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Paleopathological rigor and differential diagnosis: Case studies involving terminology, description, and diagnostic frameworks for scurvy in skeletal remains

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ABSTRACT

Diverse pathological processes can produce overlapping or even indistinguishable patterns of abnormal bone formation or destruction, representing a fundamental challenge in the understanding of ancient diseases. This paper discusses increasing rigor in differential diagnosis through the paleopathological study of scurvy. First, paleopathology's use of descriptive terminology can strive to more thoroughly incorporate international standards of anatomical terminology. Second, improved observation and description of abnormal skeletal features can help distinguish between anemia or vitamin C deficiency. Third, use of a structured rubric can assist in establishing a more systematic, replicable, and precise decision-making process in differential diagnosis. These issues are illustrated in the study of two new cases of suspected scurvy from northern Peru. From this, it appears possible that ectocranial vascular impressions may further be examined as a morphological marker of scurvy in the skeleton. Also, increased paleopathological attention to pellagra is long overdue, especially as it may produce generally comparable lesions to scurvy. This paper reflexively speaks to the process of paleopathological problem solving and the epistemology of our discipline—particularly regarding the ways in which we can continuously improve description and the construction of diagnostic arguments.

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1. Introduction

Skeletal diseases involve remarkably complex interplays between pathogens and the human body's innate reactions to biological stress, involving inflammatory responses, molecular signaling factors that direct a spectrum of osteoblastic and osteoclastic activity, and intricate osteoimmunological interactions (Ortner, 2003; Gosman, 2012; Choi, 2012; Burr and Allen, 2014; Klaus, 2014; Lorenzo et al., 2015). Despite this, skeletal phenotypic responses to disease tend to be evolutionarily and biologically constrained, such that a diverse range of pathophysiological processes may produce analogous if not sometimes indistinguishable patterns of abnormal bone formation and destruction. This reality represents one of the most important challenges in the identification of ancient disease and the study of human health in the past.

To counter this often-frequent lack of lesion specificity, paleopathology must employ a rigorous approach toward the description and differential diagnosis of skeletal abnormalities. Even in 19th century work of Sir Marc Armand Ruffer, which is widely credited as the starting point of paleopathology, he emphasized clear and detailed description, analogies with clinical literature, and multidisciplinary analysis (Sandison, 1967; Grauer, 2012). Since then, paleopathology has evolved in diverse directions. Today, we are in the midst of what can be described as the fourth wave of paleopathology, focused on ecological, evolutionary, cultural-contextual, and epidemiological dimensions of ancient disease (Angel, 1981; Ubelaker, 1982; Zuckerman et al., 2012). The strong influence of the 'new' or Americanist processual archaeology, along with its scientific and empirical trappings, helped stimulate this paradigm shift beginning in the 1960s.

Related advances continue into the 21st century. Particularly important have been the development of carefully constructed encyclopedic descriptions of skeletal diseases (Steinbock, 1976; Ortner and Putchar, 1981; Aufderheide and Rodríguez-Martín, 1998; Ortner, 2003) and the introduction of standardized

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protocols for observation and documentation of skeletal features (Buikstra and Ubelaker, 1994). Once dominated by descriptive studies and ‘snap’ diagnoses of skeletal diseases, paleopathology has also increasingly embraced the use of differential diagnostic protocols as practiced in clinical medicine. Such a systematized approach was arguably most rigorously developed by Ortner (1991, 2003, 2011, 2012). Fundamentally, differential diagnosis is a nested, multi-level problem-solving exercise. It begins on a conceptual and practical level involving an understanding of the normal and abnormal behavior and biology of bone cells (Ragsdale and Lerner, 2012). From there, detailed descriptions of lesions in an affected bone can be made before perspective shifts to the distribution of all abnormal features throughout an entire skeleton. The next level involves classification of skeletal lesions from which likely diagnostic options may be systematically weighed against each other, and various candidate conditions are ruled out. This approach has gone far to improve the scientific nature of disease identification in the past. Yet, it may be possible to continue building upon these past advances to further enhance differential diagnosis in paleopathology.

As part of this special section of the *International Journal of Paleopathology*, this paper primarily focuses on rigor in paleopathology, specifically focusing on the differential diagnosis of scurvy. This is particularly fitting, as scurvy is one of the most challenging and nuanced disease conditions to diagnose in human skeletal remains. This paper contains three linked narratives: (1) a discussion of how anatomical terminology in paleopathological description may be improved; (2) an exercise in rigorous observation and description of abnormal skeletal lesions that demonstrates how one may distinguish morphologically similar orbital lesions produced by anemia, rickets, and scurvy, and; (3) a mini-case study applying the previous two points within a framework for systematic, replicable, and precise decisions in differential diagnosis. While these three narratives happen to topically focus on scurvy, applicability to broader rigor-related issues can be considered, including disciplinary epistemology regarding the ways in which information is perceived, constructed, and organized in paleopathological thought.

