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Unusual Neck Pathology in a Nevisian prehistoric individual

Prehistoric Nevisian neck pathology

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Abstract

Over the years, a small series of skeletal remains have been collected by the Nelson Museum. Despite the poor preservation of the material, several specimens exhibited pathological changes. Severe neck pathology was found in one specimen deriving from the prehistoric site of Coconut Walk. This paper presents the changes in the vertebrae and elsewhere within this individual and proposes block vertebrae (possibly even Klippel-Feil syndrome) as the diagnosis.

Keywords: Prehistoric, Nevisian, neck pathology

Introduction

Over many years, a series of skeletal remains have been recovered from the Caribbean island of Nevis (a small member of the Leeward Islands of the Caribbean). Most of this material is currently curated in the Nelson Museum on the island and consists of skeletal material collected from around the island by interested parties. Apart from one individual, none of the material has been obtained by direct excavation, and thus most are considered as isolated finds within uncertain archaeological associations. The remains of at least twenty-two individuals were studied in 2003. Due to the poor preservation, most of the individuals could not be assigned to a sex and could only be assessed to be "adult". Nevertheless several individuals showed evidence of pathological change, with one being noted for its unusual neck pathology, as described here. This paper presents the changes in the vertebrae and elsewhere upon this individual and proposes the diagnosis of block vertebrae (and potentially Klippel-Feil syndrome).

Nevis in Context

The small island of Nevis is only 90 square kilometres in size and lies near the top of the Lesser Antilles archipelago, about 300 kilometres south-east of Puerto Rico, and immediately west of Antigua (see Fig. 1). The island is littered not only with ruins from the sugar plantation period, but also with both ceramic and aceramic prehistoric sites. The colonial period intensive cultivation of sugar radically transformed Nevisian vegetation, thereby rendering it difficult to reconstruct the prehistoric ecosystem and lifeway (Wilson, 1989: 429). The island appears to have been initially colonised by hunter-gatherers at about 1000 BC, with several later waves of migration by horticulturalists. Ceramic

prehistoric culture in the Lesser Antilles appears in the last few centuries BC (Saladoid), with later prehistoric culture (Ostionoid) appearing by about AD 600 (Wilson, 1989: 430-1). This Ostionoid culture is marked by changes in the zooarchaeological material recovered, which is hypothesised to be the result of either overexploitation of the local environment or of technological innovation (Wing & Scudder 1983) and the standardisation of the ceramic material (Wilson 1989).

Material and Methods

The skeleton discussed here was recovered from the Ostionoid period prehistoric settlement at Coconut Walk (Nevis site code JA-1), radiocarbon dated elsewhere on Nevis to AD 745 ± 135 (Wilson, 1989: 436).

The skeleton, CNW B&E, was collected by John Enwema, and given to the Nevis Historical and Conservation Society, and the material is currently stored at the Nelson Museum, in a box labelled "Skeletal material from Coconut Walk". The skeletal material was studied by the first author (SRZ). The material was stored as two separate individuals (B and E) but was reunited after inspection. Individual B consisted only of a fragmented cranium and cervical vertebrae, whereas individual E consisted only of postcrania (including thoracic & lumbar vertebrae).

For all material studied, including the specimen CNW B&E, sexing was undertaken using traditional pelvis and skull characteristics (following Brickley and McKinley (2004) and Buikstra & Ubelaker (1994)). Individuals were classified on the 1 to 5 scale from definite male, through potential male, unsexed, and potential female, through to definite female. As all material studied was of adults, age was determined by tooth wear (Brothwell 1981), pubic symphysis morphology (Brooks & Suchey

