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Anthropological characteristics of a case of microcephaly

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The article describes a female skull from the contemporary cementary of Pestkovo, Bulgaria, with morphological traits of microcephaly (skull capacity — 907 cm^3). The skull was characterised by means of measurements with reference to 60 skulls of contemporary Bulgarians. Thus the normalised data and values of natural Perkal's indices were obtained.

Obtained data indicate morphological differences of skull with microcephaly compared with the normal ones. In general, the analysed skull is of a smaller size although the values of its height (porion-bregma), total facial height (nasion-gnathion) and palatial length (orale-staphylion) are bigger than in the control group.

key words: skull, microcephaly, oligophrenia

INTRODUCTION

Authors describe the skull of a young male who deceased between 1957 and 1960. The analysed individual was a patient of a social centre for people with mental disorders (i.e. schizophrenia, oligophrenia, epilepsy and general arteriosclerosis) in a Bulgarian village — Pestkovo.

According to the information obtained from the employees of the centre, the patient suffered from oligophrenia. The morphological traits of the skull, especially its small capacity — 907 cm^3, indicate a typical case of microcephaly.

In the analysed osseous material from different ancient and contemporary burial grounds localised in Poland no case of microcephaly was found. This can be explained not only by the rare presence of the described pathology in the osseous material but also by the fact that only a small number of individuals suffering from the microcephaly reaches adult age. For these reasons every skull of an adult with microcephaly should be thoroughly analysed from the morphological perspective [4].

While recognising microcephaly most attention is paid to the small dimensions of the craniocerebral part of the skull, especially its capacity and its horizontal circumference. Also the disproportion between the level of development of the cerebral and facial parts of the cranium is emphasised [4].

Among the direct causes of microcephaly the most common are the following: serologic incompatibility, roentgenoscopy of a woman in the early phase of pregnancy, developmental disturbances of an embryo, alcoholism or syphilis of the parents, foetal infection, intrauterine compression and hormonal disturbances (for example hypophyseal microsomia). A certain role is played also by the heredity factor that rates for the new-borns between 1:25 000 and 1:50 000 [4].

All the mentioned factors can initiate the process of brain development restraint. Considering the above many scientists suggest that microcephaly is the consequence of the adaptation of the craniocerebrum to cerebral hypoplasia [1,12].

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There are many different proposals concerning classification of different forms of microcephaly. For instance the pathology can be classified into four groups: primary, secondary, craniostenosis and craniosynostosis [3], or into two basic groups — physiological microcephaly and pathological microcephaly. With reference to pathological microcephaly the following causes can be stated: craniostenosis, heredity, pre- and even postnatal acquired microcephaly [10]. According to Horácková [4], the most accurate classification was made by Koch [7], who differentiated two types of microcephaly: acquired (85% of all the cases) and inborn (about 15% of the cases).

Due to scarce data about the origin of the analysed skull we have concentrated on the detailed morphological characteristics without attempting to qualify the skull to any of the described groups.

MATERIAL AND METHODS

The information about the origin of the analysed skull is very incomplete. As far as we know it belonged to a male with recognised oligophrenia — a patient of the social centre in the Bulgarian village of Pestkovo. The individual died in early age (adultus), between 1957 and 1960 and was buried in the cemetery nearby, where his skull was found.

All the measurements of the skull were made according to Martin and Saller's code [11]. Since there is a big probability that the individual was of Bulgarian origin, 60 skulls of contemporary Bulgarians were used as a comparative set. The skulls come from the exhumation of the skeletons of partisans who died between 1941 and 1944 [8]. The craniometric characteristics of the skull with microcephaly and the control set are put together in Table 1.

