FREQUENCY OF *SPINA BIFIDA OCCULTA* AND OTHER OCCULT SPINAL DYSRAPHISMS IN THE MEDIEVAL POPULATION OF IAŞI CITY: SKELETON PALEOPATHOLOGY IN THE NECROPOLIS DISCOVERED IN THE EASTERN PART OF THE PRINCELY COURT ("*CURTEA DOMNEASĂ*"), 17TH CENTURY

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Abstract. *Spina bifida occulta* is the most common congenital malformation localized at the level of the spinal column from the atlas to the sacrum. The most frequent cases of spina bifida appear in the lumbar-sacral region, especially at the level of the sacrum bone. The maternal nutritional status, the exposure to teratogenic factors and the genetic predisposition may act together leading to the appearance of spinal dysraphisms. The osteological material exhumed in 2008 by the Center of European History and Civilization of Iaşi, from the necropolis localized in the Eastern part of the ex-Princely Court ("*Curtea domnească*") of 17th century), sums up to 111 skeletons from 60 inhumation graves. The occult spinal sacral dysraphism in case of the medieval population of Iaşi appears in 6 cases (5 men with ages between 18 and 60 and one woman with the age between 18-20 years). The percentage of the occult spinal dysraphism is of 6.59% in case of the total number of individuals where we deal with the presence of the sacrum bone, of 7.57% for the male group and of 4% if we refer to the female group.

Keywords: paleopathology, *spina bifida occulta*, medieval population, Iaşi City (Romania).

Rezumat. Frecvenţa *spinei bifida occulta* în populaţia medievală a oraşului Iaşului: paleopatologie scheletică în necropola situată în partea estică a Curţii domneşti, secolul al XVII-lea. Spina bifida occulta este cea mai comună malformaţie congenitală localizată la nivelul coloanei vertebrale de la atlas până la sacrum. Cele mai multe cazuri de spina bifida apar în regiunea lombo-sacrală, în special la nivelul osului sacrum. Statutul nutriţional matern, expunerea la factorii teratogeni şi predispoziţia genetică pot acţiona împreună conducând la apariţia dysraphismelor spinale. Materialul osteologic deshumat în 2008 de Centrul de Istorie şi Civilizaţie Europeană din Iaşi, din perimetrul necropolei localizată în partea estică a fostei Curţi Domneşti (secolul al XVII-lea), însumează 111 schelete provenite din 60 de morminte de înhumaţie. Disrafismul spinal ocult sacral în populatia medievală ieşeană apare în 6 cazuri (5 indivizi de sex masculin cu vârste cuprinse între 18 şi 60 de ani şi la o femeie cu vârsta cuprinsă între 18-20 de ani). Ponderea dysrafismului spinal occult este de 6,59% în cazul numărului total de indivizi la care este prezent osul sacrum, de 7,57% raportat la grupa masculină şi de 4% dacă ne referim la grupa feminină.

Cuvinte cheie: paleopatologie, spina bifida occulta, populaţie medievală, oraşul Iaşi (Romania).

Introduction

The anomalies or congenital malformations have been described even from the oldest of times and they are produced by the pathological modifications of the normal development during the intrauterine life (Ortner, 2003).

The genetic theory as cause of the congenital malformations developed after 1900, one with the rediscovery of Mendel's laws by Carl Correns, Hugo de Vries and Erich von Tschermak (Savona-Ventura, 2007). A great number of congenital malformations can appear at the level of the skeleton, being localized on the skull, at the level of the spinal

column and of the limbs. The congenital malformations may vary from light to severe or even lethal deformations, leading to the decrease of the expectation of life or its quality (Aufderheide & Rodriguez-Martin, 1998).

During prehistoric times, the congenital malformations were less frequent identified because most of the affected children died shortly after birth or later because their fragile skeletons could not survive the taphonomic process (Barnes, 1994).

