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Olgu Sunumu

Morgagni-Steawart-Morel syndrome: Case report

Morgagni-Steawart-Morel sendromu: Olgu sunumu

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Abstract

Hyperostosis frontalis interna (HOFI) is characterized by the benign growth of the inner plate of the frontal bone. It is most common in older women. Etiology is unknown. Morgagni-Stewart-Morel syndrome (MSM) is characterized by HOFI, obesity, hirsutism and mental illnesses but these associations are mostly based only on case reports. MSM syndrome is a misunderstood and less reported pathology. The clinical presentation is not well defined and its finding is usually coincidental. The patient may present variable symptomatology. The idea that we should keep in mind is that HOFI represent the main pattern of MSM. Thus, it is highly important to specify the presence and degree of HOFI in radiological reports.

Keywords: Morgagni-Stewart-Morel syndrome; hiperostosis frontalis interna

Öz

Hiperostozis frontalis interna(HOFI) frontal kemiğin iç tabulasının iyi huylu büyümesi ile karakterizedir. En sık yaşlı kadınlarda görülür. Etiyoloji bilinmemektedir. Morgagni-Stewart-Morel sendromu (MSM); HOFI, obezite, hirsutizm ve zihinsel rahatsızlıklar ile karakterizedir ancak bu ilişkiler çoğunlukla vaka raporlarına dayanır. MSM sendromu daha az anlaşılan ve bildirilen sendromdur. Hasta çeşitli semptomlar gösterebilir. HOFI'nin MSM'nin bir paterni olduğu akılda tutulmalıdır. Radyolojik raporlamada bu nedenle HOFI varlığını ve derecesini belirtmek önem teşkil eder.

Anahtar kelimeler: Morgagni-Stewart-Morel sendromu; hiperostozis frontalis interna

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Introduction

Morgagni-Stewart-Morel syndrome (MSM) is characterized by metabolic, endocrine and neuropsychiatric disorders accompanying hyperostosis frontalis interna (HOFI)[1,2].The main finding of the disease is HOFI; defined as the benign growth of the inner surface of the frontal bone [3].The disease can manifest with a wide variety of endocrine and neuropsychiatric conditions. Except HOFI, other findings are partially monitored in most of the cases [1,4].We present MSM patient with HOFI and neuropsychiatric findings under the light of radiological and clinical findings.

Case

Our patient is 62-year-old female who was admitted to the hospital with a long-term headache. Since 2004, she has been diagnosed with hypertension and since 2014 with diabetes mellitus. The patient was obese (BMI =30) and presented also a hirsutism. Despite of rheumatological symptoms, there was no arthritis. A dupytrien contracture on the 4th finger was present. The patient was followed and treated for systemic connective tissue disease. Although, the patient consulted psychiatry outpatient clinic several times, she did not have a psychiatric diagnosis other than recurrent depression.

In the family antecedents, the father had a history of diabetes, cirrhosis, and congestive heart disease and mother had a history of hypertension.

Abdominal ultrasonography (USG) revealed a grade 1 hepatos- teatosis in the liver while the other abdominal organs were nor- mal. The thyroid parenchyma was heterogeneous and pseudo nodular. Several smoothly heterogeneous isohypoechoic nodules were detected on the parenchyma of the thyroid. As Anti TPO rate (ATA) was high (38.2), the diagnosis of thyroiditis was set.

Cranial MRI showed an increase in intertabular distance and thickening in the parietal and frontal bones. Finger-shaped indentations were observed in the internal tabula. Appearance was compatible with HOFI (Figure).

The diagnosis of MSM was established on the base of several collected data: age group, presence of obesity, hirsutism, diabetes mellitus, thyroiditis, connective tissue disease and their association with HOFI in the MRI.

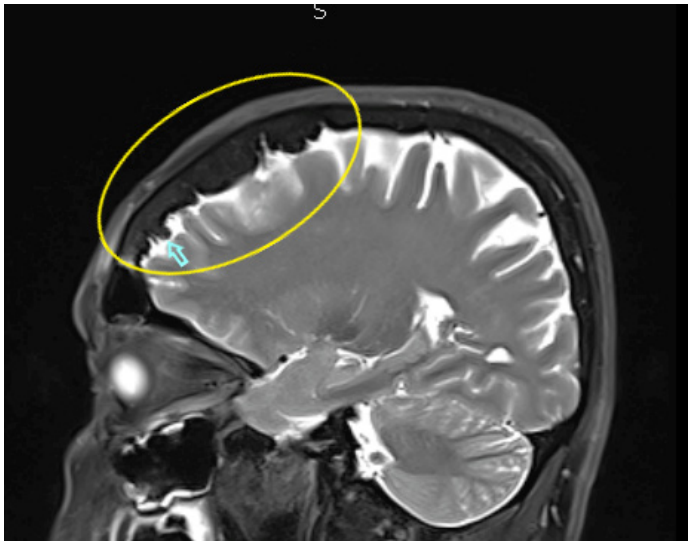

Figure: External tabulais regular,increase ofintertabular distance is compatible with HOFI (Yellow circle), finger-like bones extending to cerebrospinal fluid (Blue arrow)

Discussion

In 1719, Morgagni reported for the first time the relationship between the frontal bone thickening, obesity and hirsutism. Stewart added, in 1928, that neuropsychiatric problems may be associated with this condition. The first live case was reported by Morel in 1930. The disease is now defined as the presence of HOFI, which is variably related to metabolic, endocrine and neuropsychiatric disorders [2].

