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MORPHOLOGY OF A 19TH CENTURY ADULT MALE SKULL WITH NON-TREATED HYDROCEPHALUS — A CASE REPORT

Abstract: This report presents the craniofacial morphology of a skull afflicted with hydrocephalus, belonging to an adult male who lived in the 19th century in Vienna. The volume of the skull (2022 cm^3) exceeds normal capacity of a male skull which is estimated to be 1500 cm^3. Diameters of the neurocranium and head circumference of the specimen differ significantly from normative values, while the facial diameters remain in normal range of variation. Characteristic features of the investigated skull are numerous accessory bones and sutures of the neurocranium. Overall the morphology of the cranial bones suggests that the male suffered from congenital hydrocephalus.

Key words: cranial development; craniofacial morphology; hydrocephalus; skull.

INTRODUCTION

Hydrocephalus is the pathological enlargement of the cerebral ventricles resulting from disturbances in the production and absorption of the cerebrospinal fluid (CSF). This causes the volume of CSF to increases in the ventricular system which results in increased intracranial pressure inside the skull and progressive enlargement of the head [1]. Hydrocephalus may be congenital or acquired. The incidence of congenital hydrocephalus is estimated to be 3 per every 1,000 live births. Hydrocephalus in adults represents approximately 40% of all cases. Frequency of acquired hydrocephalus is difficult to estimate because of a variety of disorders which can cause it [1]. Infections, overgrowth of the choroid plexus and embryonic neoplasms which may occur during foetal life can induce hydrocephalus. Acquired hydrocephalus is commonly an after-effect of inflammation or subarachnoid haemorrhage caused by different factors. In turn, adult hydrocephalus is often related to neoplastic or inflammatory diseases of the central nervous system [2].

The fundamental symptom of hydrocephalus during childhood is the enlargement of the head. In extreme cases the circumference of the head may increase to 60 or even 70 cm. Radiographic studies show symptoms of increased intracranial pressure, widening of the sutures and deepening of the digital impressions. When hydrocephalus occurs in the period when the cranial sutures are closed, the volume of the head cannot enlarge. This causes the compensation mechanisms to fail. Thus, the symptoms of brain compression and increased intracranial pressure appear rapidly. In cases of long-lasting hydrocephalus compression and atrophy of the brain can occur, with intellectual development remaining at a normal level for a long time. However finally hydrocephalus leads to mental retardation [3].

The aim of this case report is to present the craniofacial morphology of an adult male who lived in the 19^{th} century, and was afflicted with hydrocephalus. From an anatomical and anthropological point of view the presented skull is a rare case. Therefore it was subjected to morphometrical analysis. This revealed to what extent craniofacial morphology can be altered by abnormal biodynamical conditions dictated by disturbances in CSF outflow.

MATERIALS AND METHODS

The investigated male skull showing signs of hydrocephalus comes from to the cranial collection of the Department of Anatomy, Jagiellonian University Medical College (Krakow, Poland). The skull is registered in the museum inventory book and dates back to 1874. There is only a brief information that it was a skull of a 40-year-old shoemaker from Vienna.

The skull is well preserved. However, it lacks the mandible, a few sutural bones and most of the teeth (Fig. 1). Marked bone muscular attachments, a well developed external occipital protuberance, a big mastoid processes, prominent supercilliary ridges and a protruded glabella indicate a male skull.

To study the craniofacial morphology of the hydrocephalic skull, measurements were taken with the use of a spreading calliper and flexible tape, appropriately to the established anthropological methodology [4]. The volume of the skull (V) was estimated using the following formula [5]:

$$V = 3.658 \cdot C + 4.577 \cdot A - 1930$$

where:

V — skull volume,

C — horizontal circumference of the skull (fronto-occipital circumference,

A — length of the transverse cranial arch (measured from the right porion via bregma to the left porion).

