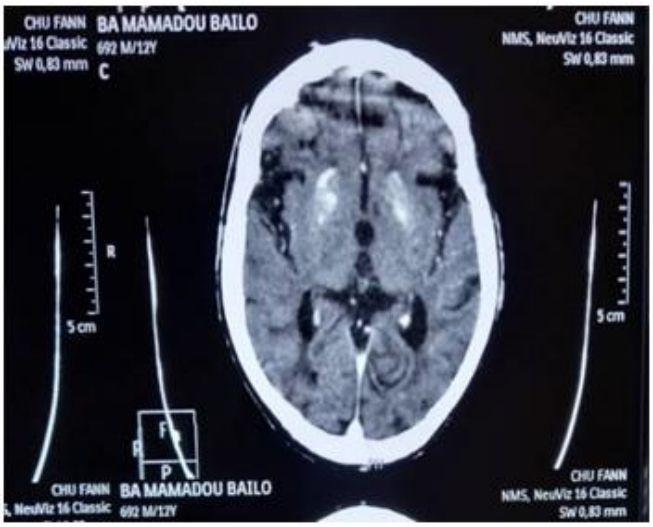

Figure 1: Brain CT axial slice, parenchymal window above tensor level

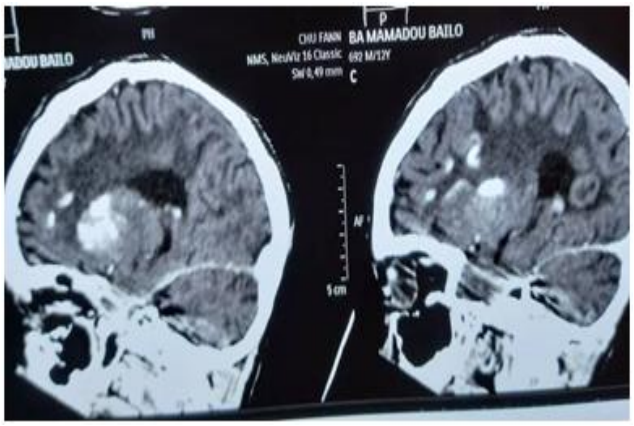

Figure 2: Brain CT, axial slice, upper and sub-tentorial stage and a parenchymal window

These two scanners objectifying:

- Objective Bilateral **macro calcification** at the level of **the lenticular nuclei**, **caudate** and at the level of the gray and white substance junction, at the frontal level
- **Peri-ventricular leukopathy**

