



Case study

An 8000-year-old case of thalassemia from the Windover, Florida skeletal population

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ABSTRACT

Thalassemia is a congenital blood disorder which destroys red blood cells at a faster rate than can be produced, resulting in anemia. Historically, this disease is found more often in Old World populations, such as Middle Eastern and Southeast Asian. The earliest reported skeletal evidence of thalassemia comes from the eastern Mediterranean (Atlit-Yam) and is correlated with early agriculturalists' exposure to malarial parasites. While there have been virtually no skeletal reports of thalassemia in prehistoric Native American populations, among the individuals from the 8000-year-old hunter-gatherer site of Windover, Florida there is a single potential case of the disease. A female in her early 20's exhibits bilateral foreshortening of the humeri with indications of premature epiphyseal fusion. Both proximal humeri are medio-laterally compressed, the gleno-humeral joint surfaces exhibit medial deformation, and bones show expansion of the medullary cavity with increased cancellous bone growth. These characteristics have been reported as indicators of thalassemia in both clinical and archaeological contexts. Alternate diagnoses such as congenital dislocation or injuries during child birth are considered but fail to account for the full set of characteristics shown. Individual #76 may, therefore, represent the oldest reported case of thalassemia from a native North American skeletal population.

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1. Introduction

The thalassemias are a group of inherited blood disorders that affect the way the body makes hemoglobin, a protein found in red blood cells that is responsible for carrying oxygen throughout the body. While there are multiple types of thalassemia, the two main groups are alpha thalassemia and beta thalassemia (referring to the globin subunits affected). In both cases red blood cells are destroyed at a rate faster than normal, causing anemia and resulting in skeletal pathologies associated with varying degrees of anemic response. Prehistoric cases of thalassemia have primarily been found among Mediterranean populations and date back as far as 6000 BCE at Atlit-Yam, Israel (Hershkovit and Edelson, 1991) and have been linked to incidence of malaria. Today the range of expression of thalassemia spans from the Mediterranean basin into East Asia. The majority of thalassemia cases have been diagnosed through the presence of cribra orbitalia or porotic hyperostosis (giving a hair-on-end appearance in x-rays) and additional anemic responses such as expanded medullary cavities in conjunction with regions known for having heavy malarial infection rates. Thalassemia has also been

tied to a deformation of the shoulder known as humerus varus (Hershkovit and Edelson 1991; Hershkovitz et al., 1991). The severities of bone deformation also vary based on the type of thalassemia an individual inherits genotypically. Major and minor versions of the disorder illustrate drastic differences in the amount of skeletal involvement and the overall life expectancy of the affected individual. Homozygous individuals generally express a greater degree of skeletal malformation compared to heterozygote counterparts (who can sometimes be asymptomatic). In addition, the heterozygote carriers of thalassemia express a level of resistance to malarial infection.

To date, there have been no recorded cases of thalassemia in the prehistoric (pre-contact) North or South American skeletal record. Theoretically, without the presence of malaria in the pre-contact New World, the primary selective factor of protection against malaria is nonexistent within those populations. This scenario has resulted in the a general lack of consideration for this disorder when differential diagnoses are being developed to explain the presence of severe anemic responses of New World skeletal material. Development of additional skeletal symptoms correlated with thalassemia will allow for the diagnosis to be applied to remains expressing these specific bony alterations regardless of geographic location or time period. This broader application may result in the reassessment of New World cases and may show that although