The greatest incidence of these anomalies was registered at the level of the spinal column, spina bifida being the most common congenital malformation where the osseous median line is incompletely closed, the majority of the cases appearing in the lumber-sacral region of the column, especially at the level of the sacrum bone. The term of "spina bifida" was first described by F. von Recklinghausen in 1882 (Aufderheide & Rodriguez-Martin, 1998). The spinal dysraphism (spina bifida, spinal defect, defect of the neural tube, opened spinal column) comprises all the forms of the congenital affections of the spinal column resulting into a defect neuronal arch through which the meninx or the neuronal elements can protrude (Dawson, 2006).

Spina bifida can provoke complications of the intestine and of the bladder, in case of the children leading to the excessive accumulation of cephalorachidian liquid in the brain (hydrocephaly). With adequate care, most of the children suffering of spina bifida can reach the adult age (Fornoff *et al.*, 2004).

Generally, spina bifida decreases with age, especially in the case of women, the exact cause of this differences between sexes still remaining unknown (Aufderheide & Rodriguez-Martin, 1998). The defects at the level of the neural tube have a genetic predisposition that manifests only in presence of certain risk factors or exogenous factors. During the last decade it was noticed that the deficit of folic acid, zinc and selenium from the maternal organism may determine the appearance of the neural tube defects at the embryo. All these three nutritive substances are necessary for the regulation of the genetic control of the cellular growth during the morphogenesis process (Barnes, 1994). Thus, the maternal nutritional status, the exposure to the teratogenic factors and the genetic predisposition can act together to provoke the spinal dysmorphisms.

According to the clinical manifestations and to the embryonic development stage the dysraphic anomalies can be divided into two categories: opened spinal dysraphisms and closed spinal dysraphisms. The closed spinal dysraphisms are of two types: with a subcutaneous mass and without a subcutaneous mass (Kumar & Shane Tubbs, 2011). Another classification of the spinal dysraphisms: spina bifida cystica or aperta (myelomeningocele or meningocele) and spina bifida occulta that manifests by an incomplete fusion of the posterior neuronal arch and it can affect several sacral segments. Spina bifida cystica or aperta is a more severe form in comparison with spina bifida occulta and it can even be fatal (Aufderheide & Rodriguez-Martin, 1998). The genetic studies show that the forms of spina bifida occulta and cystica are different expressions of the same dominant gene (McKusick, 1998).

The occult spinal dysraphism is a heterogeneous group of lesions where the neural tissular exposure is not visible (Gutierrez *et al.*, 1994). Spina bifida occulta can be localized at the level of any segment of the spinal column from the atlas to the sacrum but it is more commonly encountered in the lumbar-sacral region (L5-S1), with a reported incidence of up to 25% (Barnes, 1994).

The epidemiological studies indicated a great variation in the prevalence of spina

bifida based on ethnicity, typology, temporal and geographical tendencies (Josan *et al.*, 2008). Women are more exposed than men in cases of spina bifida and anencephaly. The Paleoanthropological researches regarding spina bifida were mostly made on the sacrum bone. Sacral spina bifida occulta has been widely recorded and reported as a congenital anomaly by paleopathologists (Kumar & Shane Tubbs, 2011).

Along the processes of osseous growth the sacral vertebrae gradually fusion and in some cases their number can vary by adding the sixth lumbar vertebra, also called transition lumbar-sacral vertebra or by sacralization of the last lumbar vertebra or of the first coccygeal vertebra. Also, the lumbarization (total/partial, symmetric/asymmetric separation of the first sacral vertebra from the sacrum bone) produces modifications at the level of the sacrum bone (Aufderheide & Rodriguez-Martin, 1998). These types of anomalies appear in the case of 3-5% of the population, of which two thirds present the sacralization L5 (López-Durán, 1995).

The sacralization of the 5^{th} lumbar vertebra (L5) is a defect in the case of which this vertebra fusions with the sacrum bone so that the lumbar spinal column loses a segment. The morphologic aspect of the sacrum bone is normal but it presents a supplementary sacral foramen (five sacral foramens). On the contrary, if the first sacral segment is separated from the sacrum bone, this defect is called lumbarization S1. In both cases the defect can be complete or incomplete, unilateral or bilateral, symmetric or asymmetric (Aufderheide & Rodriguez-Martin, 1998; Barnes, 1994).