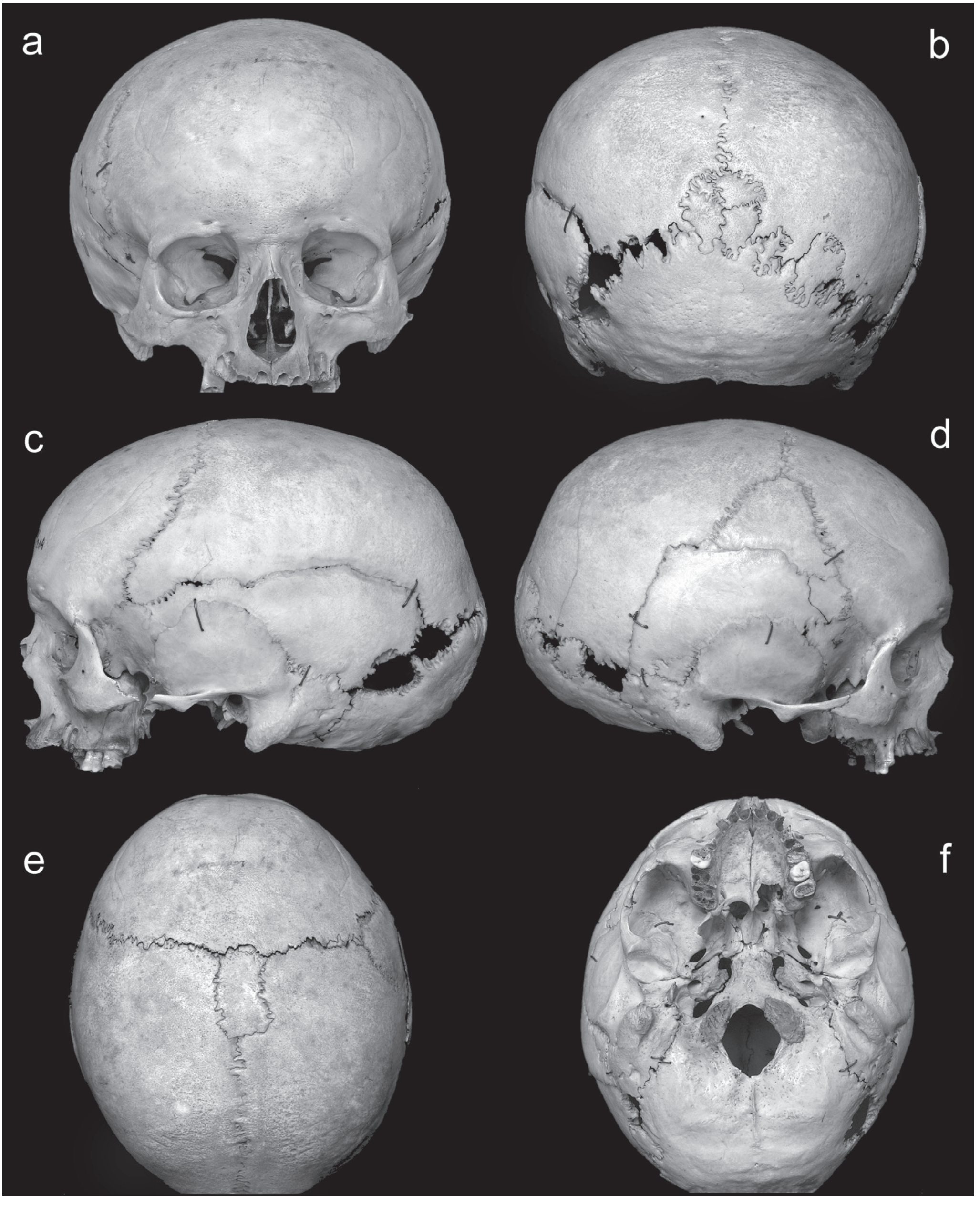

Fig. 1. Frontal (a), occipital (b), lateral (c, d), superior (e), and inferior (f) view of the hydrocephalic skull.