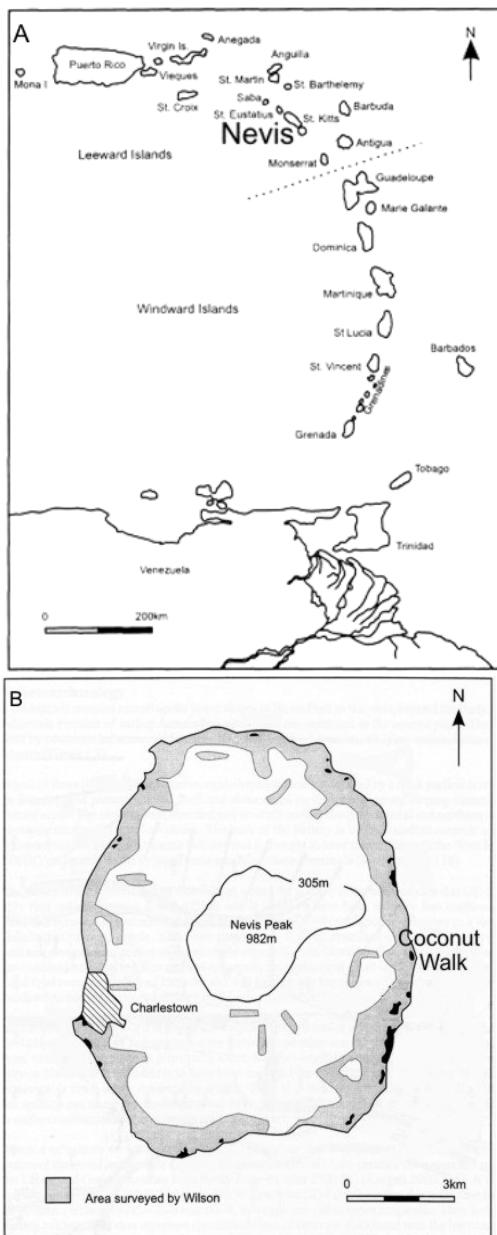


Figure 1. Location of Nevis and Coconut Walk (A: map of the Lesser Antilles; B: Map of Nevis with archaeological sites marked in black). [modified after Crosby (2003: 8-9)].

1990), auricular surface morphology (Lovejoy et al. 1985) and cranial suture closure (Perizonius 1984). As the material was fragmentary, individuals were simply classified into broad age bands (young adult, middle adult and older adult).

Description of material

CNW B&E consisted of cranial fragments, vertebrae and some other postcrania. Excluding the cervical vertebrae, all were highly fragmented. All cervical vertebrae were complete. The atlas exhibited some elongation of the articular facets associated with lippling of the superior aspect (see Fig. 2). The axis was completely fused with C3. Both the bodies and neural arches were entirely

fused together. The dens was unaffected. Severe osteophytic lippling was found on the anterior margin of the inferior surface of the body of C3 (see Figs. 3 & 4). C4 was severely affected as the body is highly curved and thinned, with increased macroporosity and associated with osteophyte development on the anterior margin (Fig. 5). C5 exhibited similar pathological changes. Overall, the cervical vertebrae exhibited mediolateral scoliotic displacement, with the lower vertebrae being displaced towards the individual's right.



Figure 2. Superior view of C1.

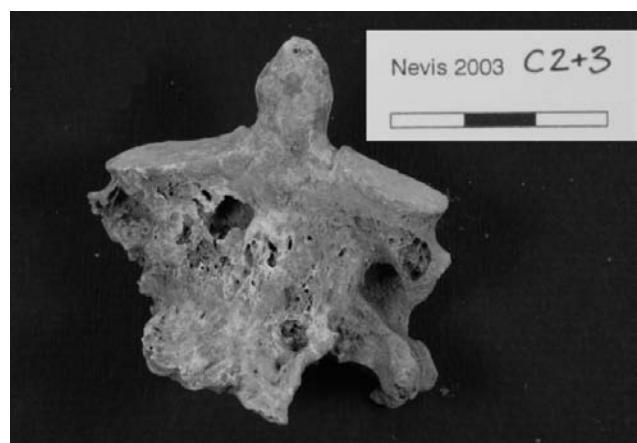


Figure 3. Anterior view of C2 & C3.