In order to achieve the format of data which enables the direct qualification of rate and direction of analysed skull traits' variations (x_a), the measurements were normalised (x'_a) according to the following formula:

$$x' = (x_a - X) : SD$$

More detailed information about which of the analysed traits stray from the standard, in ratio to the total size of the skull with microcephaly, were achieved by applying the so-called Perkal's natural index [14]. First of all the size of the skull was measured according to the formula:

$$m = \sum_{I=1}^{K} x_a : K$$

where $\sum x_a$ equals the sum of normalised skull traits' values, K — the number of traits taken into consideration. Then the natural index (W) for each trait was measured. The index is a result of subtraction of concrete normalised measurement and the general size of the skull (m):

$$W = x'_a - m$$

After having completed the measurements described above we attempted to recreate the capacity of the skull (P), taking into consideration the importance of this trait in the case of microcephaly. We applied Pearson's method, quoted by Malinowski et al. [9]. For this reason the following equation relating to male skulls was applied:

$$P = 0.000337 \text{ (g-op} - 11) \text{ x (eu-eu} - 11) \text{ x}$$
$$\text{x (ba-b} - 11) + 406.01$$

RESULTS

Metrical Features

Table I presents metrical and normalised characteristics of the analysed skull and the Perkal's natural index counted with reference to the control set. The data are also illustrated in Fig. 1.

Based on 29 features used in the comparison, a total size of the skull — m = – 2,27 was defined. According to the result the size of the skull is considerably smaller than the average size of skulls from that time.

25 normalised features (x'_a) — that is about 86 % of the features of the described skull presented lower values than the control set.

There are fundamental differences between the control group and the analysed skull. These differences were especially evident while analysing the craniocerebral measurements — 10 of 15 features strayed from the standard deviations more than –3. Among the measured traits that achieved the lowest phase of development we can name the following: skull length (g-op), parietal bones length (b-l), height measured from the basis (ba-b) and the smallest width of the forehead (ft-ft). The inside proportions measured by the natural indices also show the underdevelopment of the described parts of the skull (Table 1, Fig. 1).

The situation differs while considering the morphology of the bony face. First of all the differences with comparison to the control set are smaller, some values of normalised features achieve even positive results (f.i. gn-id).

The differences in development of the bony face and the craniocerebral part of the skull are more

