Subacute Sclerosing Panencephalitis in Children: A Reemerging Disease

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INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a slowly progressing brain disorder due to chronic infection with the Measles virus [1]. The re-emergence of this disease in recent years calls for particular vigilance in children.

OBJECTIVES

To identify characteristics of SSPE in a pediatric Tunisian population.

METHODS

Transversal study held in our department over a period of 3 years [2020-2023] including children diagnosed with SSPE according to the Dyken's modified criteria [1]. Patients were classified into four stages according to the clinical staging of SSPE [2]:

- **Stage 1:** Subtle decline in mental and scholastic performance
- **Stage 2:** Periodic myoclonus and severe mental decline
- **Stage 3:** Akinetic mutism with generalised spasticity
- **Stage 4:** Vegetative state

→ **Epidemiological, clinical, electroencephalographic, radiological and evolutionary data were analysed.**

RESULTS

CLINICAL FEATURES

- Twelve patients were included **(9 ♂ et 3 ♀)**
- Mean age of measles infection: **6 months [1- 12]**
- Mean age of SSPE onset : **45 months [21-73]**
- Acute disseminated encephalomyelitis-like presentation: **9 cases**
- Mean time to diagnosis: **80 days [7-240]**

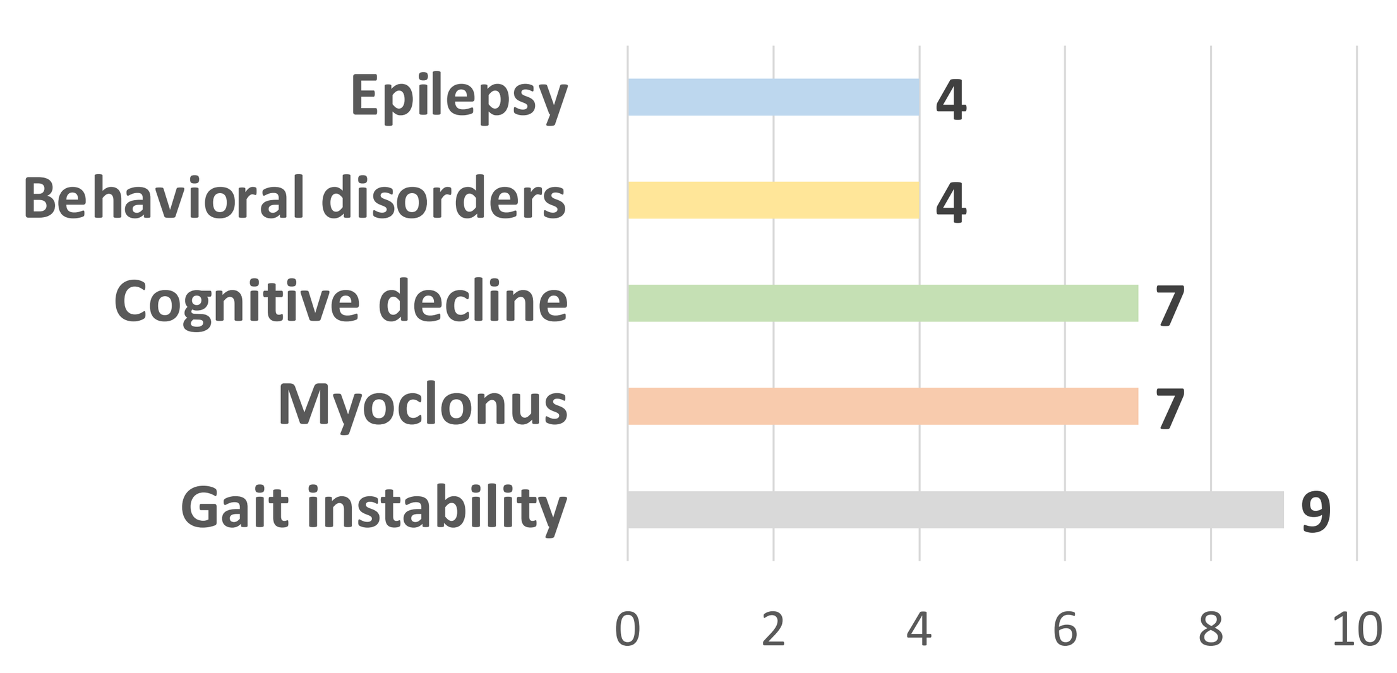


Figure 1. Onset symptoms

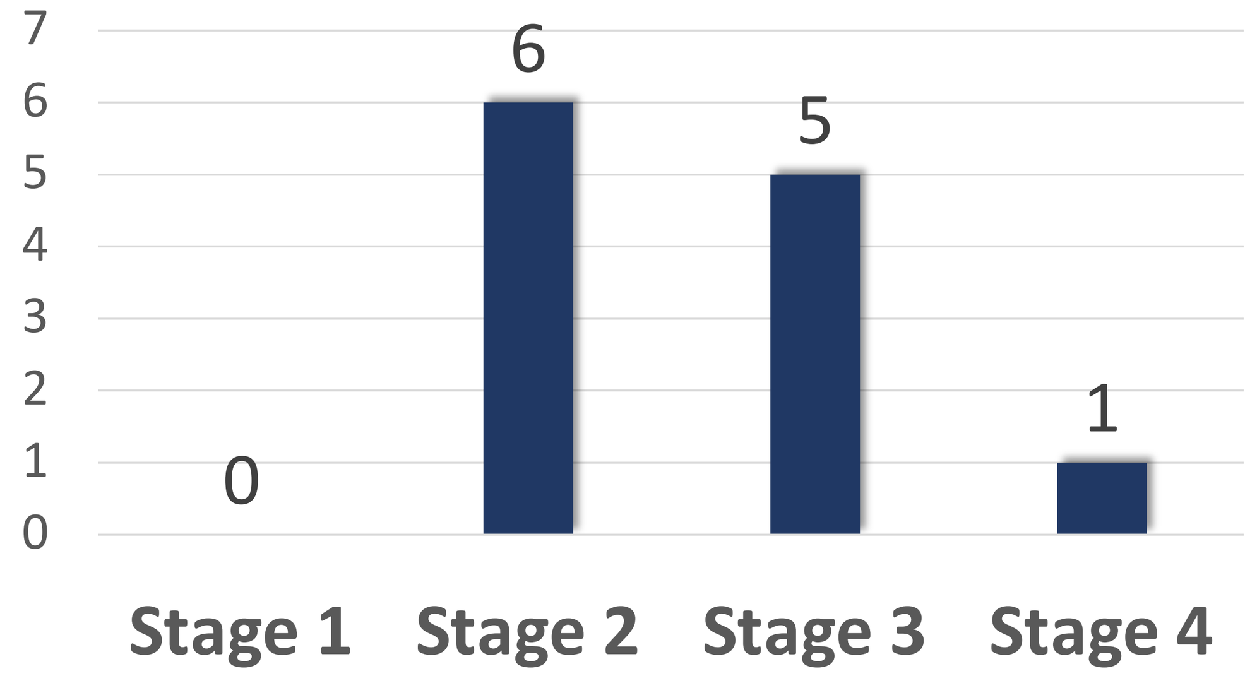


Figure 2. SSPE stages at the time of diagnosis

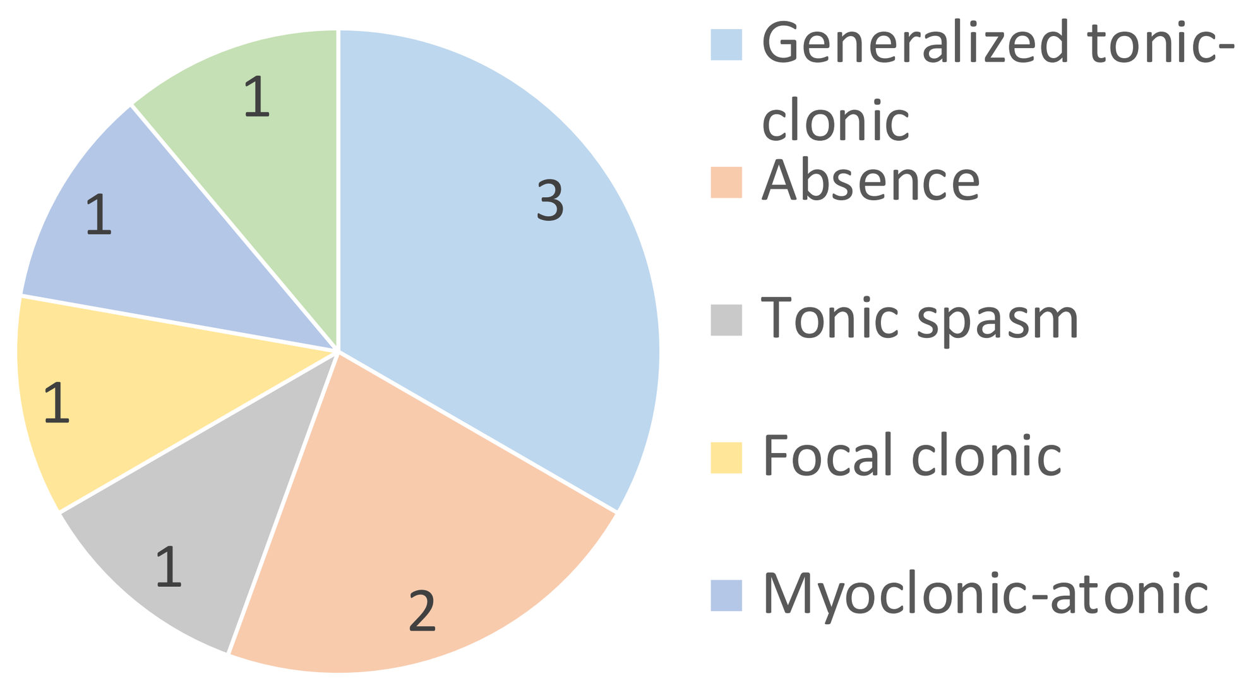


Figure 3. Seizures type found in epileptic patients

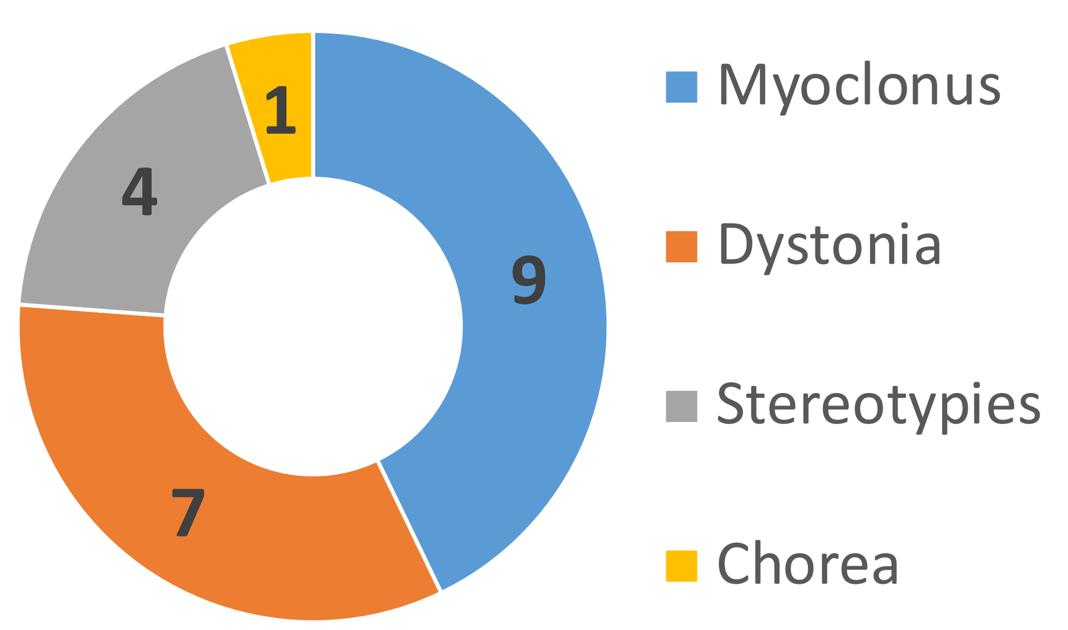
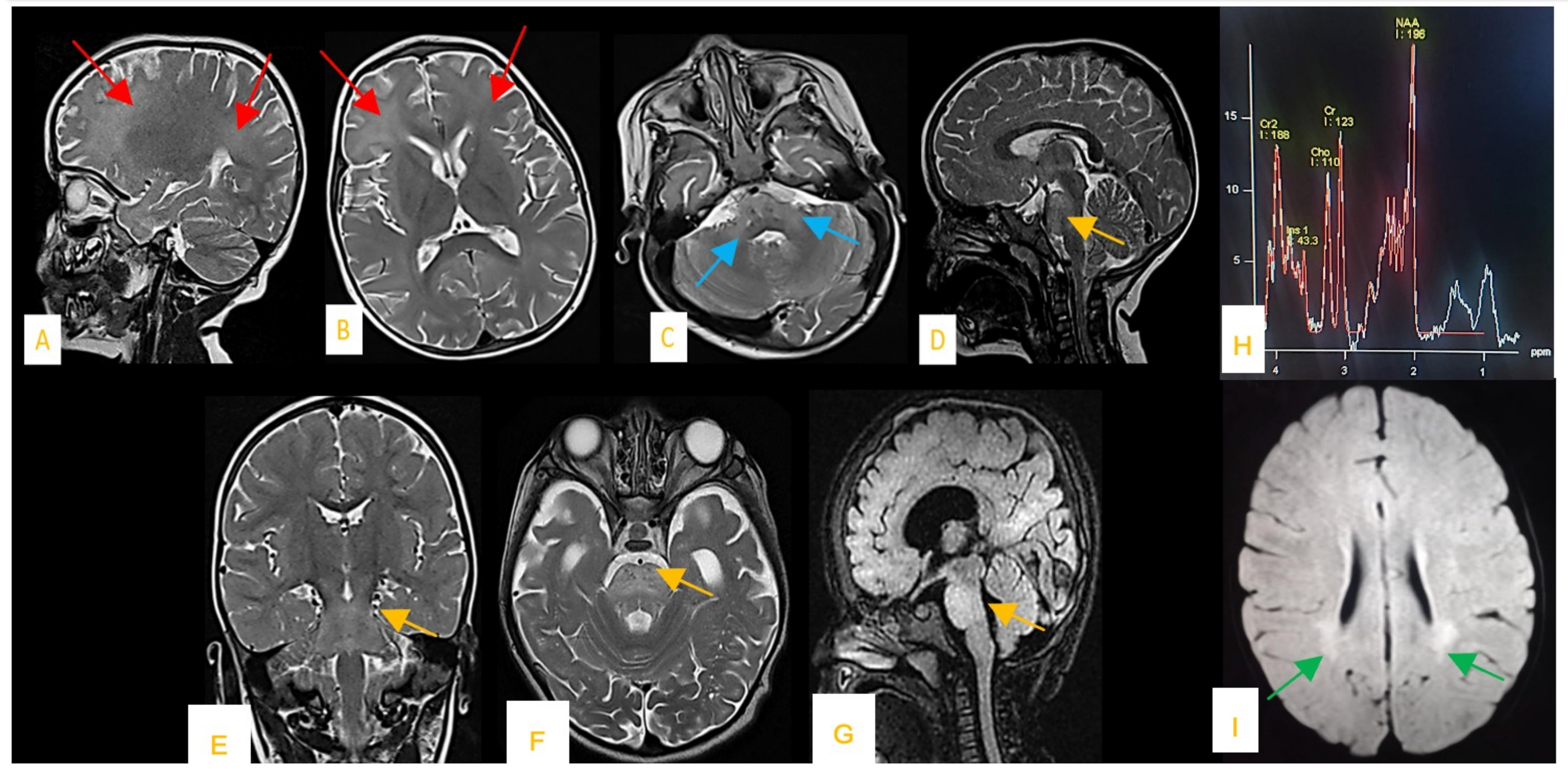


Figure 4. Movement disorders found in SSPE patients

PARACLINICAL FEATURES



Figures 5.A-F: Cerebral MRI in T2 **(A-F)** and T2-FLAIR **(G)** sequences showing fronto-parietal subcortical lesions (red arrows), in the middle cerebellar peduncles(blue arrows) and in the pons (yellow arrow) sparing the corticospinal tract; **Figures 5. H:** T2 FLAIR brain MRI **(G)** showing bilateral and symmetrical hyperintensities in periventricular regions, mainly posterior(green arrows); **Figure 5.I:** spectroscopy of patient 5.H, normal.