The archeological researches made in 2008 by the Center of European History and Civilization of Iaşi, in the central area of Iaşi (the Eastern part of the ex-Princely Court – the Area of the Palas Complex) led to the discovery of a necropolis with a number of 111 human skeletons from 60 inhumation graves. According to the data provided by the authors of the diggings, the necropolis was dated from the late Middle Ages – the 17^{th} century. From a historical point of view the evolution of the city of Iaşi took place in the conditions of permanent disasters, being considered to be a martyr city (Cloşcă, 2008).

Material and Methods

The osteological material represented by the 111 human skeletons (teenagers, adults, grown-ups and senile) present a good preservation state, the anthropological analysis as well as the paleopathological one, being easily realized. The osseous pieces were submitted to an elaborated morphoscopical analysis, determining the age and sex of each skeleton, the anomalies and pathologies. The determination of the sex and age at death of the individuals that are over 18 years old was made by using the methods recommended by Bruzek, 2002; Brothwell, 1981; Schmitt, 2005 and Walrath *et al.*, 2004. The age at death in case of the sub-adults was made on the basis of the analysis of the development stages of the dental buds, according to the methodology proposed by Ubelaker, 1979; Schaefer *et al.*, 2009, as well as through the analysis of the degree of ossification of the epiphyses, of the dimensions of the long bones and their inclusion within the corresponding age categories, according to Trotter & Peterson, 1969.

For the identification of spina bifida or of the spinal dysraphism there were monitored the anomalies from the level of the median line of the spinal column indifferently from level of its localization. In case of the absence of the hydrocephaly the anomaly is considered to be "spina bifida occulta" or "occult spinal dysraphism". Spina bifida occulta at the level of segments S3, S4 and S5 should be included within the limits of

normal variation of the sacral hiatus. Nonetheless, with regard to the identification of the dysraphism, all the cases of spina bifida must be registered indifferently from their level of appearance on the spinal column (Kumar & Shane Tubbs, 2011).

In case of the total sacral occult spina bifida, the posterior laminas of all the sacral vertebrae are completely destroyed (Senoglu *et al.*, 2008). Regarding the spondylosis, it can be defined as a pars- interarticularis failure of ossification of the vertebra, leading to its separation into two parts: one ventral part made of the vertebral body and a dorsal part represented by the spinous process and by the inferior articular process (Aufderheide & Rodriguez-Martin, 1998). In parallel with the occult spinal dysraphism we also identified other osseous anomalies (segmentation anomalies, atrophies of the appendicular skeleton, hernias etc.).

Results and Discussion

From the total of the 111 human skeletons exhumed from the necropolis situated in the Eastern part of the Princely Court (Curtea domnească), 15 subjects were determined to be sub-adults (juvenis), and 96 subjects (70♂ and 26♀) were over 20 years old (adultus, maturus, senilis). The demographic study showed that within the 0-14 year interval no death was registered within the teenage stage (14-20 years) the percentage of the deaths being of 13.51% with greater frequencies in case of the men. For the 20-x years interval, the highest mortality is registered during the mature stage (30-60 years - 65.77%), in case of the adults the percentage of the deaths is significantly reduced (18.92%). For the 60-x year age stage there were registered only two cases of men death (1.80%). The number of the male skeletons significantly exceeds the number of female skeletons (80/31), a phenomenon that can be observed for the mature and adult age level and for the teenage level. The expectation of life at birth for the entire analysed series (0-x years) is of about 36 years and the average life time for the subjects that surpassed the age of 20 is of 39.86 years in case of men and of 35.96 in case of women.

From the total number of 111 individuals with ages between 14-x years, the sacrum bone is present in 91 of the cases (66♂ and 25♀). From the total of 91, a number of 6 sacrum bones present the occult sacral dysraphism (spina bifida occulta). A number of 5 sacrum bones with occult spinal dysraphism belong to male individuals with ages between 18 and 60 years and only 1 sacrum with occult spinal dysraphism belonged to a woman aged between 18-20 years. Thus the percentage of the occult sacral dysraphism is of 6.59% in case of the total number of individuals that have the sacrum bone, of 7.57% if we refer to the male group and of 4% if we refer to the female group.