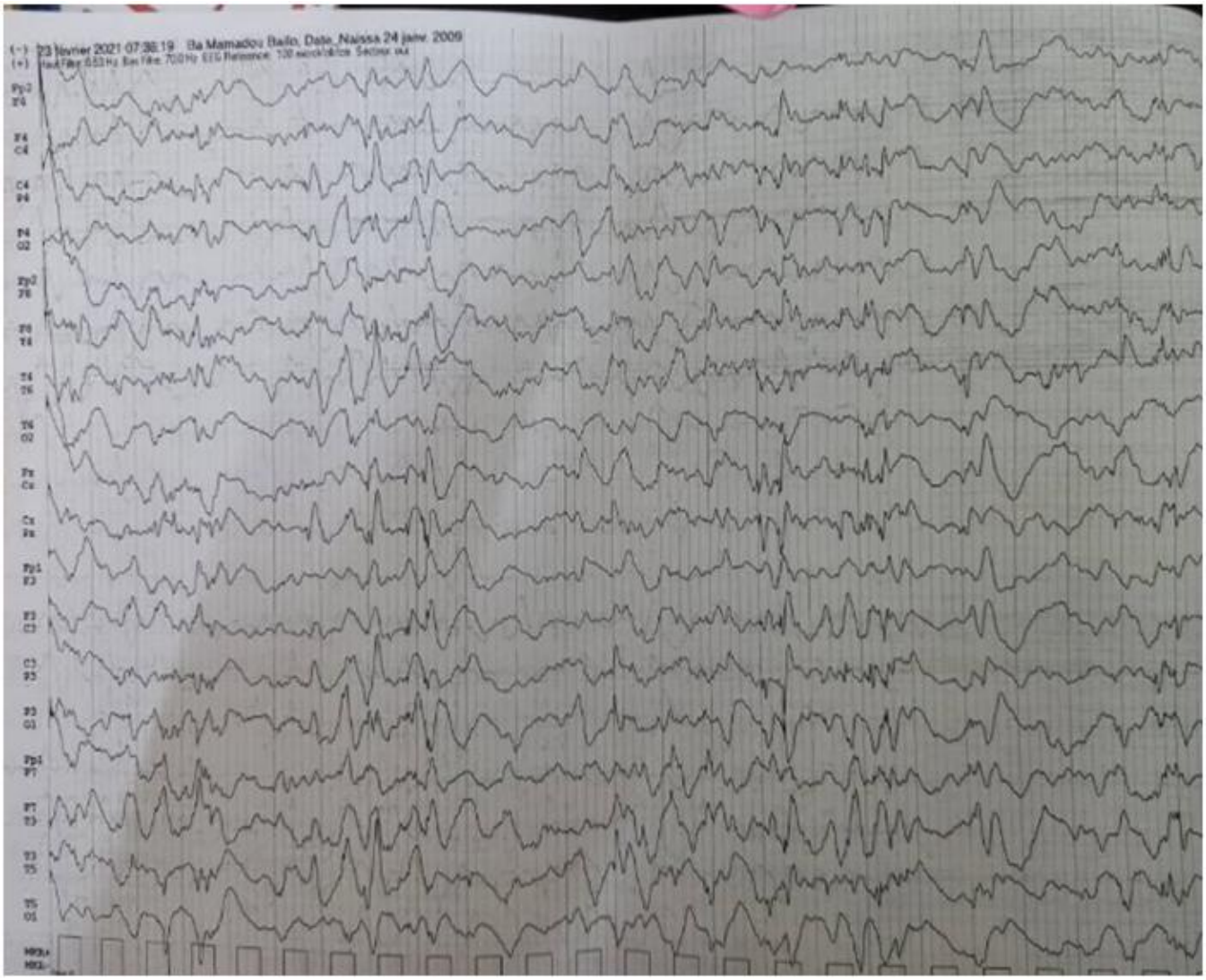

Figure 3: EEG wakefulness and slow sleep

Standby EEG tracing: with a background delta-theta rhythm at 2-4 C cycles per second, bilateral, symmetrical, synchronous and responsive to eye opening.

Hyperpnea not done and intermittent light stimulation ineffective.

Sleep: progressive with stage **I** and **II**; a background rhythm theta –delta, ample, irregular, diffuse, bilateral, symmetrical. Presence of some physiological figures of slow sleep (vertex points, outline of spindles and K complex).**Slow wave bursts notched peak in bi-parieto –temporal.**

Conclusion

EEG tracing of wakefulness and light slow sleep not very organized with irritative signs in parieto-temporal.

We have discussed metabolic encephalopathy:
1) A pseudo-hypo-parathyroid type endocrinopathy
2) Wilson's disease
3) Phare Syndrome

Paraclinical assessment

Blood:
- **Calcium**: **98** (**90-107mg /** l)normal
- **Magnesium**: 20normal
- **Creatinine:** 4.40normal
- **Protidemia**:71normal
- **ASAT: 29**normal
- **ALAT: 17**normal
- Phosphoremia: **31.8** [**27 - 45 mg** / l]normal
- Copper **1657 mg** / l [**794 - 2023] ab**normal
- Intact parathyroid hormone: **160.1pg** / ml [**1.59 - 7.21**]**ab**normal

Vit D (25 - 0H) <8,1mg / ml Deficiency **<10mg / mlab**normal

Diagnosis of a pseudohypoparathyroid complicated by rickets beingretained

Based on an increase in blood parathyroid hormone levels and a decrease in vitamin D.

Hence: Thyroid and parathyroid ultrasound
- Renal ultrasound for calcification, these two results returned to normal

