secondary subacute arthritis in children with diastrophic dysplasia and RDMD

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Background: Progressive osteoarthritis is common in skeletal dysplasias. In the most of these disorders progress is relatively slow and acute pain with severe disability secondary to osteoarthritis is rare during childhood. Diastrophic dysplasia and RDMD are quite unique among other skeletal dysplasias for secondary subacute arthritis (SSA). Protracted course of SSA can mimic JIA.

Objectives: To analyse incidence and features of SSA in group of patients with DD/RDMD.

Methods: We retrospectively analysed for SSA our series of 39 patients with DD/RDMD. Clinical, radiological and laboratory data were collected. Clinical assessment included pain (VAS), ROM (range of motion) and posture/walking disturbances. Radiological type by Yasunaga classification, neck-shaft angle (NSA) and articulo-trochanteric distance (AT distance) were estimated. MRI data regarding cartilage damage, joint effusion and bone marrow condition were noticed. Biomarkers of inflammation and immune response (ANA, RF, vimentin antibodies) were assessed in blood samples.

Results: We identified 17 patients with hip joint SSA among 39 patients with DD/RDMD (43%). Bilateral involvement was identified in 13 children with asynchronous appearance in all cases (from 2 to 18 months before the symptoms on the other side). Trauma (including iatrogenic damage during physiotherapy) preceded SSA in 9 cases. Pain, limited range of motion, limping and antalgic posture throughout the day were noticed in all the cases. Duration of these symptoms was from 4 weeks to 9 months. Progressive phase (increasing symptoms) took from 2 weeks to 4 months. General laboratory data were normal or indicated moderate inflammatory response without any specific changes. Radiological data shows predictive signs like NSA<110°, AT distance <10mm, fair or poor congruence by Yasunaga classification. Also radiological data demonstrated progressive subluxation with narrowing of articular space, osteoporosis, subchondral cysts.

MRI revealed pseudo-erosive damaged cartilage in contact area with joint effusion and bone marrow oedema. Total cartilage matrix also visualized like intermittent and multi-layered = erosive-like chondrolysis (defective articular hyaline cartilage) in 11 patients (65%).

Management included bed rest or partial weight bearing. NSAID were prescribed for 2-4 weeks with following usage according to the symptoms. Physical therapy to maintain range of motion was provided. One patient was operated (containment surgery for subluxation). In follow-up (2-7 years) 12 patients were painless at the daily life. 4 patients had intermittent pain and one patient (operated) - daily moderate pain. Range of motion was restored in 11 patients. Those patients who underwent in-hospital rehabilitation immediately after appearing of the symptoms demonstrated better outcome.

Conclusion: SSA is typical for DD/RDMD and leads to remarkable disability. Early recognition and non-surgical management are important for recovery. The quick reversibility of the clinical picture arthritis against the background of a short course of NSAIDs and rehabilitation is a characterized of non-autoimmune secondary inflammatory process (SSA). SSA not requires anti-rheumatic treatment in children with DD/RDMD.

References:

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