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A Rare Existence of Significant Number of Wormian Bones in the Lambdoid Suture

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Abstract: ***Wormian bones may be defined as those accidental or intercalated bones of the cranium having no regular relation to their ossifying centres. They are associated with cranial and central nervous system abnormalities. The present case showed the presence of significant number of wormian bones (SNWB) 34 nos. – 11 on the left, 5 – in and around lambda and 18 on the right in an adult dry Indian skull. It also showed other significant features like denticulogenesis imperfecta and exaggerated occiput, predicting it to be a case of Osteogenesis imperfecta? Knowledge of these variations (SNWB) is very important for anthropologists, forensic experts, radiologists, orthopedic and neurosurgeons to avoid misleading for multiple fractures of the skull.***

Keywords: Wormian bones, Lambdoid suture, Lambda, Inca bone, Osteogenesis Imperfecta

1. Introduction

The Wormian bones or Sutural bones or Ossicles are islands of small bones found at the sutures and fontanelles of the skull, most commonly in the lambdoid sutures. (Sanchez-Lara, Graham Junior, Hing et al, 2007). They are named after the Danish anatomist, Olaus Wormius (Pryles and Khan, 1979). The alternative names of WBs include Schaltknochen (Leichner-Weil, 1964), intercalary, sutural and intrasutural bones. They are unnamed bones because they vary from person to person in number and shape. (Nayak, 2008)

Wormian bones are formations associated with alterations in the normal development of the flat bones of the skull, insufficient rate of suture closure and regarded as "epigenetic" and "hypostotic" traits (Barberini, Bruner, Cartolari et al, 2008). They may be formed by additional ossification centers. They may occur in or near the sutures. They articulate with the surrounding bones by sutures, the dentations of which are more complex on the external side than on the internal side of the skull.

The mechanism of formation of the wormian bones is not clearly known. Some believe they are due to external influences (Hess, 1946; Bennett, 1965; Finkel, 1971). Others state they are due to normal developmental processes and are genetically determined (Murphy, 1956; El-Najjar and Dawson, 1977; Pal et al, 1986).

An independent bone at the lambda is called "Inca Bone" or "Goeth's Ossicle". Rarely they may be seen at pterion (epipteric bone), at Bregma (OS Kerckring). The sutural bones are small and bilateral. The Inca bone was common in the skull of Incas (South American Indians) and is still present in their Andrean descendants (Standring et al, 2008). Wormian bones are found in both sexes in similar percentages as well as in both sides of the skull, being predominantly symmetrical (Jeanty et al, 2000).

The morphological knowledge of WBs is important in the diagnosis of CNS disorders (Cremin, Goodman, Spranger et al, 1982). Though the occurrence of WBs is quiet common, the observational data on them are poorly reported. It is important to know about these bones because they can mislead in the diagnosis of fracture of skull bones in medico legal cases.

2. Case Report

During the routine osteology demonstration classes for Dental undergraduate students, we found a series of Ossicles (Wormian bones) along the lambdoid suture (Figure 1-3). The skull has been photographed (Fig 1, 2 & 3) and radiographed (Figure 4, 5 & 6). These bones were found in an adult Indian skull.

In the present skull, there are about 34 sutural bones. The largest among them was at the lambda and the size of rest of the bones reduced progressively from lambda to asterion. Eleven bones were on the left side of lambda (Fig. 2), 5 were in and around lambda forming the Inca bone (Goeth's Ossicle) (Fig. 1) and the remaining 18 were on the right side (Fig. 3).

The lambdoid suture itself seems to be splitted up into two borders (upper and lower) enclosing the wormian bones in between these borders (Fig. 1). In between the wormian bones indentated bony spicules can also be visualized on the external aspect of the skull (Fig. 2 – Blue Arrow marks). There are 16 bony spicules seen towards the upper border and 9 towards the lower border.

The wormian bones vary in shape and size which has been tabulated in Table 1. The shape is predominantly irregular, then in order quadrangular, triangular, circular and linear (only one - not definitive of shape). All the 34 ossicles were measured of its maximum width and maximum height using a vernier caliper.

In addition to the above features in regard to wormian bones, the sagittal suture was very wavy and is almost obliterated, the tooth sockets are also resorbed, presence of ptergospinous bar (Fig 1 A and 4 A) indicating the ossification of the ligament stretching between the spine of the sphenoid and the lateral pterygoid plate. This would authenticate the age of the skull to be more than 45. It also showed exaggerated occiput which is quite obvious both

macroscopically and radiographically (Pink arrow marks - Fig. 2, 3, 5 & 6).