The first case of occult spinal dysraphism was remarked on a male individual (noted M XIV), death age between 18-19 years. The skeleton of this individual exhumed from an individual grave is well preserved and almost complete. At the level of the cranium there are 5 wormian bones on the lambdoid suture and 3 wormian bones on the parietal-temporal suture.

The calculated waist of this subject is of 1657 mm, which corresponds to the average category. The tibiae and the humeris present osseous excrescencies in place of the muscular insertions. The sacrum of the individual M XIV (Fig.1) presents an occult spinal dysraphism type *"posterior spina bifida occulta*" with a dehiscence localized at the level of the segment S3-S5. In case of the individual M XIV, the sacral dehiscence is not associated with other anomalies or pathologies signalled at the level of the skeleton.

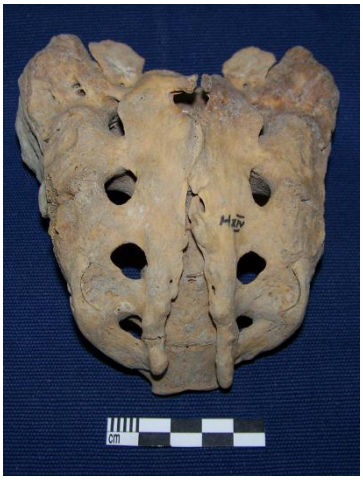

Figure 1. Male, 18-19 years old (M XIV), sacrum with occult spinal dysraphism from the segment S3 and S5.

The second case of occult spinal disraphism was signalled at a female individual (noted G8 M25) with the death age of 18-20 years. This subject was inhumed together with other two matures aged between 40 and 50. The post-cranial skeleton of this female is incomplete and from the cranial skeleton only the frontal and part of the occipital bone remained. The clavicles present very deep impressions of the costal-clavicle ligament that are very similar to an infectious process. The (incomplete) sacrum bone of this individual present a dorsal inter-laminar dehiscence at the level of segments S3-S4 (occult spinal dysraphism) in which the posterior laminas of the sacral vertebras are un-welded within this segment (Fig. 2).

Figure 2. Female, 18-20 years old (G8 M25), sacrum bone with occult spinal dysraphism in the S3-S4 region.

Because of the defective preservation of this skeleton we were not able to identify other anomalies associated to the occult spinal dysraphism so that we could not offer a precise pathological diagnosis.

The third case of occult spinal dysraphism was identified in the case of a male individual (noted M XIII), about 25-30 years old. The postcranial skeleton of the man is incomplete, but the existing osseous pieces are well preserved (the calculated waist is of 1671 mm – the over-average category). The cranium presents 10 wormian bones at the level of the lambdoid suture and one more bone at the level of the coronary suture. This individual presents a sacrum bone with a dorsal interlaminar dehiscence within the S2-S5 region, the space between the laminas being similar to a channel (Fig. 3) and the incomplete anterior central lumbarization (separation) of the first sacral vertebra S1 from the rest of the sacral body (Fig. 4).

Figure 3. Male, 25-30 years old (M XIII), sacrum bone with an occult spinal dysraphism within the segments S2-S5.

Figure 4. Male, 25-30 years old (M XIII), sacrum bone with incomplete central lumbarization S1.

The skeleton of the subject noted M IV belonged to an adult male deceased when he was 25-30 years old. The very well preserved cranium presents 9 wormian bones at the level of the lambdoid suture. The frontal bone presents a dislevelment near the coronary suture that resulted from a hit. The post-cranial skeleton lacks the tibias, the fibulas, a fragment from the left femur and a fragment from the right calcaneus. The sacral occult spinal dysraphism at the level of the sacrum bone presents 2 dehiscences: the first one is localized within the segment S1, and the second one within the segment S3-S5 (Fig. 5). The dehiscence at the level of S1 has a percentage of 20% from the total types of closed spinal dysraphisms and it does not present other associated clinical symptoms (Tortori-Donati *et al.*, 2000). Regarding the occult spinal dysraphism at the level of the segment S3-S5, this represents a frequently signalled defect from the specialty literature.

