Unidentified, multifocal joint disease from the Slovenian Kranj skeletal series

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Introduction

The Župna cerkev graveyard in Kranj contains more than 2100 archaeologically investigated burials from between the 7th and 18th centuries, which means that this is one of the largest medieval graveyards in Europe in terms of size and the duration of uninterrupted continuous use. Almost half of the graves belong to the Early Middle Ages. (Štular & Belak, 2013). From these, a total of 1169 skeletons (285 subadults, 406 female and 478 male) were available for detailed anthropological analysis. A large number of skeletons (n=50; 20 males, 28 females, and 2 subadults) exhibit irregular shaped lytic lesions varying in size from 5-20 mm on the joint surfaces. The lesions are polyarticular, generally bilateral, and lytic with no new bone formation, and no periosteal reaction.

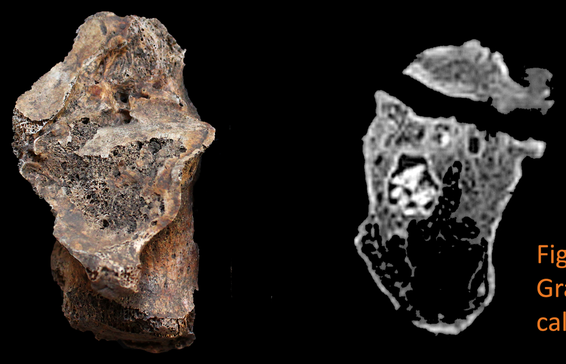


Figure 1.
Grave No. 358, right calcaneus

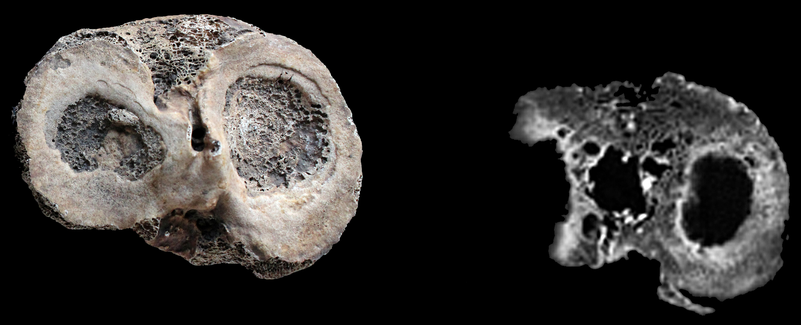
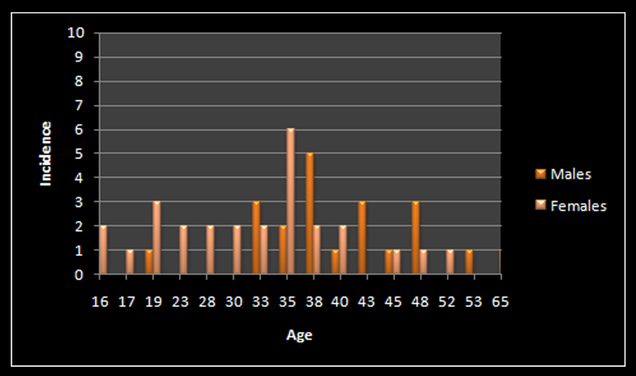


Figure 2.
Grave No. 359, both proximal tibiae.

Results

Age distribution of the individuals affected with the disease.



Frequency and distribution of the affected joints.

