Case Report

Neuropsychological profile of a male psychiatric patient with a Morgagni-Stewart-Morel syndrome


In 1765 Giovanni Morgagni described a syndrome consisting of hyperostosis frontalis interna (HFI), obesity and hirsutism. In 1928 Stewart and in 1930 Morel added neuropsychiatric symptoms, e.g. depression and dementia, which led to the definition of the Morgagni-Stewart-Morel Syndrome (MSM). Although mostly women were characterized in literature no gender specificity is demanded. This case report presents the rare case of a 66 year old male psychiatric patient with Morgagni-Stewart-Morel Syndrome. The patient complained of loss of concentration and difficulties with activities of daily living. Admission diagnosis was an opioid misuse on the basis of a chronic pain syndrome. In this case report we are describing clinical features, the patient history and technical (MRI) and neuropsychological tests. Although severe psychiatric symptoms and neuropsychological deficits are commonly seen in these patients, our patient showed only mild symptoms. This case report shows the possibility of a male patient with MSM. If MSM is a separate entity or just an epiphenomena of hormone dysregulation should be investigated in further studies.

Significant Outcomes

- The Morgagni-Stewart-Morel (MSM) syndrome can also affect men.
- An example for future research to find definite diagnostic criteria of MSM.

Limitations

- The Morgagni-Stewart-Morel syndrome is debated.
- This is only a case report.

Introduction

Giovanni Battista Morgagni described in 1765 a triad consisting of hyperostosis frontalis interna (HFI), obesity and hirsutism (1). HFI is the thickening of the inner table of the os frontale. This thickening has been found in hominids dating back 1.5 million years, like homo erectus and homo neanderthalensis (2). A large anthropological study of 3725 skulls (3) showed a prevalence of 24% of HFI in women and only of 5.2% in men. The origin of it remains unclear. One cause could be hormone dysregulation with elevated leptin levels or a testosterone–estrogen dysbalance (3–5). This HFI can lead to neuropsychological deficits without the diagnosis of a Morgagni-Stewart-Morel Syndrome (6). De Zubicaray et al. (6) examined a 74-year-old patient with an isolated HFI, which led to the following
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Neuropsychological deficits: she had selective deficits in self-ordered working memory, which was associated with the compression of the dorsolateral frontal cortex, which was the result of the HFI. Her mini-mental state examination (MMSE), WAIS-R IQ and visual and verbal short-term memory were normal. She had considerable difficulty with tasks requiring the active manipulation and monitoring in spatial and non-spatial memory.

Stewart and Morel associated the Morgagni triad with neuropsychiatric symptoms (7–9). These neuropsychiatric symptoms were not exactly specified and to this day there are no international standardised criteria for this diagnosis defined.

The concept of the MSM has sparked a long discussion if the MSM is a separate disease entity or just an epiphenomena (10,11).

MSM syndrome features the following symptoms:

1. HFI;
2. obesity;
3. hirsutism;
4. neuropsychiatric symptoms.

A new publication points to a genetic base of the MSM (12). A pair of 71-year-old female twins both with a MSM and severe neuropsychiatric impairments were found. An older publication shows the occurrence of MSM in a family (13).

Case report

Patient history

The 66-year-old male caucasian patient was voluntarily admitted at the psychiatric hospital of Muensterlingen/Switzerland on to the ward for geriatric psychiatry with an opioid abuse. He had no prior contact to a psychiatric facility. His goal was to reduce the opioid intake, as he complained of loss of concentration and difficulties with activities of daily living, for example, organising official papers. He was a widower and had two healthy siblings. In his family there is no history of dementia or psychiatric disorders. Before receiving a pension he was an electromechanical engineer with a higher degree of education.

The opioid abuse was based on a somatic symptom disorder with chronic pain. This was based on an abdominal injury owing to a car accident 30 years before the admission. This abdominal injury led to multiple operations owing to hernias. The patient also had a spinal disc herniation at the location L3/4 to S1 with a cauda equina syndrome 5 years before the admission. This led to an operation with a decompression and hemilaminectomy.