Figure 5. Male, 25-30 years old (M IV), sacrum bone with occult spinal dysraphism within the S1 and S3-S5 segments.

The mature male subject (noted G10 M33 –A), deceased at the age of 40-45 was inhumed together with other three individuals (two females with the ages of 20-22 and respectively 50-55 and a male individual with the age of 40-45). The cranium of the subject G10 M33 is absent and the postcranial skeleton lacks: the tibias, the fibulas, the left radius and the left cubitus. The waist of this subject is of 1796 mm, being part of the high category. This individual presents a sacrum with total sacral occult dysraphism (L5 respectively S1-S5) – Figure 6, and an incomplete central sacralization (welding) of the last lumbar vertebra L5, so that on the sacral body appear 5 sacral foramens (Fig. 7).

Figure 6. Male, 40-45 years old (G10 M33 -A), sacrum bone with occult spinal dysraphism within the L5 la S5 segment.

Figure 7. Male, 40-45 years old (G10 M33 -A), sacrum bone with an incomplete central sacralization in the L5 region.

The M XI skeleton, which is well preserved, belonged to a mature man (55-60 years old), average waist (1667 mm). The cranium presents an orifice resulted from an ante-mortem hit in the right parietal region, the postcranial skeleton presenting exostoses on the dorsal vertebrae. Just like in the previous case, the sacrum bone of this subject presents a total occult sacral dysraphism - S1-S5 (dorsal interlaminar dehiscence), forming a channel between the laminas (Fig. 8).

Figure 8. Male, 55-60 years old (M XI), sacrum bone with total occult dysraphism S1-S5.

Badly preserved, the skeleton G15 M53-D belonged to a male teenager (18-20 years old) whose waist could not be calculated because of the absence of the limbs bones. The cranium, which is fragmentary, incomplete and deformed post-mortem, cannot be used for a complete biometrical study. In this case also the sacrum bone of the individual G15 M53-D presents the sacralization (welding) of the fifth lumbar vertebra (L5) to the sacral body (Fig. 9). Another signalled anomaly is the bilateral spondylosis at the level of the lumbar vertebra L4 – defect of the inter-articular ossification (Fig. 10). The etiology of spondylosis represents a controversy for some authors being seen as a congenital malformation due to some defects of ossification and development of the laminas.

Turkel (1989) underlined the fact that there is a genetic element that plays very important role for the spondylosis but Stewart (1953) proved that the frequency of spondylosis increases with age and he suggested that the hyper-flexion of the spinal column simultaneously with the knees extension led to the appearance of micro-traumas – fractures in the pars-interarticularis area (Aufderheide & Rodriguez-Martin, 1998). There were not proven any clear differences between women and men although a little predilection in the case of men was observed. It was noticed that the number of the Caucasians that were affected is greater in comparison with the number of the Black people (Aufderheide & Rodriguez-Martin, 1998).

Conclusions

By this paper we underlined the frequency of the occult sacral dysraphism (sacral spina bifida occulta) in case of a medieval population exhumed from the necropolis (dating

Figure 9. Male, 18-20 years old (G15 M53-D), sacrum bone with an incomplete, central sacralization of the L5 vertebra.

Figure 10. Male, 18-20 years old (G15 M53-D), the L4 vertebra with bilateral spondylosis.

from the 17th century), situated in the Eastern part of the ex-Princely Court (Curtea domnească) of Iași. In the case of this population the percentage of the sacral occult spinal dysraphism is high (6.59% of the total number of sacrum bones, 7.57% of the total number of male sacrum bones and 4% of the number of female sacrum), in comparison with the values that were obtained for another population of the medieval Iași (1.55% of the total number of sacrum bones and 3.22% of the number of male sacrum bones) – (Simalcsik *et al.*, 2011) and lower in comparison with the values that were obtained for other medieval European populations. From the 6 cases of sacrum with occult sacral dysraphism identified within the sample we analysed, five belonged to male individuals with ages between 18- and 60 and one belonged to a female who was 18-20 years old. The occult spinal dysraphism characterized by the incomplete closure of the osseous tissues at the level of the median line has a multi-factorial origin, involving both the genetic and environmental factors that play a releasing role.

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