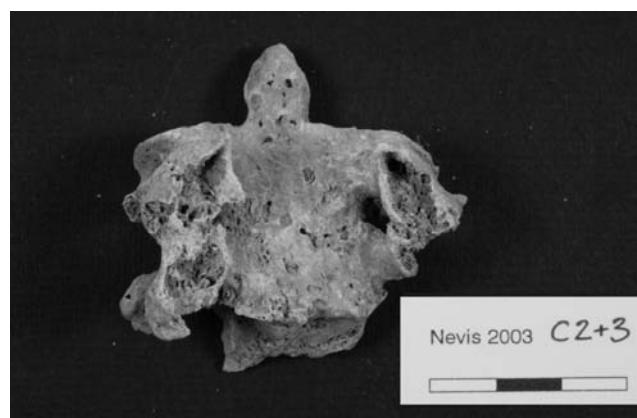


Figure 4. Posterior view of C2 & C3. Note osteophytic lippling of the inferior surface of C3.

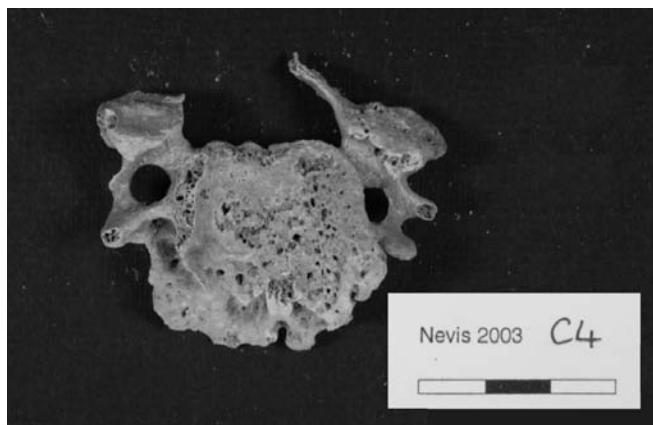


Figure 5. Superior view of C4. Note osteophytic lipping of the anterior-posterior margin

The remains of at least ten thoracic vertebrae are preserved, with the inferior-most two having osteophyte development. The remains of all five lumbar vertebrae were noted, with all expressing pathological changes. All have compression of the bodies and osteophytic lipping. Furthermore, an additional highly fragmentary lumbar vertebra was noted, likely due to lumbarisation of T12 having occurred. Given the highly fragmentary nature of this vertebra, it is possible that it is an additional lumbar or sacral vertebra, but lumbarisation is more likely (Barnes 1994: 78-9).

Eighteen rib fragments were associated with these vertebrae, of which seven could be sided (four left and three right). Lipping was found on many of the fragments, with osteophyte development inside the single costal end preserved. Unfortunately, very little pelvic material remains. The rest of the postcrania consisted of the body of the sternum, a fragmentary unsided glenoid fossa, a partial unsided humerus head and shaft, both right and left shafts and distal epiphyses of the ulnae, a fragmentary acetabulum, the medial portion of the distal articular surfaces of the left femur, the right patella and an unsided fibula midshaft. The teeth all exhibit low to moderate wear and the anterior central incisors exhibit an unusual wear pattern. This may be either accidental angled dental wear or deliberate shaping.

Age and Sex

Little suitable skeletal material for age determination was preserved. All teeth exhibited low to moderate wear, and hence this individual was provisionally classified as being a young adult through Brothwell's (1981) method.

Unfortunately, neither the cranium nor the os coxae was sufficiently complete to permit sex determination. Furthermore the postcranial material that was preserved was insufficient to allow metric sex determination methods. The maximum height of the right patella was 40mm, midway between white South African male and female values (Bidmos et al. 2005); hence this specimen remains unsexed.

Differential Diagnosis

The fusion and scoliosis in the cervical vertebrae may be caused by traumatic injury to the neck associated with healing, *spondylosis deformans*, Klippel-Feil syndrome or due to an error in segmentation leading to the development of block vertebrae. These will each be considered below.