At admission he was consuming over 1500 mg Oxycodone/day, additionally also Paracetamol up to 2 g/day. Owing to his massive amount of opioid consumption he already had complication like pruritus and constipation.

He also had multiple metabolic diseases.

These were:

- arterial hypertension;
- diabetes mellitus type 2;
- ulcus cruris arteriosus;
- obesity, BMI: 33 kg/m²;
- dermatitis seborrhoica;
- gall bladder stones;
- chronic renal disease;
- sleep apnoea syndrome.

To get to the clinic he drove by himself over 2000 km from his home in Spain to the clinic in Switzerland with his car, without any complication, for example, accidents or getting lost.

Somatic examination

There were no focal neurological signs or symptoms, but symptoms of polyneuropathy like loss of the achilles tendons reflex. He had slight oedema of his lower limbs. Besides the cured ulcera, there were no other remarkable findings of the examination of his cardiovascular system.

Psychopathology

Psychopathologically he showed a depressive mood, difficulty to remain focussed and short-term memory impairment (remembered one of three words after 15 min). Otherwise he showed no productive psychotic symptoms (e.g. hallucinations or delusions) or impairment of his abstraction ability. He complained of difficulties with his drive in the morning. Testing of his concentration revealed no pathology. The patient also complained of an anhedonia, but had no lack of motivation. There were no complaints of depersonalisation. No formal thought disorder could be found. The patient had no symptoms of anxiety or obsessive–compulsive disorder. He was fully awake and fully orientated.

He could not determine exactly when it started, but he stated that it was a minimum of 1 year. His siblings stated that his personality had changed in the last few years. The patient had a change of his social conduct and was alienated from his siblings.

We diagnosed him with the following psychiatric disorders:

- major depressive disorder;
- opioid abuse;
- persistent personality change owing to the substance abuse and the chronic pain syndrome.
Imaging

The Patient received a magnetic resonance imaging, as a standard procedure. It showed, next to the symmetric thickening of the inner table of the frontal bone, also vascular white-matter lesions (Fig. 1a and b).

Neuropsychological testing

The Patient was tested at the end of his stay using the German version of the Symptom-Checklist-90-R (SCL-90-R) (14) and with the German version of the Consortium to Establish a Registry for Alzheimer’s Disease Neuropsychological Assessment Battery (CERAD-NAB) Plus (15–17). The SCL-90-R is a self-report measure that was designed to identify the psychological stress of patients owing to their physical and psychical symptoms. With its 90 items the SCL-90-R assesses the following nine scales: somatisation, obsessive-compulsive, interpersonal sensitivity, depression, anxiety, hostility, phobic anxiety, paranoid ideation and psychoticism. The patients answer on a five-level Likert scale. The SCL-90-R testing showed the following (Table 1): while data of the most scales were within normal range (t-value ≤ 60), the scores of the patient were increased within the scale of somatisation (t-value = 67) and psychoticism (t-value = 63). Somatisation is associated with physical functional disorders; psychoticism with a feeling of isolation, as well as alienation and psychotic feelings. The feeling that something is wrong with his body (item 87) was significant within the scale of psychoticism.

The CERAD-NAB Plus describes neuropsychological abnormalities related to Alzheimer’s disease and other dementia syndromes. The Patient underwent a modified version of the battery containing the following tasks: MMSE, word list – total (the sum of learned words over three word-learning lists), figures – copy, word list – delayed recall, word list – recognition and figures – delayed recall. In addition, three subscales were formed: word list – intrusions, word list – savings and figures – savings. In addition, the neuropsychological tests phonemic fluency and Trail Making Test were added. Overall, the results of the patient were within normal range (Table 2).
Electroencephalography (EEG)

The EEG was normal with α waves, with no abnormal activity.

Laboratory tests

Laboratory tests showed a persistent elevation of Creatinine (93–127 µmol/l; norm: 62–107 µmol/l). In addition, a persistent elevation of C-reactive protein (CRP) (16–35 mg/l; norm: <5 mg/l). Sodium and Potassium was normal, as were aspartate amino transferase and alanine amino transferase. Gamma-glutamyl transferase was elevated (330–370 U/l; norm: <66 U/l). The Patient was anaemic (haemoglobin: 88–124 g/l; norm: 140–180 g/l), with normal erythrocyte indices. He also had an erythropenia (2.9–3.6 × 10¹²/l; norm: 4.6–6 × 10¹²/l). Folic acid and Vitamin B₁₂ were in normal ranges, as were albumin and brain natriuretic peptide.