Treatment initiated after endocrinology advice

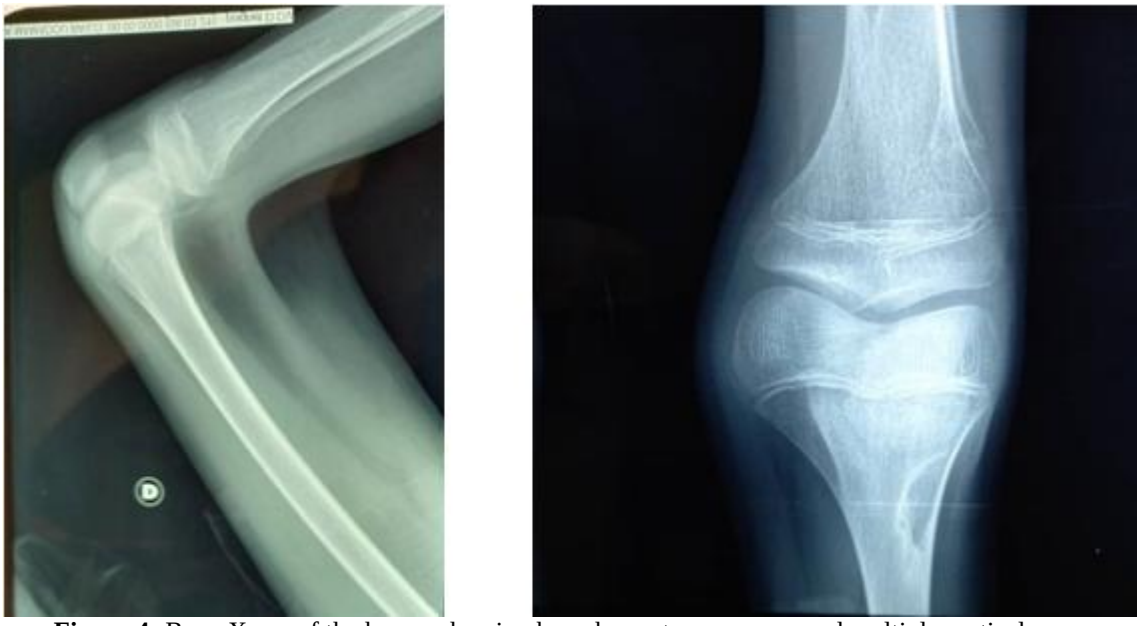

Figure 4: Bone X-ray of the knees: showing bone hyper transparency and multiple cortical gaps

- **Colecalciferol** or **vitamin D 3 100,000IU**: 2 ampoules taken once per month
- Carbamazepine suspension 1 case x 2 / day
- Physiotherapy

Evolution
Amendment of seizures
Released to meet in a month.

3. Discussion

Cerebral calcifications, when their severity exceeds a defined threshold in control individuals, can be revealed by neurological symptoms (in particular cognitive disorders or abnormal movements), psychiatric, or their association (Levine MA and Al).

They can also be discovered by chance, during a scan performed for various indications.The causes are diverse and require an etiological assessment

Extended (cf. the list of potential causes and a proposal for a diagnostic approach in (**A.Linglart et Al**). Cerebral calcifications can be the consequence of disorders of phosphocalcic metabolism, including hypoparathyroidism, hyperparathyroidism and pseudo-hypoparathyroidism.

The field of pseudo-hypoparathyroidism combines a biological picture of classic hypoparathyroidism (hypocalcemia, hyperphosphatemia, hypocalciuria, hypophosphaturia) with a normal or even increased dosage of serum PTH.

Clinical manifestations of rickets: hypotonia, delay in psychomotor acquisitions, kyphosis, convulsions or tetany. The phosphocalcic balance initially shows levels of calcium and phosphorus which are normal and which subsequently decrease. Alkaline phosphatases are almost constantly high. The **25 (OH) D** level is very low and **1, 25 (OH) 2** can have very variable levels (**Rojbi et al**).

Our patient corroborates with the data of the literature his rickets could be secondary to a dietary deficiency and the non-exposure to the sun caused by his immobility.

4. Conclusion

The discovery of bilateral cerebral calcifications, symmetrical must make evokes a metabolic pathological in the first place requires an examination the realization of a paraclinical assessment oriented in order to identify an etiology in order to prevent certain complication linked to the ulna which cause.

References

[1] **Albright F, Burnett CH, Smith PH, Parson W**. Pseudo-hypoparathyroidism - an example of "Seabright-Bantam syndrome". Endocrinology 1942; 30: 922-32.2.
[2] **A.Linglartet Al**, Clinical characteristics of the different forms of pseudo-hypo-parathyroidismIb (PHP Ib): autosomal dominant or sporadic? Annales d´endocrinologie, 200
[3] **Levine MA, Downs RW Jr, Moses AM et al**. Resistance to multiple hormones in patients with pseudo hypoparathyroidism. Association with deficient activity of guanine nucleotide regulatory protein. Am J Med 1983; 74 (4): 545-56.
[4] **Rojbi (Dr) ∗, I. Kammoun (Pr), R. Ben Mohamed (Dr), A. Morchdi (Dr), M. Ben Cheikh (Dr), L. Ben Salem (Pr), C. Ben Slama (Pr)** Pseudo familial hypoparathyroidism Department of endocrinology and metabolic diseases, national institute of nutrition, Tunis, Tunisia SFE Angers 2015 / Annalesendocrinology (2015) 474-492- P358
[5] **Weinstein LS**. Albright hereditary osteodystrophy, pseudo- hypoparathyroidism, and Gs deficiency. In Spiegel AM, ed. G proteins, receptors, and disease. Totowa, New Jersey: Humana Press, 1998: 23-56.3.