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CASE REPORT

Hyperostosis frontalis interna: An Egyptian case referred to the second dynasty (2890–2650 BC) from Tarkhan-Egypt

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Abstract  Aim of the work and case presentation: This article is to present the evidence of hyperostosis frontalis interna (HFI) in a young adult male, approximately (25–26) years old, recovered from a second dynasty site (2890–2650 BC) at Tarkhan in the south of Dahshur-Egypt. HFI is rare in historic populations. It is most commonly found among females with prolonged estrogen stimulation. Males with hormonal disturbances like primary hypogonadism were found to manifest with HFI.

Conclusion: This study is a further contribution to the case history of HFI in osteoarcheological studies. Further researches are recommended to have an overview on such an old disease in Egypt.

1. Introduction

Hyperostosis frontalis interna (HFI) is a generalized pathological condition with an unknown etiology and variable clinical association. It is characterized by excess bone growth and manifests on the inner table of the frontal bone, occasionally extending onto the temporals, parietals and the occipital. It is seldom found in males, but it is a common phenomenon among post-menopausal females in modern societies [1]. The etiology of HFI is uncertain: it may be due to an unknown genetic predisposition, a common environmental exposure, or special metabolic diseases [2]. Endocrine disturbances, aging, and dietary factors are also listed among the causes of HFI. ‘Morgagni–Stewart–Morel–Lavallée syndrome’ was first described in an obese female patient during autopsy that had hirsutism and thickening of inner table of a skull [3]. HFI may be accompanied by myxoedema [4], acromegaly or prolactinemia [5,6]. Sometimes it is associated with headache and some neuropsychiatric diseases such as epilepsy and dementia [7]. Its association with other pathologic conditions like giant cell arteritis [8] and Klinefelter’s syndrome has been reported [9].
In clinical medicine, HFI is diagnosed from its X-ray appearance, whereas in palaeopathology it is diagnosed by a direct observation of the skull [10].

The association of HFI in a male accompanied by primary hypogonadism has been reported [4]. The male fraction increasing prevalence in the old pre-Christian samples in comparison to the modern samples suggested a hormonal basis for microevolutionary trends in HFI [11].

The aim of the present paper is to present the oldest known Egyptian male individual with HFI who is referred to the second dynasty (2890–2650 BC).

2. Case presentation

A skeleton of an adult individual was found at a second dynasty site (2890–2650 BC) of 288 burials at Tarkhan in the south of Dahshur. Dahshur is a royal necropolis located in the desert on the west bank of the Nile approximately 40 km south of Cairo. This burial was located at the south side of the location, 1.5 m below the earth surface.

The skeleton was found in a squatting position, on its left side, and directed from north to south. Remnants of linen wrapping were found around some parts of the body which was kept in a mat basket (Fig. 1). The condition of the skeleton was not good as the surrounding area was invaded by agriculture.

The male sex was diagnosed by Phenice’s technique for sex determination [12].

The individual was a young adult, approximately (25–26) years old at the time of death. This was identified by using the Todd pubic symphysis scoring system [13].

The height of the individual was calculated to be between 164.80 ± 4.57 and 167.38 ± 4.66 cm, by using the length of humerus and radius (30 and 24 cm respectively), by applying the stature formulae of humerus and radius for male sex settled by Trotter and Gleser, 1958 [14].

The skeleton was investigated by conventional radiology. No invasive investigation was used.

The skeleton was robust showing diffuse irregular bilateral bone overgrowth at the sites of muscle attachments. The endocranial surface of the frontal bone, medial parts of the parietal bones and occipital bone show irregular thickening with a maximum thickness of 8 mm which was measured along the coronal suture 4.5 cm medial to the left temporal line (Fig. 2). The lateral parts of the parietal bones and the temporal bones were unaffected and showed taphonomic changes. The continuous bony overgrowth involved more than 50% of the frontal endocranial surface and the irregular elevation of the entire region with sharp clearly demarcated borders classified this case as HFI type D according to the classification of Hershkovitz et al., 1999 [15].