3. Discussion

It was opined that WBs can be found in healthy individuals as a normal variant (Burgener and Kormano, 1997). When WBs occur as a normal variant, they tend to be smaller and less numerous than when they are associated with skeletal dysplasias (Kaplan and Kemp, 1991). But most authors opine that they are not pathognomonic, as they occur in normal individuals, but the presence of more than ten sutural bones is unusual.

In the 16th century, the anatomists Andernach and Vesale were the first to associate wormian bones with cerebral disorders (Pryles and Khan, 1979). Usually not more than several are found in single individual but more than hundred have been found in hydrocephalic adult skull (Standring, 2008).

Although the exact mechanisms responsible for the formation of wormian bones are unknown, Wormian bones are the markers for various diseases and are important in the primary diagnosis of brittle bone disease, *osteogenesis imperfecta* (Glorieux, 2008). Wormian bones may also be seen in: *Pycnodysostosis, Osteogenesis imperfecta, Rickets, "Kinky-hair" Menke's syndrome, Cleidocranial dysostosis, Hypoparathyroidism and hypophosphatasia, Otopalatodigital syndrome, Primary acro-osteolysis, Down's syndrome.* These causes can be remembered by the mnemonic “PORKCHOPS” (26).

Skull growth is affected by dural attachments and is related to brain development. Presence of sutural bones is almost invariably associated with abnormal development of the CNS and may serve as a useful marker for the early identification and treatment of the affected infant or child (Pryles and Khan, 1979). According to these authors, the prevalence of central nervous system abnormalities in a population with wormian bones varies from 93% to 100% in a random group and reaches 100% in a mentally retarded population.

According to Bergman et al, (1988) nearly 40% of skulls have sutural bones in the vicinity of the lambdoid suture. The next most common site of WB is the pterion ossicle, which has a high incidence among Indians. Although they are most commonly found in the posterior sutures (lamboidal and occipito-mastoid sutures), they can occur in any cranial suture and fontanelles. They can develop either from independent ossification centers or by their separation from primary centers (Sanchez Lara et al, 2007).

Wormian bone along the lambdoid suture is more common. The sutural bone at lambdoid suture is also termed as preinterparietal bone or Inca bone. A possible explanation given to the occurrence of this bone in lambda is that the interparietal part of the squamous occipital bone above the highest nuchal line develops in membrane, usually from two pairs of centres (Pal et al., 1984).

Sreekanth et al, (2013) reported 14 WB in the lambdoid suture. There were six sutural bones on the left half and two sutural bones on the right half of the lambdoid suture. All the sutural bones were irregular in shape. There were six interparietal bones.

The present report is a rare case of the presence of Multiple WBs (34 nos) in the lambdoid suture not reported earlier in world literature, suggestive of any one of the above mentioned clinical conditions. Our uncertainty is that it may be a condition of Osteogenesis imperfecta due to the presence of multiple wormian bones, absence of teeth (denticulogenesis imperfecta) and exaggerated occiput. It may necessitate further investigations to identify an underlying pathology or hereditary disorder that has affected the skull growth at an early stage of development as suggested by Cremin, Goodman, Spranger et al, (1982).

The presence of a SNWB in OI is a well-documented fact. Semler stated that the more severe the manifestation of OI the higher the number of Wormian bones. These finds agree with those presented by Kovero et al, in 2006 where SNWB were recorded in 63% of adult OI patients.

Denticulogenesis imperfecta represents the disturbance in tooth formation associated with Osteogenesis imperfecta. The radiographic hallmarks of Osteogenesis imperfecta include osteopenia, multiple fractures and wormian bones in the skull. (Rajendran and Sivapathasundaram, 2012).

4. Conclusion

The Wormian bones were more frequent at the lambdoid suture. The clinical importances of these variant bones were emphasized with relevant review of literature.

The present case of series of wormian bones like this gives us an ambiguity of Osteogenesis imperfecta which has to be delineated. Other obscurity is that these bones may lead to problems in posterior approach to the cranial cavity.

The detailed knowledge of WBs is enlightening for the neuroanatomists, neurosurgeons, orthopedicians, radiologists, anthropologists and morphologists as these bones might lead to confusions in reading the radiographs in the case of head injuries. The Wormian bones like this may be mistaken for multiple fractures.