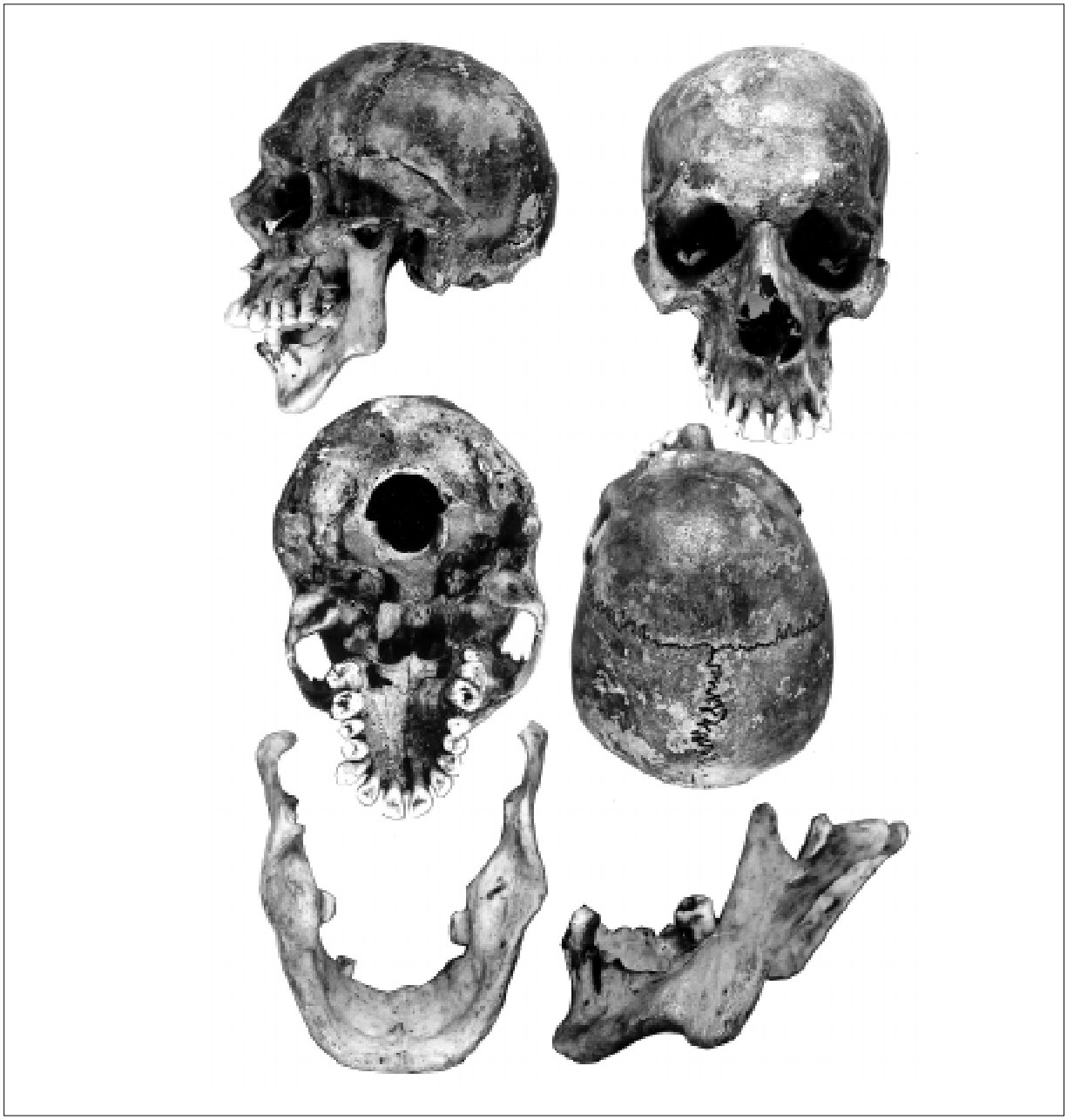

Figure 1. Skull with microcephaly in lateral, frontal, basilar and vertical views as well as the inferior and lateral views of the mandibula.

accentuated by Perkal's indices (Table 1, Fig. 2). Analysis of the indices shows that 8 from 15 linear elements taken into consideration during the research on the general skull size reached higher (positive) values than expected. The highest values of the natural indices were obtained from: facial length (n-gn), nasal length (n-ns), piriform aperture width (apt-apt), upper jaw length (ol-sta) and mandible body height (gn-id).

Table 2 presents the measurements complementary to the skull characteristics, the equivalents of which we have not found in accessible literature, together with the quotient indices which describe the proportions of the skull. According to the classification of Malinowski and Wolański [9] the analysed skull can be described as long (dolichocephalus), moderately high (orthokranius), strongly arched (akrokranius) with very narrow face (hyperleptoprosopus), very high cheek (hyperlepten), very wide mandible, moderately high orbital cavities (mesokonch), moderately wide nose (mesorrhinus) and narrow forehead. It is worth emphasising that only two in-

Table 1. Statistical characteristics of the analysed skull and the control group

Feature and number according to Martin	Control set		Skull of the analysed individual		
	X	SD	x_a	$x'_a = (x_a - X) : SD$	W
1. g-op	182.3	6.5	154	–4.35	–2.08
7. ba-o	38.0	2.7	31	–2.59	–0.32
8. eu–eu	140.1	6.6	114	–3.95	–1.68
9. ft-ft	96.5	4.0	79	–4.38	–2.11
10. co-co	119.7	6.4	99	–3.23	–0.96
11. au-au	117.6	5.8	97	–3.55	–1.28
12. ast-ast	110.1	4.3	92	–4.21	–1.94
13. ms-ms	102.9	4.8	91	–2.48	–0.21
16. szer. F. m.	32.3	2.1	31	–0.62	+1.65
17. ba-b	136.8	5.6	112	–4.43	–2.16
20. po-b	118.1	4.6	122	+0.85	+3.12
29 n-b	112.9	5.0	96	+3.38	–1.11
30. b-l	112.8	5.4	82	–5.70	–3.43
31. i-o	47.8	5.4	32	–2.93	–0.66
44. ek-ek	95.9	2.7	84	–4.41	–2.14
45. zy-zy	129.0	4.9	109	–4.08	–1.81
46. zm-zm	91.6	4.2	78	–3.24	–0.97
47. n-gn	120.0	6.4	122	+0.31	+2.58
48. n-pr	70.3	4.5	70	–0.07	+2.20
50. mf-mf	21.1	2.3	17	–1.78	0.49
51. mf-ek	43.0	2.3	42	–0.43	1.84
52. orbit. height	34.4	1.8	32	–1.33	+0.94
54. apt-apt	25.2	2.0	25	–0.10	+2.17
55. n-ns	53.3	2.9	53	–0.10	+2.17
61. ekm-ekm	61.4	4.0	51	–2.60	–0.33
62. ol-sta	43.5	3.3	45	0.45	+2.72
63. enm-enm	39.1	3.4	30	–2.68	–0.41
66. go-go	98.6	6.1	83	–2.56	–0.29
69. gn-id	32.5	2.5	37	+1.80	+4.07
			Total: –65.77 K = 29 m= –2.27		