X-raying of the frontal bone revealed diffuse thickening of bone with increased opacity (Fig. 3).

3. Discussion

Possible differential diagnosis of Tarkhan male includes Paget’s disease which is a common condition characterized by increased and disorganized bone turnover which can affect one or several bones throughout the skeleton. HFI has a frequency of 8% in Paget’s disease [16]. Paget’s disease disrupts normal bone architecture, and spacing is sometimes present between the teeth due to the increase in the size of the jaws [17]. Paget’s disease in Tarkhan male was excluded by the different age group and the different macroscopic appearance of the skull.

Multiple osteoid osteomas almost affect the flat bones of the skull and face [18]. The thickening of the bone in Tarkhan male was not localized and it was homogenous. No signs of meningioma like reactive hyperostosis, intraosseous meningioma or sclerotic metastasis were detected. The possibility of fibrous dysplasia was excluded by the macroscopic picture of the medullary cavity.

CT scan was suggested as a radiological tool to provide enough information to distinguish HFI from other bony overgrowths e.g. osteomas, ossified hemangioma, etc. [19].

Diffuse idiopathic skeletal hyperostosis (DISH) is a condition which is characterized by the presence of new bone formation. HFI may be seen in these cases [20]. Harris et al. (1974) detected one patient with HFI out of 34 patients with DISH, with no evidence of diabetes mellitus or acromegaly [21]. The
association of HFI, gout and DISH was reported in the paleopathological study made by Fornaciari et al. (2009) [22]. They studied the remains of the famous Medici family of Renaissance Florence that suffered from an arthritis disease. These three abnormalities were found in Cosimo’s remains. The possibility of DISH was excluded in this skeleton by the absence of the calcification and ossification of spinal ligaments and the entheses.

Ciocci et al. (1985) performed a routine roentgenographic examination of the spine and the skull of 690 patients with rheumatologic disorders. They reported HFI in 63% of cases, more common in females, with no common pathogenic mechanism [23].

HFI was detected in 87% of 30 acromegalic patients in a study made by Littlejohn et al. (1986) [24]. Elevated somatomedin-C (insulin-like growth factor 1) levels were suggested to be responsible for both the generalized and the localized skeletal overgrowth. The strong association of moderate and severe HFI with acromegaly and with hyperprolactinemia suggests that HFI may be a further marker of pituitary dysfunction [25]. Although the sella turcica was not found due to the bad condition of the skeleton, acromegaly was excluded by the absence of prognathism or enlargement of the mandible and the absence of gaping teeth.

HFI was reported to occur in Klinefelter’s syndrome [26], but the gross features of Tarkhan male excluded this possibility.

Morgagni–Stewart–Morel syndrome is a disease of abnormal bone growth or osteochondrodysplasia which is characterized by the thickening of the frontal bone of the skull and

Figure 2  Thickening of the endocranial surface of the frontal bone along the coronal suture medial to the left temporal line.

Figure 3  X-raying of the frontal bone revealed diffuse thickening of bone with increased opacity.
crown, obesity, impotence in males and masculinization, menstrual disturbances and amenorrhea in females. Its cause is unknown [27]. Morgagni–Stewart–Morel syndrome has been reported in a 32-year-old man with increased serum concentrations of prolactin and thyrotrrophic hormone [28].

HFI was also described in association with toxic goiter and diabetes mellitus [29]. It was thought to be caused by endocrine disturbance of the pituitary gland [4].

HFI is not a purely female phenomenon as males with hormonal disturbances such as primary hypogonadism were found to manifest it, especially type D. Such individuals often suffer from primary or secondary testicular atrophy [4].

It has been suggested that functional disturbance of the gonads, i.e., faulty estrogen stimulation of or abnormal progesterone effect on the ovaries, or inadequate androgen stimulation by the testes, are the main causes of HFI. Calame (1951) [30] claimed the symptoms of HFI to be the same as those of infundibulo-pituitary disturbance (e.g., adiposity, genital dystrophy, disturbance of sugar metabolism). He placed particular emphasis on gonadal factors and noted that male patients with HFI were commonly feminized, with atrophic testes. Perou (1964) [31] found that 5 out of 6 HFI cases, among males, presented with testicular under development or atrophy. Males probably developed HFI type D under extreme conditions of hormonal imbalance such as atrophied testes [15].