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Fig. 1- The Posterior View of the Skull showing Multiple wormian bones (1 -27) at the Lambdoid suture

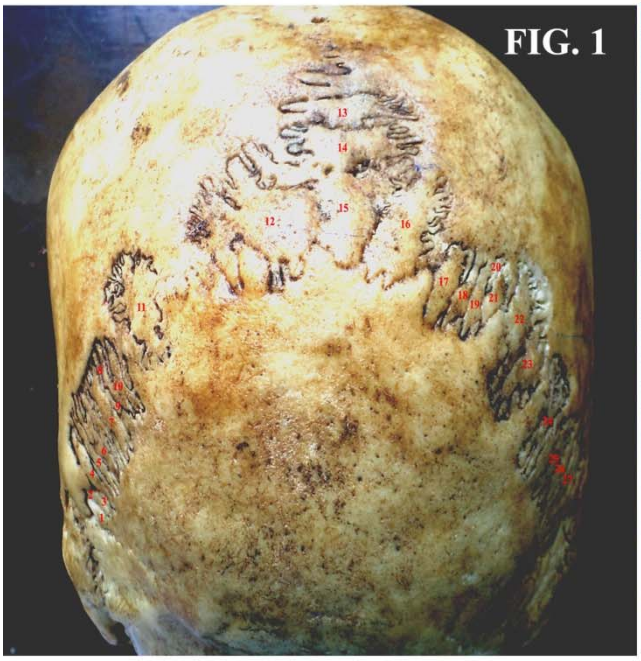


Fig. 1 A - Oblique view of the skull showing absence of teeth (Black arrow), presence of pterygoalar bar ((grey arrow) and SNWB (green arrow)

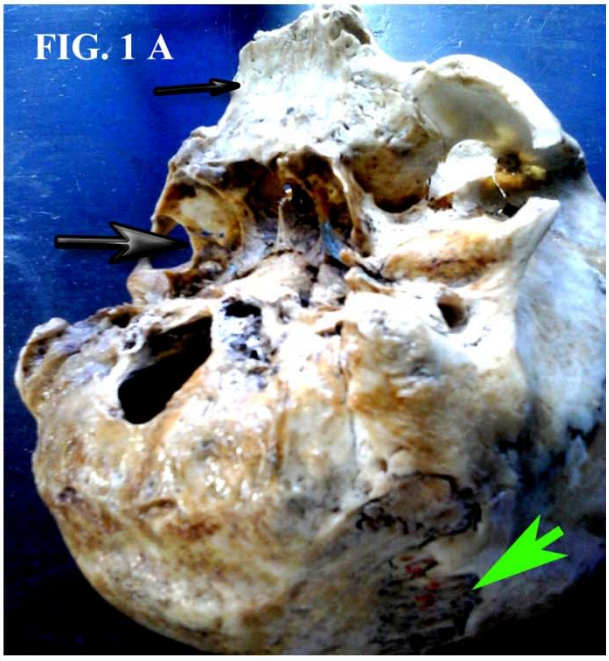


Fig. 2 - Left Lateral View of the skull showing multiple WBs (1--11)

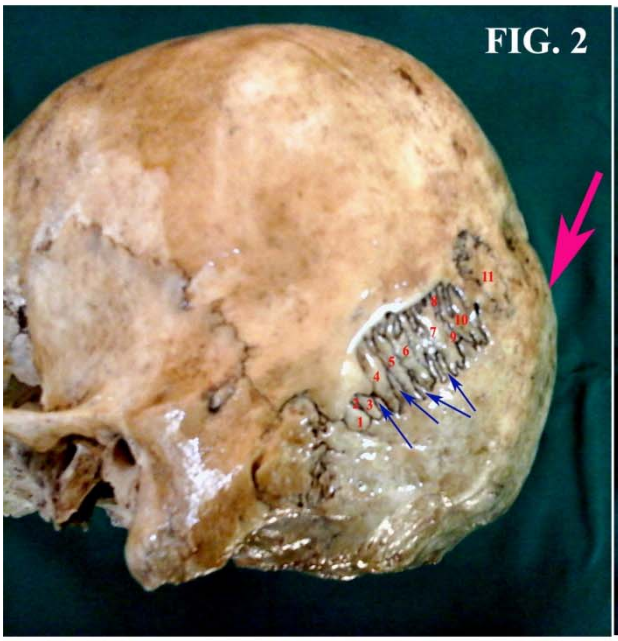
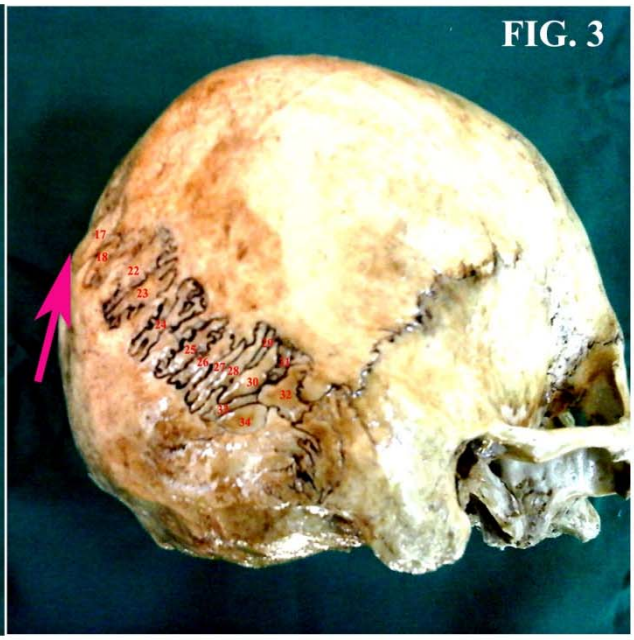


