Pediatric Rheumatology

Poster presentation

Behçet Syndrome and Hypogonadotropic Hypogonadism: case report

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Background

Behçet Disease (BD) is a sistemic chronic vasculitis

Clinical case

A sixteen year old boy presented, three years before, gastrointestinal (abdominal pain, vomiting, diarrhea) and neurologic symptoms (headache, diplopia, ataxia, VI nerve paralysis), with fever, oral and genital aphtosis. Laboratory tests showed increased ESR, CRP). He carried B51HLA. Physical examination showed svere obesity, pseudomicropenis, rare pubic hair, pubertal stage G2P2. Thus BD was diagnosed. Treatment with corticosteroids and antiplatelet agents was started.

After 2 months, azathioprine and infliximab were added; but biological agent was stopped, because of adverse reactions; thus thalidomide was started. After the diagnosis, the patient presented a progressive weight increase (BMI 39) and a delayed puberty. He presented acanthosis nigricans, pseudoginecomasty, pseudomicropenis with testicular volumes of 2 mL, bone age was of 14.5 years (vs 15.6 cronologic age). The boy underwent several endocrinological tests that showed a short increase of GH to glucagon and clonidine Tests, low testosterone levels, a short increase of gonadotropins to LHRH test, normal answer to IGF-I generation test, normal glucidic tolerance with severe and prolonged hyperinsulinism.

Conclusion

We suggest that the boy had presented a neurologic involvement, that could explain his hypogonadotropic hypogonadism. We performed a cerebral MRI and a dosage of anti-hypophysis antibodies, which are normal.

Although no cases of BD and hypogonadotropic hypogonadism have been described, we report this case for the numerous patogenetic hypotheses that it could generate.

References

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