Traumatic injury

Trauma was suggested by the complete fusion of the C2 and C3 vertebrae and the associated osteophyte development and macroporosity on the rest of the cervical vertebrae (Ortner & Putschar 1981). No complete fracture was found, therefore a potential traumatic injury would be infraction to either C2 or C3 with a prolonged period of bony growth. In order to form the scoliosis observed, the force required would need to be bending in nature and affect both vertebrae. It was not possible to X-ray the specimen and, from visual examination, no macroscopic fracture line was found. Furthermore no callus was noted. If fracturing had occurred, the episode must have occurred long before the death of the individual as the vertebrae were fully fused and healthy. Furthermore, for fusion to occur, the vertebrae would likely need to have become immobilised together. However, this diagnosis, potentially associated with spondyloarthropathy, is supported by the lower vertebral compression and osteophyte development. Traumatically-induced fusion of the vertebrae may occur as a result of compression injuries, such as those resulting from hyper-flexion of the neck (Resnick & Niwayama 1981). This potential cause of the palaeopathology is considered unlikely in the Nevisian population.

Spondylosis deformans

Spondylosis deformans may occur in any segment of the vertebral column. The disease is characterised by bridging osteophytes, arising from the junction of the body and the fused marginal vertebral plate which bulges at the level of the intervertebral disc. This is the most common degenerative spinal disease to affect males in greater proportion than females (Mann & Murphy 1990: 57). Over a prolonged period, remodelling may give the bridges a smooth appearance, thereby making the condition resemble ankylosing spondylitis¹ (Ortner & Putschar 1981: 421).

This diagnosis is supported by the proliferation of osteophytes elsewhere within the vertebral column, the macroporosity noted in the cervical vertebral bodies and the potentially age-related compression of the lumbar bodies. It is unlikely that the complete fusion would occur in only two vertebrae, especially C2 and C3,

¹ Ankylosing spondylitis itself was not considered a possible diagnosis as the condition starts in the lower back and sacroiliac region, and then moves up the vertebral column, whereas the fusion noted affects only C2 and C3.

without other fusion of vertebrae occurring unless this is an early stage. The diagnosis of *spondylosis deformans* is unlikely as there is no inflammation observable, and hence it would be required to be secondary to trauma or related to a bone-forming degenerative condition such as diffuse idiopathic skeletal hyperostosis (DISH) or ankylosing spondylitis.

Block vertebrae

The block formation of two adjacent vertebrae occurs as a result of incomplete segmentation of the vertebrae. The fusion may be a result of congenital absence of the intervertebral disc (Aufderheide & Rodríguez-Martin 1998: 62-3) due to caudal shifting during the sclerotome phase of development (Barnes 1994). Clinical incidence rates are reported to be 2-4%, with the most commonly affected vertebrae being in the cervical spine (Aufderheide & Rodríguez-Martin 1998) and lumbar regions. Complete unity is found between the centra and is associated with either complete or incomplete unity of the neural arches, although, usually the neural arches are affected (Barnes 1994: 66-7).

For this individual, this diagnosis is supported, as the vertebrae affected are C2 and C3, with no separation between the bodies and with associated fusion of the transverse processes and pedicles.

Klippel-Feil syndrome

This condition, also known as brevicolis (Kaplan et al. 2005: 573), consists of the “congenital fusion”² of two or more vertebral segments into a block vertebra with a single spinous process, neural arch and vertebral body. This is the result of block vertebrae forming due to segmentation failure in embryogenesis (Aufderheide & Rodríguez-Martin 1998: 60). The neck thus becomes shortened and has limited mobility (Barnes 1994: 67). The prevalence is less than 0.01%, and so thus may be considered as a specialised form of block vertebrae.