Fig. 3. Right Lateral View of the skull showing multiple WBs (17 --34)



Blue Arrow indicates the indentations between the Wormian Bones (WBs).

Pink Arrow shows the exaggerated Occiput.

SNWB - Significant number of wormian bones

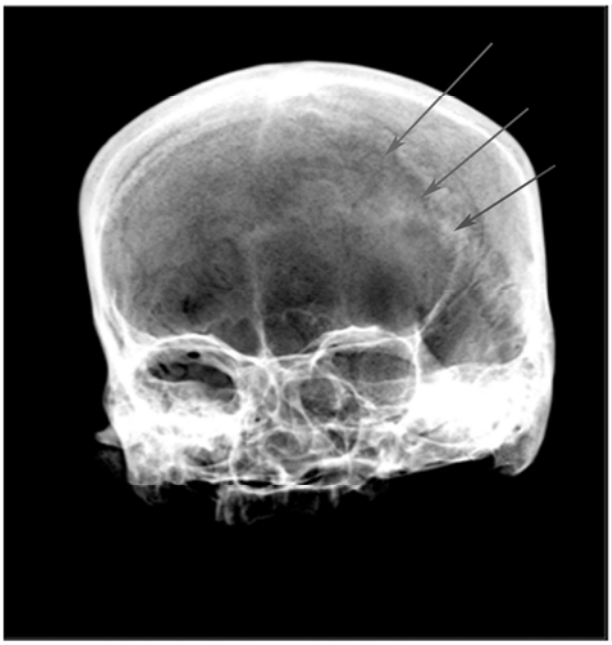


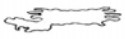
Fig. 4 - Anterior Radiographic view of the Skull showing the Multiple Wormian Bones (Arrow mark)

Fig. 4 A - Basal view of the skull showing absence of teeth (Arrow marks) and presence of pterygoalar bar (thin probe)

Fig. 5 - Left Radiographic View of the skull showing the presence of Multiple WBs (Arrow marks) and Exaggerated Occiput

Fig. 6 - Right Radiographic View of the skull showing the presence of Multiple WBs (Arrow marks) and Exaggerated Occiput

Table 1 - Various Shapes and Dimensions of the Wormian Bones (1 - 34)

WB.NO.	SHAPE OF THE WORMIAN BONES	MAX. WIDTH (cms)	MAX. HEIGHT (cms)
1.	 - Circular	0.5	0.8
2.	- Triangular	0.2	0.4
3.	 - Quadrangular	0.3	0.9
4.	 - Quadrangular	0.4	1.8
5.	 - Quadrangular	0.4	1.9
6.	 - Irregular	0.4	1.9
7.	 - Irregular	1	1.8
8.	- Triangular	0.2	0.7
9.	 - Quadrangular	0.2	0.6
10.	 - Irregular	0.6	1.7
11.	 - Irregular	0.9	1.9
12.	 - Irregular	2	1.3
13.	- Irregular	2.2	0.7
14.	 - Irregular	2.5	0.8
15.	 - Irregular	1.1	1.6
16.	 - Irregular	0.9	2.3
17.	 - Quadrangular	0.5	1.5
18.	 - Triangular	0.3	1.6
19.	 - Triangular	0.3	1.5
20.	 - Triangular	0.1	0.8
21.	 - Quadrangular	0.3	1.3
22.	 - Irregular	0.7	2.2
23.	 - Irregular	0.6	2
24.	 - Irregular	0.4	2.2
25.	 - Irregular	0.5	2.1
26.	 - Irregular	0.4	1.7
27.	 - Triangular	0.3	1.9
28.	 - Linear	0.3	2.4
29.	 - Triangular	0.1	1
30.	 - Oval	0.5	1.9
31.	 - Triangular	1	0.7
32.	 - Irregular	1.1	1.4
33.	 - Quadrangular	0.3	1.5
34.	 - Circular	0.6	0.9

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