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INFANTILE OSTEOPATHOLOGICAL CONDITIONS IN ONE CASE OF FORCED INANITION

Luis Ríos
Mario Vásquez
Doménec Campillo
Mary Lewis
Jason Wiersema

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This osteopathological research is a derivation of the multidisciplinary work between the team of Forensic Anthropology of the Human Rights Bureau of the Archbishopric of Guatemala (ODHAG), Luis Ríos and Jason Wiersema, carried out in the month of June, 2003. We are presenting here a case of collective forced inanition which took place in 1982 at Ilom, a village located in Chajul, Quiché, where according to testimonies of those who survived, on the 23 of March the army stormed in this community as a part of the "swept land" strategy. At that time, men were locked in the school, women and children in the chapel; immediately after, around 90 men were murdered and the homes of the villagers burnt. Following these events, the survivors were forced to move to *finca* Santa Delfina –owned by José Luis Arenas, known as *The Ixcan Tiger (el Tigre del Ixcan)*-, where an army detail was stationed. The residents of Ilom were forced to work at the *finca* without a salary, and therefore access to food was extremely limited. According to the testimonies of the surviving parents, it was estimated that 150 children died, as they were the most vulnerable in this situation, and many of them had no food at all. In the case of the youngest ones, the mothers referred that due to the massacre they had witnessed, they panicked and their bodies dried out of milk, the only nourishment their children could receive, which led to their death; this came together with different symptoms such as fever, diarrhea, vomiting, phlegm, change in skin color, abundant crying, etc. The infants were buried in the village cemetery, with the previous authorization of the officer in charge of the army detail.

CASE 1

We shall now describe a sub-adult skeleton with a severe pathological condition. In the cases discussed here, diagnosis is complicated due to the fact that the skeletons must be considered within the context of an acute episode of malnutrition, probably famine. This makes it even harder to establish the pathological condition caused by a genetic mutation from another one resulting of nutritional deprivation. In the skeleton, acute malnutrition is manifested by different forms of pathological lesions, some of which may resemble the combined lesions resulting of some genetic diseases, including imperfect osteogenesis. Therefore, they will be considered as

genetic and nutritional originated diseases, whose bone manifestations are consistent with those presented in this case, though we shall describe in a more detailed manner what imperfect osteogenesis type II and achondrogenesis are about.

IMPERFECT OSTEOGENESIS, TYPE II

Imperfect osteogenesis is a congenital disease which produces a deficiency in the type II collagen. Imperfect osteogenesis of the type II (IO type II) is characterized by a generalized and intense osteopenia, to the point that it makes the skeleton susceptible of fracture with a minimum prenatal stress. Typically, all the long bones, as well as the ribs and collarbones, show multiple fractures with the subsequent deformation. Although the formation of the bone callus is generally normal, the replacement of lamellar bone is not adequate, so that bones are susceptible to additional fractures. The nature of this disease makes the bones which support the weight to be more susceptible to fractures. The content of collagen in the dentine is also abnormal, often resulting in a debilitated joining of the enamel with the dentine, and the consequent flaking of the latter. This condition is known as imperfect dentinogenesis.

ACHONDROGENESIS

It is possible that the skeleton in question suffered from achondrogenesis, a hereditary disease of a recessive character and of the dysplasic type, somehow related to achondroplasia. In this pathology, in general, neonates are born dead or die within a short period of time. According to its definition, achondrogenesis is a *"hereditary disorder characterized by bone hypoplasia, resulting in very short extremities, where the head and trunk are normal".* It corresponds to a polysystemic, hereditary, recessive disease, and is not related to sex (Puigdollers et al. 1970). According to Gorlind et al. (1979), Parenti, in 1936, separated it from achondroplasia for being characterized by: 1) lethality; 2) a disproportionate large head; and 3) accentuated shortening of the limbs and trunk. There are descriptions of cases involving siblings and kinship.

The pathological proximity of this condition with achromegalia is clear, and probably, according to different authors, it is a variety of that disease; however, several particularities would point to the presence of other congenital factors that could account for symptomatic heterogeneity, while still other authors include them among the skeletal dysplasias (Dugoff et al. 2000).

Jaffe (1978) makes no separation with achondroplasia, and includes it in a chapter denominated *"Findings in children and adults with achondroplasia"*, where he outlines that the surviving individuals *"represent slighter cases of achondroplasia"*, and considers that the anatomical studies made so far are insufficient: *"… we know that in the affected children, the postnatal centers of ossification develop in several cartilogenous epiphysis and in the precursors of the tarsal and carpal bones… atrophy in their large bones is largely due to a feeble growth of the epiphysis*

cartilaginous discs… a contributing factor in these cases could be the premature, partial or full joining of many epiphyses". Furthermore, it is noted that *"limbs are too short and convex…their length rarely exceeds 20 cm… the degree of ossification of the long bones varies fairly… the metaphysis of the humerus bone is broadened and presents the form of a cup…"* (Gorlind 1979). Although reference is made to macrocephalia, this is not always present and not all infants are born dead, though their passing usually takes place within a few days or weeks. In addition, there is a published classification based on radiographic criteria, where there are comments in the sense that the more common diseases are *"… achondroplasia, thanatophoric dysplasia, imperfect osteogenesis, and achondrogenesis"* (Dugoff et al. 2000).

