Chronic nonbacterial osteomyelitis in childhood: five years’ experience of imaging at a tertiary hospital

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Objective

- Chronic nonbacterial osteomyelitis (CNO) is a relatively rare autoinflammatory bone disease of unknown etiology characterized by sterile bone inflammation.
- Chronic recurrent multifocal osteomyelitis (CRMO) is the most severe form of CNO.
- CNO mainly occurs in children and adolescents, predominantly affects the metaphyses of the long bones, but single or multiple lesions can occur at any site of the skeleton.
- Delays in diagnosis may lead to prolonged courses of antibiotics, unnecessary radiation exposure from multiple radiographs or bone scans and repeated surgery including bone biopsies.
- The aim of the study was to determine patient characteristics, clinical presentation, pattern of bone involvement and imaging strategies of patients with CNO.

Methods

- We did retrospective analysis of the electronic medical records of children diagnosed with CNO at the Tartu University Hospital Children’s Clinic and department of Maxillofacial Surgery from 2012 to 2017.
- The complete record of each patient — including demographic, clinical, laboratory and histological data — was reviewed.
- All performed images were analysed for the presence and number of bone lesions.
- To classify patients the CNO clinical score proposed by Jansson et al. in 2009 was applied.

Results

- Patients characteristics (Table 1).
  - A total of nine children, five boys and four girls, with CNO/CRMO were enrolled in the study.
  - The median age at the onset of first symptoms was 11 ± 3.9 (range 5.3-17.1) years and the median age at diagnosis was 12.4 (range 5.5 – 17.5) years. The median delay to the CNO diagnosis was 5 (range 1-17) months.
  - There was not significant difference in the median diagnostic delay between multifocal and unifocal involvement groups.
  - The median time to diagnosis of the first two patients diagnosed in 2012 was 15 months versus 3.9 months in other seven patients diagnosed in later years what confirms importan change in the diagnostics skills and knowledge.
  - 56% of patients with unifocal disease and/or mandibular involvement were biopsied and in all cases biopsy confirmed the diagnosis of CNO.
- Clinical presentation.
  - All patients complained about pain, six had localized swelling and only one presented with fever. One child had associated Crohn’s disease, none of the patients had pusulosis. Five patients presented with clinically unifocal disease but two of them had radiologically more than one lesion. Median CNO clinical score at the diagnosis was 47 ± 10.
- Skeletal involvement according to MRI and radiographs is summarized in the Table 2.
- Imaging strategies. Although in some cases also CT, bone scintigraphy and PET-CT was done, in this study we reviewed only radiographs which are usually the first radiological assessment tool in everyday practice, and targeted MRI and WBMR images. For follow-up of patients with multifocal CNO whole body MR was used.

Conclusions

- In the recent years CNO has been more recognized and diagnosed in our institution and mean time to the CNO diagnosis has decreased.
- Symmetry, multifocality and particularly specific patterns of lesions appear suggestive to CNO.
- For imaging of CNO MRI should be preferred instead of CT and nuclear imaging.
- WBMR is a valuable and radiation-free imaging modality of choice in patients with clinical multifocal symptoms and in detection of subclinical lesions.

Key words

Chronic nonbacterial osteomyelitis, CNO, chronic recurrent multifocal osteomyelitis, CRMO, whole body MR, WBMR