RESULTS

The main craniofacial measurements of the hydrocephalic skull together with appropiate normative values are presented in Table 1. Measurements performed have shown a marked increase of the following parameters — cranial circumference, cranial length and width, transverse cranial arch, width of the forehead and the occiput. The volume of the hydrocephalic skull was calculated to be 2022 cm^3.

Table 1

Summary of craniometric data of the hydroecphalic skull (x), and normative values.

Measurement (in millimeters)	x	x_n	sd	min	max
Cranial circumference	605	518.6	14.7	480	550
Transverse cranial arc length (po-b-po)	380	336.6	12.3	310	365
Length of the skull (gl-op)	214	177.1	6.4	162	194
Height of the skull (ba-b)	143	133.4	7.2	118	146
Width of the skull (eu-eu)	176	147.2	4.9	137	158
Cranial base length (n-ba)	107	98.4	5.9	86	110
Width of the forehead (ft-ft)	104	98.3	4.7	86	108
Occipital width (ast-ast)	120	110.7	5.1	97	120
Width of the face (zy-zy)	131	133.3	5.5	122	146
Length of the face (ba-pr)	100	93.8	6.2	82	108
Height of the face (n-pr)	60	67.4	3.8	58	74

xn — mean value; sd — standard deviation; range of variation: min-max values; po — porion; b — bregma; gl — glabella; op — opisthocranion; ba — basion; eu — eurion; n — nasion; ft — frontotemporale; ast — asterion; zy — zygion; pr — prosthion.

In the case of the studied skull, diameters of both the splanchnocranium and basicranium are within normal biological range of variation. Contrary, the investigated skull shows considerably altered diameters of the calvarial bones (Tab. 2). This refers particularly to the parietal chord (141 mm) and the parietal arc (160 mm) which are extremely long.

The outstanding morphological character of the calvarium was found to be caused by the abnormal size of the bones, as well as their number. We found an accessory suture, which divided the left parietal bone into two parts. This suture ran almost horizontally from the sphenoparietal suture towards the lambdoid suture. On the right side, the number and organisation of the bones was also found to be uncommon. There are three accessory bones between the parietal, frontal and temporal bones, connected by non-obliterated sutures. The first is

Table 2

Measurements of the calvaria of the hydrocephalus (x), and normative values (xn).

Measurement (in millimeters)	Chord distance					Arc length				
	x	x_n	sd	min	max	x	x_n	sd	min	max
Frontal bone (n-b)	120	110,8	4,2	102	121	140	126,1	5,8	113	142
Parietal bone (b-l)	141	110,9	7,6	95	131	160	124,3	9,8	102	148
Occipital bone (l-op)	100	93,6	5,5	81	107	125	114,6	7,2	98	130

a big triangular bone (50 × 55 × 65 mm). The second accessory bone is wide and sickle shaped. The third accessory bone is small and located between the two mentioned bones, the frontal bone and the great wing of the sphenoid bone. Moreover, a single, large bone (bregmatic bone) is embedded in the region of the bregma (junction between the sagittal and coronal sutures). Numerous big sutural bones were present within the lambdoid suture.

DISCUSSION

The aim of this case report is to presents the craniofacial morphology of an adult male who lived in the 19th century and suffered from hydrocephalus.

Measurements of the hydrocephalic skull have shown excessive growth in most of the cranial diameters when comparing with a normal male skull [6]. A typical feature of hydrocephalus is the considerable expansion of the neurocranium, while the facial morphology remains unaltered [7]. The smallest width of the forehead of the investigated hydrocephalic skull is within normal variation range of this feature, while the largest width of the forehead (145 mm) significantly exceeds the normative value. The widening of the frontal bone in the horizontal plane was probably facilitated by the growth along the coronal suture, rather than towards the antero-posterior direction, as this would have been associated with the bossing of the forehead which was not observed in the skull. In turn the presence of enlarged parietal bones was certainly associated with increased osteogenesis and activity of the sagittal suture [8]. The function of the suture was stimulated by an extensive growth of the brain. Presumably the pressure exerted by the brain onto the posterior part of the skull caused protrusion and backward bending of the occiput. This could also stimulate the process of new bone formation within the lambdoid suture [9–11]. Although this suture may contain accessory bones in healthy individuals as the epigenetic features, the sutural bones of the studied skull derive apparently from pathological cranial development dictated by hydrocephalus. The final result was that cranial dimension enlarged, adjusting to new bone morphology, which allowed for complete brain tissue protection.