RACHITISM

This is a non-lethal disease characterized by an anomalous curvature of the long bones, and it rarely shows up prior to the fourth month of life.

HYPOPHOSPHATASIA

Fractures may occur in the long bones, but in a lesser degree that in the cases of IO type II. The vertebrae of the affected children are extremely thin.

CAMPOMELIC DYSPLASIA

There is a reduced bone density, though restricted in most cases to the long bones, sparing other bone elements.

CONGENIT SYPHILIS

Frequently, it is lethal prior to becoming evident in the bone tissue. The fracture of the long bones is restricted to those regions whose structure is connected with syphilitic tissue. The long bones are often thickened as a consequence of bone deposition (periostitis).

CONCLUSION

It is suggested that OI type II and maybe achondrogenesis are the more probable explanations for the combined pathologies present in this skeleton. However, the situation of a deficient nutritional environment around this child makes it very possible that the multiple pathological processes that influenced the morphology of this skeleton may have remained hidden.

CASE II

We shall now describe the possible lesions found in other skeletons originated in the same exhumation. The lesions were divided according to their localization in the endocranium and the long bones. The endocranial lesions appear as layers of new bone on the original cortical surface, expanding around the meningeal vases in the form of isolated plates, like “frozen” extensions of the diploe or in the form of capillary impressions that reach the internal plate of the brain. These lesions are mainly located in the occipital bone, though there were also observed in the parietal and frontal bones, and it would seem that they are distributed across the areas of the venous draining system of the cranium (Lewis 2004). Though there is general consensus in recognizing that these lesions are the result of hemorrhage and swelling of the meningeal vases, its precise etiology is still been debated (chronic meningitis has been suggested, as well as trauma, anemia, neoplasia, scurvy, rachitism, and tuberculosis, like possible causes). For an illustration of these lesions, one type of the four described by Lewis (2004) may be observed, in cases derived of the study of a children’s sample recovered in four medieval and post-medieval English archaeological deposits.

In the case of the occipital bone, we may note a cavitary exostosic reaction that reminds that of the sieve, this said without the intention of including this highly controversial disorder in the field of paleopathologic investigations. Actually, it seems to be a secondary reaction to an inflammatory process, but it would be risky to provide an etiology, although phlebitis of an infectious etiology could be suggested. According to Lewis’ conclusions (2004), the most adequate thing to do would be to refer to these lesions as non-specific indicators of hemorrhage or infection.

The long bones also presented lesions on their outer surface. These long bones feature interesting alterations present in different sectors of the bone cortical, showing a moderate exostosic reaction with small cavitations, suggesting a periostitis of the type 5 and an uncertain etiology, of which for now, an infectious etiology was excluded.

CONCLUSIONS

This research has presented several preliminary observations on the pathological conditions observed in a forensic case. In this case, only sub-adult skeletons were exhumed, and according to testimonies, the population was subjected to forced famine, one that caused the death of several tens of children. The present examination of the skeletons recovered show some of the pathological conditions found on them. It is difficult to associate these pathological conditions with the famine event due to the discussion on their etiology. Different conditions, such as the Harris lines or the hypoplasia of the enamel are not of any use at this time, as it is necessary that the individual continues to grow in order to show such conditions. However, it is interesting to note that in the exhumation, six sub-adult skeletons were recovered, thus allowing us to observe the endocraniums, and that four of them presented lesions of the type described above.

If the number of exhumed skeletons would have been higher, the presence of this type of lesions in an elevated percentage of cases could suggest a generalized deficiency of the population during the same time-interval, and this could in fact have been associated with the episode of forced famine.

This work is dedicated to the victims of the armed conflict, particularly to the girls and boys of Ilom and their families.

REFERENCES

Dugoff, L., G. Thieme, and J.C. Hobbins
2000 Anomalías esqueléticas (Skeletal Anomalies). *Clin Parinatol* 27 (4): 979-1006.

Gorlind, R.J., J.J. Pindborg, and J.R. Cohen
1979 *Síndromes de la cabeza y del cuello.* Barcelona.

Jaffe, H.J.
1978 *Enfermedades metabólicas, degenerativas e inflamatorias de huesos y articulaciones.* La Prensa Médica Mexicana, México.

Lewis, M.
2004 Endocranial Lesions in Non-Adult Skeletons: Understanding Their Aetiology. *International Journal of Osteoarchaeology* 14: 82-97.

Parenti, G.C.
1936 Una varietá della osteogenesi imperfecta. *Patológica* 28: 241-262.

Puigdollers, J.M., Ribas, M., and Gri, E.
1970 Enfermedades hereditarias sistémicas. In *Tratado de patología y clínicas médicas* III (edited by A. Pedro Pons), pp. 1157. Salvat, Barcelona.