Three types have been defined (Barnes 1994: 69):

Type I – involving several cervical and thoracic vertebrae forming one grossly abnormal osseous block, Type II – involving only 2 or 3 vertebrae, with C2 and C3 being most common, and believed to be an autosomally recessive trait, and,

Type III – involving cervical vertebrae and associated with other segment errors in the thoracic and lumbar regions.

The type II form is the most common and is usually asymptomatic.

With this syndrome, the vertebral bodies become flattened or widened and the disc space is reduced. The

syndrome is also associated with scoliosis (especially of the cervical vertebrae), malformation of the occipital, elevation of the scapula, spina bifida, cleft palate, extra cervical vertebrae, fusion of the ribs, facial asymmetry and torticollis (Barnes 1994; Douglas 1991).

Klippel-Feil type II is a potential diagnosis of the pathology noted in the individual above. The specimen has C2 and C3 completely united, with associated scoliosis. Unfortunately, the occipital is very poorly preserved and so cannot be assessed. Only the portions of the left side of the maxilla are preserved and so cleft palate cannot be assessed. Only one fragmentary glenoid of the scapula and no sacral material were preserved, hence the other typically associated pathologies cannot be assessed.

Discussion

The evidence presented suggests that the upper cervical pathology described arose from type II Klippel-Feil syndrome as a result of block vertebral formation of C2 and C3. In living persons, individuals exhibiting Klippel-Feil syndrome are noted for having short necks, low posterior hairlines, limited neck mobility and occasional skin webs from the neck to the shoulder (Riseborough & Herndon 1975: 197). Although believed to be the result of an autosomally recessive trait (Barnes 1994: 69), potentially through the PAX1 gene locus on the long arm of chromosome 8 (McGaughran et al. 2003), Klippel-Feil syndrome is a heterogeneous disorder, exhibiting different expressions in different families (González-Reimers et al. 2001).

Archaeologically, Klippel-Feil syndrome has been noted among the Anasazi Pueblo groups (Barnes 1994: 69-71), individuals from prehistoric Peru (Ortner & Putschar 1981: 357), prehistoric Mexico, Ptolemaic or Late Period Egypt (Aufderheide & Rodríguez-Martin 1998: 60), prehispanic Canary Islanders (González-Reimers et al. 2001), and Austrian Magyar period groups (Pany et al. 2004).

The individual also appears to have had “lumbarisation” of T12. This is associated with cranial shifting during embryogenesis (Barnes 1994: 104-109), and may have a similar aetiology to the C2 and C3 block vertebrae.

The impact upon the individual’s life is hard to predict. As torticollis and facial asymmetry affect almost half of sufferers, it is possible that this individual looked different to other people in the population. If this is the case in the Coconut Walk individual, this may have led to the differential treatment by the population. Unfortunately, this hypothesis cannot be tested as the cranial remains were too fragmentary to study, and the specimen itself derives from a potentially disturbed context. Neurological and renal problems occur in many individuals with the disorder (Sullivan 2005), and hearing loss is common in about one third of sufferers (Hensiger et al. 1974; Kaplan et al. 2005: 574). It is, therefore, possible that this individual appeared in some

² This is not truly fusion of the two vertebrae as the segments never actually separated. For detail regarding embryogenesis of the vertebrae, see Barnes (1994), Kaplan et al. (2005) or Scheuer & Black (2000).

way different to the rest of the local population and hence may have acted or been treated differently.

Conclusion

An individual has been described who has severe pathological changes to the vertebral column, especially the upper vertebral column. These changes consist of “fusion” of C2 and C3, cervical scoliosis, increased cervical macroporosity and osteophytosis. These are associated with osteophyte development in the lower thoracic and lumbar regions and lumbarisation of T12. Although it is possible that this pathology results from traumatic insult, we argue that the most likely diagnosis is type II Klippel-Feil syndrome. This is an autosomally recessive trait, and thus, if found in other prehistoric Nevisian (or other Caribbean) islanders, may be used to aid in genetic population reconstruction.

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