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HOMO — Journal of Comparative Human Biology 57 (2006) 19–27
Journal HOMO of Comparative Human Biology
www.elsevier.de/jchb

Hyperostosis frontalis interna, a genetic disease?: Two medieval cases from Southern Poland

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Received 16 March 2005; accepted 30 August 2005

Abstract

Two cases of thickening of the internal tables of the frontal bones (hyperostosis frontalis interna, (HFI)) have been examined. These were two female skeletons from the 16th century Dominican Church in Raciborz (Southwest Poland). The similarity of their morphological and metrical traits indicates that they could be related, and suggests that HFI is likely to have a genetic base. These two skeletons are the subject of an analysis which may possibly throw some new light on the controversial and continually disputed nature of this illness.

Introduction

Hyperostosis frontalis interna – etiology and pathogenesis

Paleopathological studies are important for the assessment of both the biological condition of particular individuals and the quality of life of human populations in

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doi:10.1016/j.jchb.2005.08.001

the past. They provide us with a vast body of information on the history and development of particular diseases in both time and space.

During menopause in women, on the inner surface of the frontal bone there frequently occurs pachycephaly in the form of prominent bulges, separated by grooves of different depths. These features, located symmetrically on both sides of the frontal crest, do not reach beyond the coronal suture.

The bulges are present in the inner lamina and occasionally in the diploë, while the thickness of the external lamina remains unchanged. Occasionally there also occur calcifications in the falx cerebri and along the sagittal suture (Bidziński, 1988).

Hyperostosis frontalis interna (HFI) is classified into a broad category of hyperostoses termed either hyperostosis cranii diffusa (HCD), in which there appears a uniform thickening of all cranial vault bones (Murczyński, 1952), or hyperostosis calvariae interna (HCI), in which osteal bulges overlap parietal bones (Anton, 1997). Osteal bulges may also be encountered in bones of one half of the skull only, this condition being called hemicraniosis (Murczyński, 1952). All the above-mentioned illnesses comprise a large number of pathological states termed hyperostosis cranii (HC) (Larsen, 1997).

From the anatomical viewpoint, degenerative osteal hypertrophies in part of the skull do not show histologic changes and maintain a regular system of Haversian canals and laminar spaces (Anton, 1997). Such changes were first described over 250 years ago by pathologist Giovanni Batistta Morgagni, a Padua University professor of anatomy (Morgagni, 1719). In the course of his studies he observed the co-occurrence of HFI-type changes with obesity and excessive hair growth (hirsutism, virilism). More than 200 years later, Stewart (1928), Morel (1929) and Moore (1955) observed that HFI individuals showed – besides obesity and hirsutism – also neuropsychiatric symptoms and a persistent headache. The co-occurrence of all these symptoms, linked by the above-mentioned authors with hormonal disorders, is described in the literature as the Morgagni–Stewart–Morel–Moore (MSMM) syndrome (Anton, 1997).

The clinical symptoms of the MSMM syndrome are characterized by headaches, psychic disorders, dysopia, female hirsutism related to lesions of tuber cinereum and infundibulum and obesity (Rożniatowski, 1981; Murczyński, 1952; Anton, 1997). Given that these symptoms are most frequently observed in postmenopausal women, they should be ascribed to menopausal vegetative disorders. The origin of these changes is often linked with hypophyseal disturbances, since an increased number of acido- and basophilic cells and the occurrence of small adenomas in the anterior lobe of the pituitary gland have been reported (Henschen, 1949). On the other hand, Gegick et al. (1973) observed an elevated level of alkaline phosphatase in the plasma of individuals with the MSMM syndrome. Rühli and Henneberg (2002) have shown hormonal influence on this phenomenon. In the etiology of the illness, the role of hypophysis-diencephalon system disorders is often stressed (Rożniatowski, 1981; Gładykowska-Rzeczycka, 1990). However, it has not been determined so far to what extent the osteal bulges that manifest themselves as HFI determine the occurrence of headaches and psychic disorders. The pressure of the expanded inner lamina of the frontal bone on frontal lobes does not seem to play any direct role in the