	Males	%	Females	%	Total	%
Knees	20/28	71.4	40/43	93.0	60/71	84.5
Hands	9/18	50.0	20/28	71.4	29/46	63.0
Ankles	17/29	58.6	20/32	62.5	37/61	60.7
Feet	17/23	73.9	14/29	48.3	31/52	59.6
Elbows	7/19	36.8	24/36	66.7	31/55	56.4
Hips	9/26	34.6	19/39	48.7	28/69	43.1
Wrists	5/18	27.8	4/11	36.4	9/29	31.0
Shoulders	2/18	11.1	5/25	20.0	7/43	16.3

The grouping of individuals affected by the disease in the cemetery does not appear to be random and may reflect a genetic component to the disease (Pleterski & Štular - personal communication).
The lesions are polyarticular, generally bilateral, and lytic with no new bone formation, or periostesal reaction.
Most commonly affected are the knees, hands, ankles, feet, elbows, hips, wrists and shoulders. The lesions were more frequent in females than males (28/406 or 6.9% compared to 20/478 or 4.2%), giving a male/female ratio is 1:1.6, which is marginally not siginificant (χ^2= 3.145, P=0.07616). The highest frequencies in females are in the knees (93%) and hands (71.4%), while in males the lesions are most commonly found in the feet (73.9%) and knees (71.4%). In terms of the age distribution of the disease the affected individuals cover all age groups, the youngest being a subadult of 7-8 years at time of death, while the oldest is a female older than 60 years.
Although, most of the lesions could be observed macroscopically, radiographic images revealed numerous cavities and subchondral cysts within the trabecular bone, which in some cases exhibit a calcified formation (Figures 1-2). Radiographic analysis was carried out using MDCT unit (Emotion 16; Siemens Healthcare, Erlangen, Germany). Isometric 1.5 mm slices were obtained using 16x1.2 mm with reconstruction increment 1.5. 3D maximum intensity projection (MIP), volume rendering technique (VRT) and multiplanar reconstructions (MPR) provided more details of the bone cavities.

Conclusion

At this moment the aetiology of the disease is unclear. The lack of osteophytes, peri-articular erosions, osteoporosis, sclerosis of the affected part of the joint and bony ankylosis rules out more recognizable joint diseases such as rheumatoid arthritis, juvenile idiopathic arthritis, psoriatic arthritis, Reiter's syndrome, osteochondritis dissecans, and gout.

Differential diagnosis

	Rheumatoid arthritis	Juvenile idiopathic arthritis	Psoriatic arhritis	Reiter's syndrome	Gout	Osteochondritis dissecans	Multicentric reticulohistiocytosis
Age of onset	20-50	<16	20-50	20-40	50+	10-40	During the fourth decade of life
Sex predilection	Females	Females	Females	Males	Males	Males	Females
Hallmark lesion	Marginal bone erosion of the joint	Epiphyseal overgrowth in knee	Erosion of distal intherphalangeal joints	Overhanging edge lesions	Eburnation of the joint and osteophytes at joint margin	Separation of articular cartilage and subchondral bone fragment	Sharply demarcated marginal erosions
Main location	Small joints of hands (IP&MCP)	Large joints (knee), cervical spine	Distal IP joints of hands and feet	Joints of lower limb	MTP joint of the great toe	Knee	Distal IP joints
Other joints	Knee, foot, wrist, elbow, shoulder	Wrist, ankle, carpals, tarsals	Knee, ankle, foot	Feet, hands, wrist, elbow, knee	Any joint	Elbows, ankles, other	Knee, foot, wrist, elbow, shoulder
Number of joints involved	Multiple	One or few	Few or multiple	Few	Usually only one	One, few, multiple	Multiple
Symmetry of the lesion	Symmetrical	Asymmetrical	Asymmetrical	Asymmetrical	Asymmetrical	Symmetrical	Symmetrical

*table partially modeled after Rajić Šikanjić and Vlak 2010.

Due to the fact that the lesions are mostly bilateral, most frequent in hands and knees, include severe joint destruction, sharply demarcated marginal erosion, and the absence of osteopenia, osteoporosis, ankylosis, and periosteal reaction the proper diagnosis for the disease affecting these individuals may be multicentric reticulohistiocytosis, also known as lipoid dermatoarthritis, which is a rare, systemic histiocytic polyarhritis disease that affects the skin and joins (Shah et al.). If so, this would be the first example of this disease in an archaeological series.

Figures 3-4.
Grave No. 397, phallanx and capitate.

Figure 5.
Grave No. 349, distal tibia.

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