2. Scurvy: pathophysiology and skeletal manifestations

The interface between subsistence behavior and metabolic disease in the human past represents a principal concern of paleopathological research. Scurvy, or vitamin C deficiency, provides particularly important perspectives (Ortner, 2003; Brickley and Ives, 2008; Armelagos et al., 2014; Crandall and Klaus, 2014a). Not only is scurvy quite challenging to diagnose in human skeletal remains, it has additionally been a “neglected” skeletal condition in paleopathology until carefully composed diagnostic criteria emerged in the late 1990s (Ortner and Eriksen, 1997). Since then, paleopathological studies of scurvy have expanded significantly, with evidence of vitamin C deficiency being found around the world dating back at least a few thousand years (Brickley and Ives, 2008; Crandall and Klaus, 2014a).

Scurvy, sometimes alternatively referred to as Moller–Barlow’s disease, is the result of a physiologic deficiency of ascorbic acid, which is required for multiple biological functions including collagen formation. Before the divergence of Platyrrhines and Catarrhines, a mutation occurred approximately 35 million years ago in the L-gulonolactone oxidase gene (Nishikimi and Udenfriend, 1976). This error produces the transcription of a premature stop codon in the final set of instructions for production of an enzyme in vitamin C biosynthesis (Nishikimi and Yagi, 1991). Vitamin C must thus be obtained entirely from our diets, and various fruits, vegetables, and even animal sources often contain

sufficient quantities of vitamin C. Understanding the conditions under which deficiencies in vitamin C occurs informs much larger issues involving adaptation, subsistence economy, human–ecology synergisms, urbanism, and socioeconomic inequality (Melikian and Waldron, 2003; Lewis, 2004, 2010; Brickley and Ives, 2006, 2008; Mays, 2008, 2014; Waldron, 2009; van der Merwe et al., 2010a,b; Brown and Ortner, 2011; Geber and Murphy, 2012; papers in Crandall and Klaus, 2014b; Tiesler et al., 2014).

If somatic stores of ascorbic acid fall below 350 mg, a person becomes prone to defective Type 1 collagen and osteoid formation. Consequently, blood vessels become fragile and susceptible to rupture. Periosteal membranes equally develop a propensity to tear and bleed, immune function falters, and suboptimal metabolism of iron and folate occurs (Tamura et al., 2000; Weinstein et al., 2001; Akikusa et al., 2003; Lewis, 2007). Hemorrhage is a pathological hallmark of scurvy. Outside the circulatory system, blood elicits a robust inflammatory response. When hemorrhages form around bones, skeletal signs of inflammatory reactions can be observed. For example, a vascular response in the cranium may be initiated by an incursion of osteoclasts into existing cortical bone that creates channels for newly formed capillaries (usually less than 1 mm in diameter Ortner et al., 1999; Kozłowski and Witas, 2012). This provides pathways for white cells to reach and remove extravasated blood. If formed under scorbutic conditions, new capillaries are themselves prone to tearing and can exacerbate bleeding and inflammation in a feedback loop (Brown and Ortner, 2011, 198).

Should hemorrhage form between the periosteum and the surface of a bone, clotted blood will quickly begin to organize into connective tissue (Ragsdale and Lehmer, 2012). In chronic scurvy, common skeletal sites manifesting new bone formation include the superior orbits, ecto- and endocranial regions of the cranial vault, the alveolar bone of the maxilla and mandible, the hard palate of the maxilla, and the posterior maxilla and mandible (Table 1). Ortner et al. (1999, 2001) argued that abnormal bilateral porosity of the greater wing of the sphenoid bone is virtually pathognomonic for scurvy—a function of chronic bleeding of ruptured connective tissue stemming from minor trauma related to normal chewing (Ortner, 2012). Endocranially, scurvy can produce epidural bleeding as hematomas may tear the dura and periosteum from the bone along with the bridging vessels between the arachnoid and dura layers of the meninges (Kumar et al., 2009; also Lewis, 2004). In the post-cranial skeleton, normal movement of the rotator cuff is linked to bleeding, formation of porous lesions, and new bone deposition in the supra- and infraspinatus fossa of the scapula. Osteochondral junctions of ribs and long bone metaphyses may fracture. New bone ≤ 1 cm thick may be deposited on affected regions of long bone diaphyses. The most massive subperiosteal hematomas are associated with the weight-bearing long bones of the lower limb (Ortner, 2003, 384). Brown and Ortner (2011) and Geber and Murphy (2012) also identified the ilium and the foramen rotundum of the sphenoid bone as additional sites of scorbutic inflammation.

However, scorbutic lesions are often exceedingly subtle in expression and can be easily overlooked (Crandall and Klaus, 2014a). Lesions may mimic normal growth processes especially in subadult skeletons, and may otherwise be challenging to differentiate from normal anatomical variation. Scurvy can produce lesions that morphologically resemble or even directly overlap those produced by anemia, infectious diseases, and myriad other processes (see the following). It is thus a disease condition well-suited to highlight issues surrounding rigor in paleopathology.

3. Terminological rigor: why words matter

Ortner (2011, 2012) made the point that words matter in differential diagnosis, as the terms we use “. . . often make a big difference

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