The etiology of MSM syndrome is not yet fully explained. However, some authors have reported that the mechanism behind the syndrome is due to endocrine imbalance involving sex hormones. The proof which support their theorem is that HOFI is only seen in male patients with gonadal disorders [1].

Ruhli and Hanneberg presented another theorem incriminating the leptin hormone. It was reported that leptin metabolism contributes to HOFI formation by the increase of bone metabolism [1,5].

Rosatti evaluated four generations of patients' family and reported experimentally that the disease had an autosomal dominant genetic transition [6]. Genetic theorem is supported in the literature by the case of twins. However, the genetic infrastructure of MSM is still unclear [1,7].

The main finding of the disease is HOFI which is a morphological form of frontal bone and usually appear as single or multiple

bilateral nodules in the inner tabula. Diploe and calvarial mid line characteristic are unchanged[1]. It is most commonly seen in older women. The etiology is unknown. This condition is usually not important clinically and finding is coincidental. [3].

HOFI is divided into 4 groups according to the morphological and histopathological classification of Hershkovitz:

Type A - Single or multiple, single-sided or double-sided, isolated bone thickness, less than 1 cm. usually occupying less than 25% of the frontal bone surface area.

Type B - Increased bone thickness without clear margins. Usually less than 25% of the frontal endocranial surface area is affected.

Type C - wider nodular bone overgrowth associated with irregular thickening up to 50% of the frontal endocranial surface (more than one nodular bone extension).

Type D - Large effected area bone, involving more than 50% of the frontal endocranial surface [8].

MSM syndrome is also called metabolic craniopathy. It is a condition associated with a wide range of endocrine problems such as diabetes mellitus, diabetes insipidus, and hyperparathyroidism. Other signs and symptoms include headache, dizziness, hirsutism, menstrual problems, galactorrhea, obesity, depression, and seizures [2]. The data are mostly based on case reports and there is no clear consensus on the definition of the syndrome [1].

In our case, patient had headache, hirsutism, obesity, diabetes mellitus, recurrent depression and additional symptoms related to thyroiditis and connective tissue disease which were not previously described. According to Hershkovitz, our case was classified as a HOFI type C.

Conclusion

Morgagni-Stewart-Morel is a less understood and reported syndrome. It should be kept in mind that HOFI is the main pattern of MSM therefore it is important to indicate the presence and the degree of HOFI in radiological reports.

Declaration of conflict of interest

The authors received no financial support for the research and/or authorship of this article. There is no conflict of interest.

*This case report was approved by the local ethics committee. Informed constent was collected from the patient.

References

1. Attanasio F,Granziera S, Giantin V, Manzato E. Full Penetrance of Morgagni-Stewart-Morel Syndrome in a 75-Year-Old Woman: Case Report and Review of the Literature. J Clin Endocrinol Metab 2013; 98: 453-57.
2. Nallegowda M, Singh U, Khanna M, Yadav SL, Choudhary AR, ThakarA .Morgagni Stewart Morel syndrome-additional features. Neurol India 2005; 53: 117-19.
3. Smith S, Hemphill RE. Hyperostosis frontalis interna. J. Neurol. Neurosurg Psychiatr 1956; 19: 42-45.
4. Klein D, Rosatti P, Mach RS, Ferrier PE, Rauch S: Morgagni-Morel syndrome: an independent clinical and genetic entity, demonstrated in a family study covering 4 generations. Schweiz Arch Neurol Neurochir Psychiatr 1973;112: 239-50.
5. Ruhli FJ, Henneberg M. Are hyperostosis frontalis interna and leptin linked? A hypothetical approach about hormonal influence on human microevolution. Med Hypotheses. 2002; 58: 378-81.
6. Rosatti. Family affected by hyperostosis frontalis interna (Morgagni-Morel Syndrome) through 4 successive generations. J Genet Hum 1972; 20: 207-52.
7. Koller MF, Papassotiropoulos A, Henke K et al. Evidence of a genetic basis of Morgagni-Stewart-Morel syndrome. A case report of identical twins. Neurodegener Dis. 2005; 2: 56-60.
8. Hershkovitz I, Greenwald C, Rothschild BM, et al. Hyperostosis frontalis interna: an anthropological perspective. Am J Phys Anthropol 1999; 109: 303-25.