Pediatric Rheumatology

Poster presentation

Polyarteritis Nodosa (PAN) in childhood: a report of two siblings with intractable disease controlled by mycophenolate mofetil (MMF)

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Background

PAN is a systemic vasculitis with multisystem involvement rarely observed in childhood. We report two siblings (1 boy, 1 girl) with aggressive PAN who had a persistent sustained improvement with MMF.

Patients

The boy developed at 8 yrs musculoskeletal pain and painful nodules on the legs, than vertigo, tinnitus, and diplopia, neurosensorial ipoacusia, left central facial palsy and hypertension. Angio MRI revealed ischemic alterations on Willis's circle and prompted to diagnose PAN. Despite aggressive therapy the boy developed ischemic lesions of 3 digits of hands and Iloprost was introduced; over a short time multiple ulcerative deep cutaneous lesions appeared on the legs. Due to persistent active disease, MMF (2 g/day) was started and up to now the clinical symptoms are stable and laboratory work up normalised.

The girl, at 30 months had neurological manifestations (head and eye rotation with no consciousness, optical bilateral neuritis); over time similar episodes recurred and periodic ataxia was diagnosed. Cerebral MRI showed areas of hyperintensity on the thalamus. At 7 yrs maculopapular rash on the face and upper extremities, arthralgia/mialgia and hypertension were complained. Skin biopsy confirmed necrotizing vasculitis of medium sided vessels and PAN was diagnosed. Deep cutaneous ulcers appeared on the legs. After a severe ischemic attack, confirmed by MRI lesions at the pons area, MMF was started. Since then the girl is stable.

Conclusion

MMF used in the treatment of SLE and primary vasculitis in children, should be considered as either alternative or adjunctive therapy in intractable severe persistent active PAN.