A lumbar puncture was not possible, owing to the operation on the spine of the patient.

Medication

At admission the patient had the following medication: 25 mg Hydroxin/day, 5 mg Bisoprolol/day, 30 mg Duloxetine/day, 4 mg Dimentidin/day, 1400 mg Oxycodone/day, 40 mg Pantoprazole/day, 1.5 mg Risperidon/day, 10 mg Torasemid/day, 30 UI Insulin/day.

We changed the medication to: 5 mg Bisoprolol/day, 5 mg Amlodipine/day, 25 mg Hydroxin/day, 4 mg Dimentidin/day, 100 mg Torasemid/day, 50 mg Allopurinol/day, 50 UI Insulin/day, Budesonide 400 mg/day and 320 mg Oxycodone/day.

Table 2. Z-values the patient reached in the tasks of the CERAD-NAB Plus (German version): 1 Mini-Mental State Examination, 2 word list – total, 3a-c tree trials of the word-learning list, 4 word list – delayed recall, 5 word list – intrusions, 6 word list – savings, 7 word list – recognition, 8 figures – copy, 9 figures – delayed recall, 10 figures – savings, 11 phonemic fluency, 12 Trail Making Test (Part A), 13 TrailMaking Test (Part B) a 14 Trail Making Test (B/A)

![Cerad-Batterie](https://www.cambridge.org/core/terms).

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Table 2. Z-values the patient reached in the tasks of the CERAD-NAB Plus (German version):

<table>
<thead>
<tr>
<th>Task</th>
<th>Z-value</th>
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<tbody>
<tr>
<td>Mini-Mental State Examination</td>
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<td>Word list – total</td>
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<td>Tree trials of the word-learning list</td>
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<td>Word list – delayed recall</td>
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<td>Word list – intrusions</td>
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<td>Word list – savings</td>
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<td>Word list – recognition</td>
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<td>Figures – copy</td>
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<td>Figures – delayed recall</td>
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<td>Figures – savings</td>
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<td>Phonemic fluency</td>
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<td>Trail Making Test (Part A)</td>
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<td>Trail Making Test (Part B)</td>
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<tr>
<td>Trail Making Test (B/A)</td>
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Discussion

We diagnosed the patient with a MSM, as he showed the main symptoms of it. As there is no gender specificity, men also can have a MSM.

The elevation of the CRP is of unknown significance, as he had no elevation of leukocytes, fever or any other symptoms of inflammation.

The subjective complaints of the patients could not be shown in the neuropsychological testing. This could be because either the tests were not sensitive enough or the disease was not progressed so much. Considering that the patients, which are featured in the literature were older (e.g. 71 year (12)), the patient could develop severer symptoms later. The remaining medication could also be responsible for his complaints. Considering the amount and the duration the patient was taking the medication, and his ability of him to organise the drive to the clinic, it is highly likely that the patient was already accustomed to this medication. An isolated HFI can also lead to mental impairment (9).

We presume that the impression of the frontal lobes of the brain and the remaining medication, but not the chronic opioid abuse, were responsible for the complaints of the patient. Further testing of the patient will illuminate if there is a progression.

This case report also demonstrates that the medication of geriatric patients should be continually monitored, especially when they suffer chronic pain. A higher opioid medication did not have a significant effect on his pain level.
As the HFI seems to exist since prehistoric times (2,3) and there may be a genetic basis for the MSM (12,13), it could be that the MSM is a separate pathological entity. As this disease is not well-known or characterised, further studies and definite diagnostic criteria could provide further insight into this disease. What we would also like to achieve, is to raise awareness, so a HFI is not classified as a random radiological finding, but is seen in a greater context.

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Conflicts of Interest

All authors declare no conflicts of interest.

References