X — arithmetic mean of the control group,
SD — standard deviations,
x_a — values of the analysed skull measurements,
x'_a — normalised values of the measurements,
W — natural Perkal indices. Measurements in mm. Control group according to the data by Malinowski et al. [8],
N = 60 individuals

dices i.e. morphological facial index and zygomatico — mandibular index reached extreme values while the other ones can be placed within the limits of the normal variability of features.

Cranioscopy features

Taking into consideration the most complete description of microcephalic skull morphological properties the cranioscopic features were also analysed. As a comparative sample the unpublished cranioscopic tables of Michalski [9], modified by Wierciński, were used.

Basic skull sutures: coronal suture, interparietal suture and occipitoparietal suture are free, abrasion of mastication teeth surface is only lightly marked. Vertical skull outline is ovoides obtusus (Fig. 1), the forehead of rounded shape and biconvex profile is rather poorly inclined. The supraorbital arches are strongly shaped and mastoid bones are big and extended (Fig. 1).

Facial part of the skull shows weakly formed prognathism — the angle between the Frankfort horizontal plane and the line connecting nasion-prostion points is about 100°. As the mandible is retreated one can have the impression that the prognathism of the skull is much stronger than it is in reality. Strong left-sided asymmetry of the maxillary bones, piriform aperture, and the basal part of the occipital bone were observed (Fig. 1). Incisura maxillaris is very deep, the root of the nose is high, nasal bones protrude poorly with slightly flexuous-concave profile. Spina nasalis is poorly lowered and protrudes strongly.

Mandibular arch shape is broadly trapezoid, mentum protrudes extremely strongly.

Analysing the above description together with the photographs added to the documentation the following anomalies and pathologies attract special attention: first of all a very strong distoclusion is marked together with partial apertognathia (occlusioaperta partialis seu infraoclusio) — (Fig. 1), as well as the reduction of third molar (M_3) (Fig. 1). Also two other features were observed: a hypertrophy of bony tissue around the angle of the mandible (Fig. 1) and extremely protruding mentum, which makes the mandible resemble the type observed in old people (Fig. 2).

DISCUSSION

Professional literature concerning studies of skulls with microcephaly draws attention to the particular form of certain morphological traits — typical for such an anomaly. Apart from small skull capacity (less than 1100 cm^3) pithecoidal morphology of the skull is most often accentuated. This is caused by simultaneous as-

Table 2. Quotient measurements and indices completing the morphological characteristics

Feature and number according to Martin	Measures (x_a)	Feature type	Measure or index		
5. n-ba	93	**angles:** mandible branch		127°	
ba-l	91	mandible opening		87°	
Po-v	92	mentum protrusion		60°	
l-i	59	**quotient indices:**	x_a	x_b	X
40. ba-pr	90	eu-eu/g-op x 100	74.0	77.0	77.0
42. ba-gn	97	ba-b/g-op x 100	72.7	78.7	73.9
60. pr-al	60	ba-b/eu-eu x 100	98.2	102.1	97.8
65. kdl-kdl	94	n-gn/zy-zy x 100	111.9	92.0	93.0
68. go-gn	68	n-pr/zy-zy x 100	64.2	60.0	54.5
68 (1). go-kdl	65	go-go/zy-zy x 100	92.2	78.0	76.5
Arches: i-o	35	mf-ek/orbit. height x100	76.2	97.0	80.3
n-b	107	apt-apt/n-ns x100	47.2	45.5	47.5
b-l	90	zy-zy/eu-eu x100	95.6	106.4	92.1
l-i	65	ft-ft/zy-zy x100	72.5	56.0	74.8

