Tension-Type Headache Revealing Morgagni-Stewart-Morel's Syndrome: A Case of 71-Year-Woman

Diallo SH^{1,2*}, Sogoba Y^{2,3}, Kotchene C¹, Yalcouye A², Traoré I¹, Sidibé M⁴,
Tolo N⁴, Guindo A⁴ and Maiga Y^{1,2}

¹Service de Neurologie CHU Gabriel Touré

²Faculté de Médecine et d'odontostomatologie, USTTB

³Service de Neurochirurgie CHU Gabriel Touré

⁴Polyclinique Guido, Bamako

***Corresponding Author:** Diallo SH, Service de Neurologie CHU Gabriel Touré.

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Abstract

Morel Stewart Morgagni syndrome or Internal Hyperostosis Frontal (HFI) is a metabolic craniopathy, most often asymptomatic, it can however be associated with a various manifestations including neuropsychiatric symptoms. We report the case of a 71-year-old patient with chronic tension-type headache seen in neurology consultation. The diagnosis was established based on the clinical and radiological criteria, and the patient benefitted symptomatic treatment and psychosocial support.

Keywords: Morgagni Morel Syndrome; Tension-Type Headache; Neurology Consultation

Introduction

Morgagni Morel's syndrome or hyperostosis frontalis interna (HFI) is characterized by a progressive thickening of the frontal bones with unknown a etiology, affecting mainly women over 35 years of age. It was first described by Morgagni in 1719 [1,2]. Stewart in 1928 and Morel in 1951 added the neuropsychiatric symptoms, which define the Morgagni-Stewart-Morel syndrome (MSM). Its manifestations include HFI, endocrine and metabolic disorders such obesity, dyslipidemia, hypertension, hyperglycemia and neuropsychiatric symptoms including headache, seizures, anxiety and cognitive impairment [3-6]. Its pathophysiology is yet to be elucidated and the hypothesis of the increased leptin levels or prolonged stimulation of estrogen are not universally accepted. However, genetic and environmental factors are interestingly being discussed [6,7].

We report the case of an outpatient followed for chronic tension-type headache.

Clinical case

We report the case of a 71-year-old woman who presented with moderate diffuse tension-type headache for several years, dizziness reported by the patient as unsteadiness when walking, generalized anxiety that worsened over the time. Symptoms continued to be embarrassing to the patient which led to her hospitalization. Her past medical history noted a gastropathy, and obesity as risk factor (BMI = 38) and hirsutism.

The metabolic assessment was normal except a dyslipidemia on Low Density Lipoprotein (LDL = 1.89/l) and Triglycerides (2.15g/l). The brain MRI revealed internal hyperostosis of the bilateral frontal bones associated with vascular leukopathy and early senile cortical atrophy (Figure 1 to 3).

We did not perform the EEG since she never reported seizure nor complained about it.

Figure 1: Coronal cut of the brain MRI showing hypo intensity T1 of the internal frontal bones after Gadolinium injection.

Figure 2: Axial cut of the brain MRI showing bilateral hypo intensity T2 of the internal frontal bones.

Figure 3: Axial cut of the brain MRI showing predominant bilateral hypo intensity T2 of the internal frontal.

Tension-type headache was suspected following the criteria of the International Headache Society (HIS, 2013), and the patient was followed on an outpatient basis. She was put on treatment with an analgesic level 2, Amitriptyline 25 mg and Paroxetine 20 mg (1 pill a day), and Acetyl leucine 500mg (2 pills three times daily). For the metabolic disorders, she received Fenofibrate 500 mg (1 pill two times daily). In addition, a psychosocial support was provided to the patient. On this treatment, the disease course was marked by a progressive improvement of the symptoms and she started with the daily work.

Discussion

Morgagni-Stewart-Morel's syndrome is mostly asymptomatic but can be associated with diabetes, osteoporosis, epilepsy, cognitive impairment or psychological symptoms and frontal syndrome [2,3]. Also, cases associated with migraine, depression and polyarthropathy have been reported in the literature [7-9]. In the most severe cases, it associates cortical atrophy and metabolic disorders [3].

The diagnosis is confirmed with brain imaging that can be either incidental or revealed by the symptoms of the disease. Its severity is assessed by Hershkowitz's anatomical classification which identifies five stages ranged from A to E [3].

Our patient presented with the classic symptoms commonly reported in the literature on Morgagni Stewart Morel's syndrome, including headache, generalized anxiety, hirsutism, diabetes. We did not find any depressive syndrome and the anxiety was quite embarrassing for the patient. We found diabetes, obesity and hirsutism among the endocrine disorders as reported in most of the cases. However, according to Moore, thyroid disorders are only reported in 4.4% of cases [2,3].

Although no gender specificity has been described, Morgagni Stewart Morel's syndrome is mainly observed in women from the age of 21. The first neuropsychiatric symptoms associated with IFH occur after the age 35 in most of the cases like the case we present here [2,5].

The radiological lesions are located on the internal table respecting the diploe, the frontal bone. The skull is known for being a privileged hormonal target; the estrogenic stimulus can reactivate the primary centers of ossification of the frontal bone which causes the abnormal bone growth [7,5]. In the case we report here, there was a bilateral frontal bone involvement.

The prevalence of HFI in relation with the body weight and identified the disease in 49% of cases, with a prevalence of 84%

in obese individuals and 16% in thin individuals [2,7], our patient presented with a severe obesity.

The etiopathogenesis of Morgagni Stewart Morel's syndrome is not fully elucidated, although many authors argued that endocrine imbalances involving sex hormones and genetic factors may be implicated [6]. The common neuropsychiatric symptoms reported in the literature includes depression up to melancholy and schizophrenia, senile dementia [2], these symptoms were not seen in our patient.

Conclusion

Pathology of incidental discovery with unclear physiopathology, the clinical and radiological correlation helps to diagnose this disease and the management is multidisciplinary.

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