CASE REPORT

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Morgagni-Stewart-Morel syndrome: A new case

Lahoussaine Abainou, Hamza El Jadi, Sanae El Hadri, Hicham Baizri

ABSTRACT

Introduction: Morgagni–Stewart–Morel (MSM) syndrome is defined as the presence of hyperostosis frontalis interna (HFI) associated with metabolic, endocrine, and neuropsychiatric disorders. Here, we present an MSM syndrome case involving a woman with symptoms of depression and infertility. Case Report: A 38-yearold woman admitted for inaugural diabetic ketosis and she reported a history of severe frontal headache of which she had suffered for several years associated to depression, obesity, and infertility. Axial cranial computed tomography (CT) scan showed bilateral thickening of the inner table of the frontal bone. Her endocrine dysfunctions and neuropsychiatric symptoms are interpreted as components of Morgagni-Stewart-Morel syndrome and may correlate with the severity of the frontal hyperostosis and the cortical atrophy. Conclusion: We describe a case of MSM syndrome, presenting many of the clinical features described in the literature. The exact etiology of HFI and MSM syndrome remains unclear; the most interesting theories relate to estrogen dysfunction, obesity and leptin dysfunction, and genetics.

Keywords: Endocrine and neuropsychiatric disorders, Hyperostosis frontalis interna, Morgagni– Stewart–Morel syndrome

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INTRODUCTION

Morgagni–Stewart–Morel syndrome is an association of clinical features with radiological findings (HFI). The common clinical features are headache (migrainous), psychotic symptoms, metabolic and endocrine disorders (obesity, diabetes mellitus, and thyroid disease), hirsutism, osteoporosis, and menstrual disorders, but these associations are mostly based on case reports, and no clear consensus exists on the definition of the syndrome.

CASE REPORT

A 38-year-old woman, married without children, admitted in February 2019 to the Endocrinology Department for an inaugural diabetic ketosis. During the interrogation the patient complained that she had an eight-year history of severe chronic headaches never explored and she was being followed up by a psychiatrist for depression. She also reported a medical history of obesity, infertility for several years. On examination she was obese, depressed, and her body mass index (BMI) was 36 kg/m². Axial cranial CT scan showed bilateral thickening of the inner table of the frontal bone (Figure 1A and B). Lab values are given in Table 1.

These endocrine dysfunctions and neuropsychiatric symptoms are interpreted as components of MSM syndrome. For the severe headache, the surgical indication was discussed with the neurosurgeons but the patient refused immediately surgery and she

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Figure 1: Axial cranial CT scan evidences the bilateral thickening of the inner table of the frontal bone (red arrows).

was treated by analgesics with improvement under tramadol.

DISCUSSION

Morgagni–Stewart–Morel syndrome was first described in 1719 by Giovanni Battista Morgagni in an obese female patient during autopsy who had hirsutism and HFI defined as thickening of inner table of a skull [1].

The modern concept of its clinical significance was initiated in 1928 after Stewart reported three cases of the syndrome diagnosed at autopsy. The first living case was reported by Morel in 1930 [2, 3].

Table 1: Laboratory studies

Hyperostosis frontalis interna is a morphological pattern of the frontal bone that is usually an incidental finding in X-ray, cranial computed tomography (CCT), or magnetic resonance imaging (MRI) studies [4].

Morgagni–Stewart–Morel syndrome is defined as the association of HFI with metabolic, endocrine, and neuropsychiatric disorders. But these associations are mostly based on case reports, and no clear consensus exists on the definition of the syndrome [4].

The etiology and pathogenesis behind skull thickening and MSM syndrome are till now not clear; the most interesting theories relate to extended exposure to estrogen, obesity and elevated leptin level, and genetics [5, 6].

The common clinical features are headache, hirsutism, menstrual disorders, galactorrhoea, obesity, diabetes mellitus, depression, irritability, fatigability, transitory hemiplegias, hearing impairment, cranial nerve palsies, muscle weakness, and seizures [7–9].

The treatment is generally symptomatic but there is a case of MSM syndrome described in the literature with violent headaches predominant in the clinical course treated surgically and the pain ceased immediately after the operation. The authors discuss surgical treatment especially in cases with high intensity headaches [10].

Morgagni–Stewart–Morel syndrome is also called as metabolic craniopathy due to its wide range of symptoms and endocrinal dysfunctions [11].

Biohumoral marker patient's value	Normal value	
Glycated hemoglobin (HbA1c) (%)	8.14	0-5.6
Calcium (mmol/L)	2.13	2.12-2.55
Phosphorus (mmol/L)	1.02	0.2–1.6
Albuminemia (g/L)	40.5	35-50
Calciuria (mmol/24 h)	1.58	0.5-7.5
Parathyroid hormone (PTH) (ng/L)	50	15-65
25-Hydroxyvitamin D (nmol/L)	61	>75
Total cholesterol (mmol/L)	6.5	<5.10
Uric acid (mmol/L)	0.50	0.15-0.35
Creatininemia (mol/L)	57	50-90
C-reactive protein (mg/L)	8.73	<5
Serum glutamic-oxaloacetic transaminase (SGOT) (U/L)	18	<50
Serum glutamic-pyruvic transaminase (SGPT) (U/L)	12	<65
Alkaline phosphatase (U/L)	56	35–104
Thyroid-stimulating hormone (TSH) (µIU/mL)	1.86	0.27-4.2
Follicle-stimulating hormone (FSH) (IU/L)	2.5	1.7-7.7
Luteinizing hormone (LH) (IU/L)	6.8	1–11.4
17 b Estradiol (pg/mL)	19.4	12.5-366
Prolactin (µIU/mL)	412	102–496

CONCLUSION

The known symptoms of MSM syndrome are mostly based on case reports, and they tend to be nonspecific. Patient with MSM syndrome may present varied symptoms associated to thickening of the inner table of the frontal bone, thus lead to difficulty in diagnosis.

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Author Contributions

Lahoussaine Abainou – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hamza El Jadi – Conception of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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