

Poster presentation

A phenotypic shift after initiation of IL-1 receptor blockade in a boy with systemic juvenile arthritis

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Case presentation

A boy with systemic juvenile arthritis (sJIA) for 7 years, treated with prednisone 1 mg/kg/day, NSAIDs, Methotrexate and TNF- α antagonism, presented at age 15 with severe incapacitating back pain.

Clinically, a cachectic and stunted boy, weight and height SD scores were -5,05 and -3,65 respectively. He had fever, anaemia, splenomegaly and polyarthritides. There was complete immobility and severe pain of his lumbar spine. X-rays revealed compression fractures of all lumbar vertebrae. Bone mass densitometry confirmed severe osteoporosis (Z-score -4.5 SD). ESR: 33, Hb: 7.4 g/dl, CRP 112 mg/dl.

IL-1 receptor antagonism resulted in a rapid major improvement. Within a few weeks, there was no fever, no arthritis, no splenomegaly, with normal CRP, ESR and Haemoglobin.

Body composition changed dramatically; weight increasing by 10 kg (SDS -2,9) and body fat fraction (DEXA) from 17% to 35%, despite weaning steroids from 1 to 0,3 mg/kg/day. Bone Z-score improved to -3,3 SD.

Discussion

The present case illustrates the striking effectiveness of IL-1 receptor antagonism in a patient with severe refractory sJIA, and steroid dependency (with devastating side effects, esp on bone mineralization). IL1-ra has been associated with obesity in adults [1,2]. IL1 receptor antagonism was associated with an important change of body

composition in this patient. It remains to be clarified how the interplay between inflammation, IL-1 blockade and corticosteroid therapy impacts on body composition and fat metabolism.

References

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