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AUTOIMMUNE DEAFNESS ASSOCIATED WITH NON-SYPHILITIC INTERSTITIAL KERATITIS IN PEDIATRICS

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BACKGROUND

About 250 cases of Cogan's syndrome were reported. With no preference for sex or age, most diagnosed in average age at 29, with no known etiology and no specific treatment, classified as typical and atypical according to the place and time of appearance of the symptoms and systemic manifestations.

CASE REPORT

Female, 12years old, previously healthy. She had acute abdominal pain associated with vomiting lasting 3 weeks. Ocular hyperemia and mild bilateral itching, painless in improvement with eye drops, but recurring. Evolved with otalgia bilateral, dizziness, imbalances and decreased hearing acuity, social withdrawal with behavioral change. Three months after she started daily fever (T. axillary 39 °C), 1 peak per day; weight loss of 5 kg in 6 months, fatigue, asthenia, polyarthralgia of large and small joints, symmetrical, additive, without phlogistic signs or morning stiffness.

Worsened neurosensory symptoms and toxemic constitutional symptoms. Sequential audiometry with progressive hearing loss from moderate to bilateral severe. In ophthalmological evaluation in the presence of red eye: interstitial keratitis in both eyes and episcleritis in the left eye. Laboratory tests: CRP: 240 mg/L, ESR 130 mm/h, Hb 9.2 g/dL (normocytic and normochromic). Normal leukometry. Platelets: $695 \times 10^3/\text{mm}^3$.

Discarded processes neoplastic and infectious (central nervous system and hematological, syphilis, HIV, rubella, toxoplasmosis, cytomegalovirus, tuberculosis). Rheumatological investigation presented autoantibodies negative (antinuclear antibody, rheumatoid factor, anti-Ro, anti-La, anti-Sm, anti-dsDNA, lupus anticoagulant, anticardiolipins, antibodies neutrophil cytoplasm, direct antiglobulin test); add-ons normal serum. Normal echocardiogram.

The hypothesis of Cogan's syndrome was made. Oral prednisolone 1 mg/kg/day with improved pattern behavioral and auditory acuity, resolution of fever, ocular, articular as well as platelets and anemia, and decreased inflammatory tests. However, after starting to reduce the dose of prednisone, observed worsening of hearing acuity that had stabilized. Indicated a pulse therapy of methylprednisolone 1 g/day for 3 days followed by prednisolone 1 mg/kg/day and methotrexate (9.06 mg/m²/week), bilateral tympanic infiltration with 1.5 mg of dexamethasone.

Once again, she relapsed and recurred from audiovestibular symptoms on the end of corticosteroid therapy, even when methotrexate is in effect. A new pulse therapy with 1 g/day of methylprednisolone for 3 days was indicated followed by 1 mg/kg/day in oral prednisone and infliximab 6mg/kg/dose; doses at weeks 0, 2 and 6, and maintenance with doses every 8 weeks, with clinical improvement and drop in inflammatory tests.

CONCLUSION

This work reports a rare case of typical Cogan's syndrome, poorly described in pediatric population, refractory to high doses of corticoid with stabilization after infliximab introduction.