Classification of non-bacterial osteitis: retrospective study of clinical, immunological and genetic aspects in 89 patients.

OBJECTIVE: To define non-bacterial osteitis (NBO) as a clinical entity possibly associated with autoimmune manifestations. Patients with sterile osteitis were analysed to develop diagnostic criteria. METHODS: A total of 89 patients with non-bacterial inflammatory bone lesions were observed for a median of 49 months. History, diagnostic imaging, laboratory and histological data were obtained. Mutation analysis in the genes PSTPIP1 and PSTPIP2 was performed. RESULTS: Patients had an onset of disease at a median age of 10 yrs [interquartile range (IQR) 7.5-12] and suffered a median period of 21 (IQR 9-52) months with a median of three foci per patient. Twenty percent of all the patients demonstrated associated autoimmune disorders, particularly of the skin and bowel. The majority of bone lesions were located in the vertebrae and metaphyses. Slight-to-moderate elevation of inflammation values were found in all the patients and antinuclear antibodies were elevated in 30%. Non-steroidal anti-inflammatory drugs (NSAIDs) were effective in 85% of the patients. HLA-B27 and Human Leukocyte Antigen-DR (HLA-DR)-classification did not differ from the general population. Autoimmune diseases in 40% of all the families, multiply affected family members, linkage to 18q21 and mouse models strongly...
indicate a genetic basis for NBO. We observed three different courses of disease regarding the
duration of complaints, rate of complications and associated autoimmune manifestations leading to a
new classification of NBO. CONCLUSIONS: Clinical analysis of our cohort leads us to define NBO as
a distinct disease entity with three clinical presentations: acute NBO, chronic recurrent multifocal
osteomyelitis or persistent chronic NBO. Diagnostic criteria were proposed to differentiate NBO from
diseases with similar clinical presentation.