x_a — analysed skull features, x_b — comparative skull features according to the data by Horackowa [4] and the mean of the features (x) of Bulgarians by Malinowski et al. [8]

sociation of several features as for instance: prognathism, strong forehead inclination, strong development of supraorbital arch and strong external occipital protuberance. The last feature has usually been localised over the Frankfort plane [6]. Also the disproportion between the level of development of the craniocerebral and facial part of cranium is emphasised.

Analysis of the described case of microcephaly confirmed some of these observations. There are obvious similarities with reference to prognathism, the shape of supraorbital arches, and external occipital protuberance. However it must be stated that external occipital protuberance is situated under the Frankfort plane i.e. in the same position as in contemporary, normally developed skulls.

Also the disproportion between the development of calvaria and visceral cranium was confirmed. The underdevelopment of calvaria was distinctly marked as well as tendency to diminish the size deflection of visceral cranium.

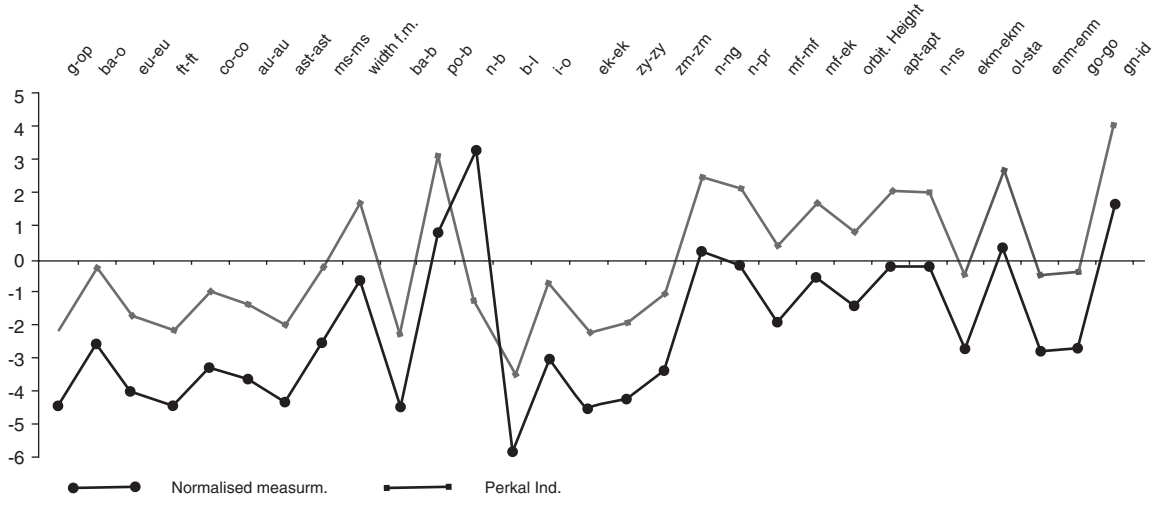

Figure 2. Diagram presenting features of skulls with microcephaly compared with natural Perkal indices.

During the comparison of the morphological construction of different skulls with microcephaly one more peculiarity attracted our attention. Most of the skulls are characterised by dissoclusion and partial apertognathia [2,4,13,15]. Such reciprocal position of masticatory system often causes wrong evaluation of the degree of prognathism, which may seem bigger than in reality.

At the end of our consideration we would like to state that further analyses of skulls with microcephaly will lead to establishing a more detailed list of features referring to that type of pathology. They will also contribute to the determination of the morphological variability of these features within the group, based on statistically reliable numbers.

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