Basing only on skull morphology we are not able to define the etiopathology of the studied hydrocephalic case [12]. We can only hypothesize that observed cranial dysmorphology was caused by:

Congenital hydrocephalus, which could have appeared during the period of prenatal or early postnatal development.

Pathological growth of the skull that resulted from acquired hydrocephalus. This might have been caused by perinatal injury (i.e. haemorrhage to the brain ventricles followed by formation of a hematoma) leading to disturbances in the circulation of CSF.

The first hypothesis is supported by cranial morphology of the examined case. Altered morphology of the selected cranial bones, presence of numerous sutural bones and additional sutures suggest that the hydrocephalus origins from early childhood. Contrary, a hydrocephalus in adults can induce only minimal osseous variations. The investigated skull is an example of orchestrated growth between the cranial bones and the brain, and demonstrates plasticity and adaptability to abnormal physiological conditions. As it was stated before, the hydrocephalic skull provided sufficient protection for the expanding brain. The fact that the sutures of the cranial vault remained open and widened is of importance. They could serve as the sites of growth for the cranial bones and provided space for the bone to slightly move [13].

The morphological appearance of the studied hydrocephalic skull signifies that during its development specific factors must have acted to stimulate its growth in an abnormal way [14]. These factors provoked the formation of additional bones that might have played a compensatory role in the abnormal growth of the neurocranium. Certainly, development of the cranial vault of the examined hydrocephalic skull was disturbed and accelerated more than in normal growth of the head. However the pattern of the cranial sutures in the investigated specimen is not altered as in the cases of different skull deformities [15]. The cranial sutures do not show extensive complication or widening as it is commonly observed in hydrocephalus. The presence and osseous activity of accessory sutures and supernumerary bones could efficiently compensate the extensive enlargement of the brain and thus the cranial vault could completely envelop the brain. In the studied skull, the enormous expansion of the brain influenced mainly the size and shape of the upper part of the neurocraniu. The cranial base and facial skeleton were not considerably altered [10, 16, 17]. As it was stated by Scott, the cranial base is not directly correlated with growth of the brain and larger skulls tend to have larger cranial bases [18]. The length of our specimens basicranium did not differ significantly from the normal range. Also the posterior part of the cranial base (the occipital width) remained within the normal range of variation. This cranial region is closely related with the growth of the cerebellum [13]. Thus, we presume that overgrowth of the neurocranium did not affect the size of the basicranium. However, larger diameters of our specimens basicranium might be secondary to increased intracranial pressure.

It seems interesting that in the 19th century an individual with hydrocephalus survived until the age of 40. It should be stressed that in that time the therapeutic possibilities were limited [19] and foetal hydrocephalus was lethal. When hydrocephalus occurs during prenatal or early postnatal development, it significantly changes the anatomy of the skull.

The relationship between craniofacial deformity, elevated intracranial pressure, and mental retardation remains controversial [15]. Therefore, we presume that in the case of the individual whose skull we have described could have preserved appropriate mental functions and was able to work as a shoemaker, as it was stated in the inventory book. Hence, if we accept the theory that the male suffered from congenital hydrocephalus, we can safely say that the skull and the brain developed in anatomical accordance. The skull did not pose any restrictions for the growing brain, therefore the male was able to survive till adulthood without surgical treatment.

CONFLICT OF INTEREST

The authors have no conflict of interest or financial relationship to disclose.

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