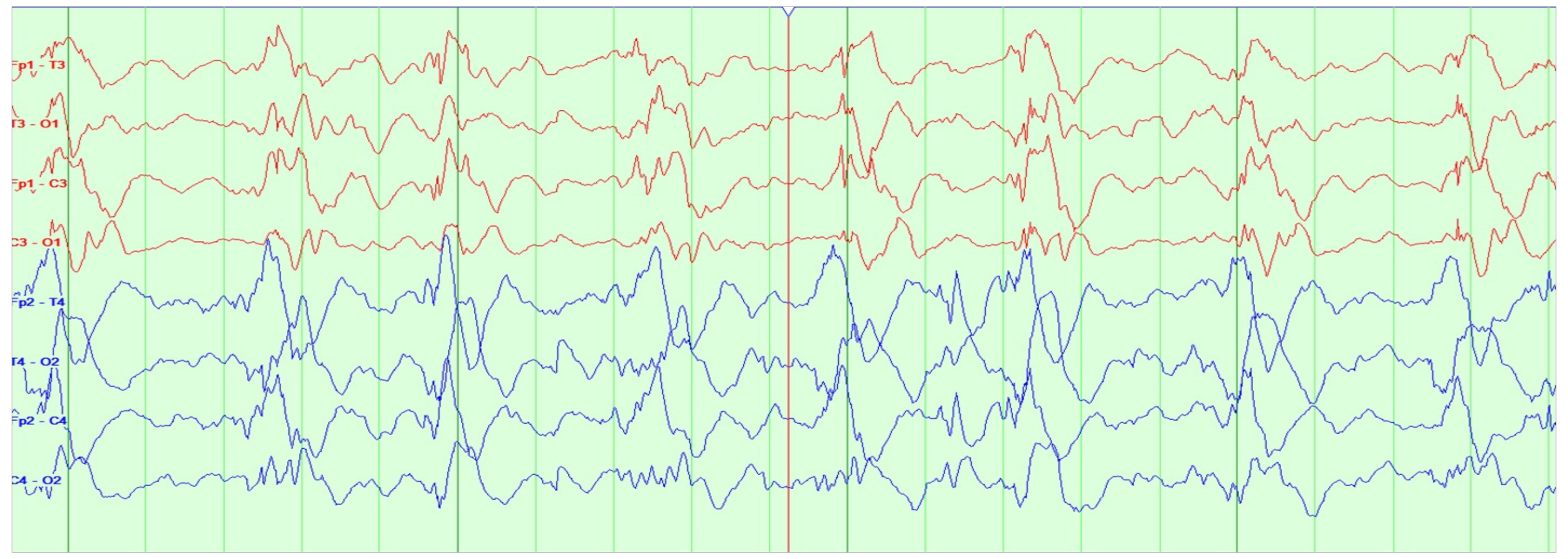


Figure 6. Electroencephalogram showing periodic complexes with generalised high amplitude slow waves, found in six patients

Table 1. Paraclinical findings

Paraclinical findings	n
Normal standard CSF analysis	12
Oligoclonal bands with high IgG index	6
Periodic complexes with generalised high amplitude slow waves (**Figure 6**)	6

TREATMENT AND OUTCOME

Table 2. Ethiopatogenic treatment

Ethiopatogenic treatment	n
Intravenous immunoglobulins	9
Isoprinosine	4

Table 3. Outcome of SSPE patients

Outcome	n
Vegetative state within 3 months	12
Swallowing difficulties	7
Status epilepticus	4
Drug resistant epilepsy	3
Dysautonomia	3
Aspiration pneumonia	2
Death	1

DISCUSSION & CONCLUSION

From January to April 2019, **3,141 cases** of Measles were reported in Tunisia [3]. Our patients contracted the infection during this outbreak, before reaching the age of vaccination. A World Health Organization expert group reported the global incidence as **4 to 11 SSPE** patients per **100 000** measles cases [1]. Since our study is single-centre, it is unfortunately not possible to estimate this incidence in Tunisia. Clinically, SSPE is characterized by **cognitive decline**, **periodic myoclonus**, **epilepsy, gait abnormalities**, and eventually, a **vegetative state**. Atonic seizures were the most frequently found type of seizures in our patients. This type of seizure remains rare in SSPE [1]. **Movement disorders** (MD) are key features in SSPE. Myoclonus is the most frequent MD followed by dystonia, stereotypies and chorea, [5] which is inline with our results. Typically **brain MRI** is normal at early stages [1]. At later stages, abnormalities are usually found in subcortical and periventricular white matter. Spectroscopy shows metabolic abnormalities of the brain. Low N-acetyl aspartate and elevated myoinositol values are characteristic in SSPE [1]. Unfortunately, **treatment options remain limited** and no therapy is known to be curative. Currently, the only hope against SSPE lies in its prevention.

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