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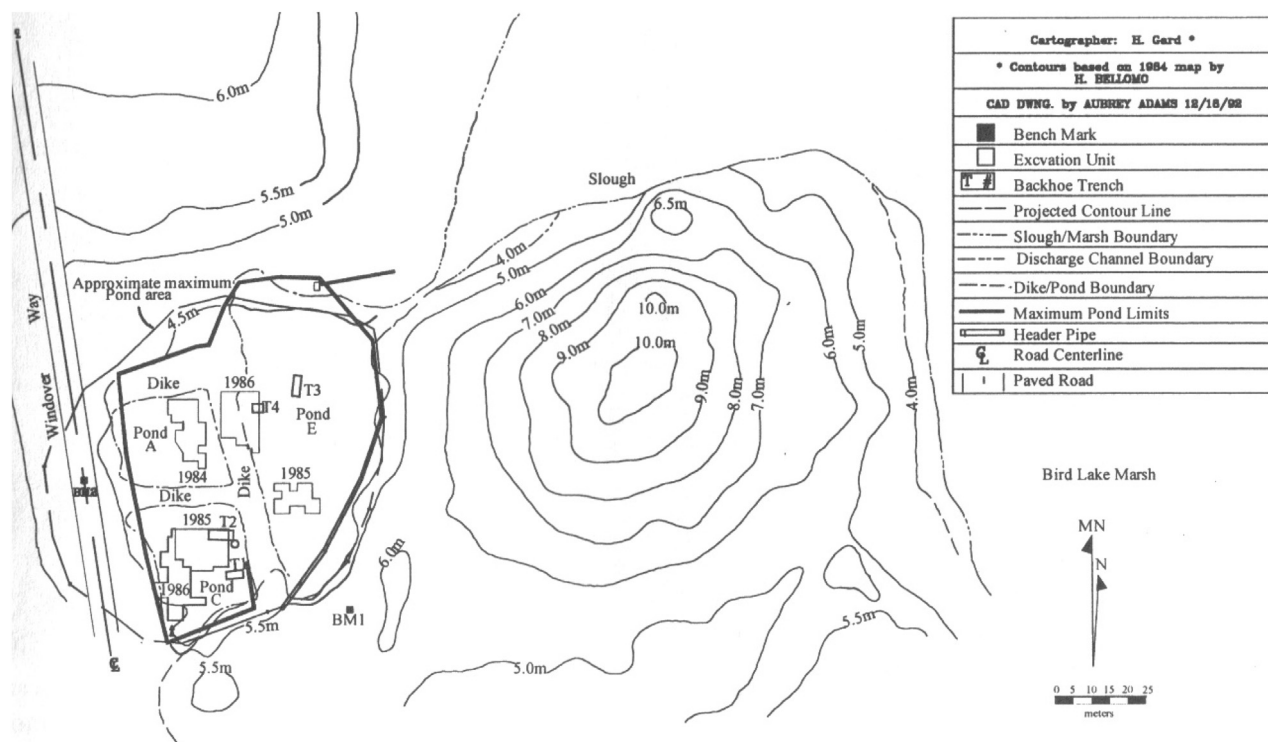


Fig. 1. Windover site map (Doran, 2002).

thalassemia is largely related to the degree of malaria in the environment, it is not the only cause for the disease. It may be rarely seen as a purely genetic disorder, but if the symptoms are present among pre-Columbian Native American remains, thalassemia should not be ruled out as a potential diagnosis purely based on time and location.

2. Materials and methods

2.1. Windover site

North American skeletal collections older than 5000 years are relatively rare, and of these, 60% of the individuals ($N=194$) are from Florida and date to before 7000 years BP. The Windover site yielded a minimum of 168 adult and subadult individuals and dates to the Early Archaic period (8522–7421 BP calibrated) (Doran, 2002). The site, near present day Titusville, was discovered in 1982 during the cleaning of a small pond for road construction in the Windover Farms housing development. During the process a backhoe operator discovered human remains and artifacts in the black peat spoil banks coming from the bottom of the pond. After several excavation seasons (1984–1986) it was discovered that the wet site was a charnel (mortuary) pond containing peat sediments in which burials were placed (Fig. 1). The skeletal preservation was greatly enhanced through the combination of water chemistry, rapid burial, and physical stability maintained by the peat sediments (Doran, 2002). Through the examination of the burials and associated materials it was found that after death the bodies were placed in shallow graves in flexed positions on their sides around the margin of what was a small pond (Doran, 2002). The burials were deep enough to create anaerobic conditions and the context of the preserved textiles indicates that the bodies were possibly wrapped in matting or fabric and marked by protruding wooden stakes driven into the pond floor to delineate individual burials or groups (Dickel, 2002).

There seems to be very little compelling evidence of a significantly marine orientated diet in Florida's early Archaic period (Dickel and Doran, 2002). Through the analysis of stomach contents and bone isotopes (Tuross et al., 1994), the Windover population was found to have a hunter-gatherer subsistence strategy focused on inland riverine, pond, and marsh resources. The primary sources of food included duck, turtle, and catfish as well as large and small terrestrial resources such as deer and rabbit (Dickel and Doran, 2002). In addition, floral remains indicate the use of a wide array of fresh fruits (grapes, prickly pear, and elderberry), nuts, greens, seeds, and tubers (Newsom, 2002).

Due to road construction leading to the discovery of the site, an estimated 58 individuals were recovered as disarticulated scatter. Of the remaining in situ burials now curated at Florida State University, 63 adults (36 males, 27 females) and 30 subadults (under the age of 18) were found in varying degrees of preservation and completeness. Age and sex estimates were calculated based on traditional non-metric pelvic and cranial assessments, as well as subadult dental eruption (Doran, 2002). Antler tools, deer ulna awls, and atlatls were found to be associated with male burials, while antler punches, modified shell, and simple bone pins were associated with females (Doran, 2002). Subadults were found with a mixture of artifacts including awls, a single atlatl handle, ulna scrapers, drilled fish vertebra beads, modified shell, and lithics (Dickel, 2002). Among the individuals from the 8000-year-old hunter-gatherer site of Windover, Florida there is a single individual that expresses bilateral humerus varus deformity.

2.2. Individual 76

2.2.1. Age and sex

The biological sex of this individual was estimated to be female based on the degree of sexual dimorphism present in this population and the gracility of the skull and long bones in comparison to other members of the Windover population. She died in her early-

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