The study of the skeleton of the famous singer Farinelli (1705–1782) who was castrated before puberty showed prominent HFI affecting his frontal bone. Androgen deficiency was accused for causing this abnormality [32].

HFI may occur as an independent condition. It was considered as a phenomenon distinct from hyperostosis cranialis diffusa (HCD) and other endosteoses [15].

The association of headache with HFI is well established in the Morgagni–Stewart–Morel syndrome. It is presumed to be due to bone overgrowth of the inner table of the skull. The persistence of headache after effective treatment of pituitary adenoma may be due to the presence of HFI [25].

Hyperostosis frontalis interna (HFI), first described in 1765 by Santorini and Morgagni, has been defined as “a disorder of the endocranial plate which remodels into a more cancellous phenotype” [25].

Two skeletons of old individual dating to early middle ages from necropolis of Vicenne-Campochiaro (Italy) showed advanced HFI [33].

The frequency of HFI increases with age. It appears in women after the age of 40. It was believed to be associated with prolonged estrogen stimulation. It is much less frequent in females under 40 years of age. However, advanced cases of HFI (types C and D) have been observed in individuals as young as 40 years of age [15].

HFI is fairly common in modern populations, affecting 5–12% of the general population [7]. It has rarely been documented in the archaeological record [34], but with a much lower frequency than in modern groups [35].

However, HFI shows its increased prevalence and change in sex ratio at the start of the industrial revolution in the mid 18th century AD [15]. Hershkovitz et al. (1999) examined approximately 2000 skulls from various geographic locations and ethnic groups, extending from the 4th millennium B.C. to the 7th century A.D., as well as a large group of European skulls prior to the 19th century. None of these skulls displayed HFI. On the other hand, 24% of female skulls and 5% of male skulls showed HFI in a group of 1706 skulls from the 20th century. They classified HFI into types A–D, based on the involvement of other bones and the extent, appearance, border type, shape, and location of the lesions.

HFI prevalence has increased during the last century, especially among young individuals, possibly indicating a profound change in human fertility patterns, together with the introduction of various hormonal treatments. Prolonged estrogen stimulation is related to HFI [36].

Rahli and Henneberg (2002) [37] suggested that during human evolution, a wider availability of food favored an increased metabolic rate and increased leptin levels, which is a hormone that signals the feeling of satiety to the hypothalamus and helps to control the metabolic rate. Serum leptin levels are correlated with body mass index and may have caused a higher incidence of HFI.

HFI has been clearly identified in Homo erectus and Neandertalians [38]. It was also observed as bone nodules on the endocranial surface of the frontal bone of two elderly females coming from the excavation of the Neolithic hypoguem of Boilleau (France) [39].

The earliest European male with HFI in the historic era was referred to the late late period (150–80 BC) at Basel-Gasfabrik in Northwestern Switzerland [11].

To our knowledge, the earliest example of HFI in the historic era was reported in a 40-year-old female recovered from a meroitic cemetery (ca. 300 AD) in Sudanese Nubia [40]. The authors suggested the changes in the skull fragment consistent with the diagnosis of Morgagni–Stewart–Morel (MSM) syndrome.

The young adult male from Tarkhan-Egypt with HFI presented in the recent work is referred to the second dynasty (2890–2650 BC), and raises the pre-Christian reported male HFI percentage, which was considered as 43% of the reported HFI cases and was notably different from the recent male modern prevalence of 0.3% [11].

Presented in this article, to our knowledge and till now, is the oldest known Egyptian male with HFI referred to the second dynasty (2890–2650 BC). This report contributes to the case history of HFI in osteoarcheological material, and is added to the male individuals with HFI in the pre-Christian historic era.

Conflict of interest

The authors declare no conflict of interest.

References

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