Pediatric Rheumatology

Poster presentation

Efficacy and safety of thalidomide in two sisters with severe refractory polyarteritis nodosa (PAN)

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from 15th Paediatric Rheumatology European Society (PreS) Congress
London, UK. 14–17 September 2008

Published: 15 September 2008

Pediatric Rheumatology 2008, **6**(Suppl 1):P267 doi:10.1186/1546-0096-6-S1-P267

This abstract is available from: http://www.ped-rheum.com/content/6/S1/P267

Background

Polyarteritis nodosa (PAN) is a necrotizing vasculitis seldom reported on childhood, with a severe prognosis.

Patients

We report two sisters (16 and 8 years old) with PAN refractory to different drugs who had a persistent improvement over 5-years of thalidomide.

In the older, the presenting symptoms were fever and migrant arthralgias. At onset, she was 6 year old. Laboratory work-up showed increased ESR, CRP, PTL count, and anaemia. Over time she developed ischemic colitis and a transient cerebral ischemic attack. Cerebral Angio MRI revealed multiple periventricular hyperintensities prompting the diagnosis of PAN. Steroids, cyclophosphamide, methotrexate, and azathioprine were unsuccessful, and the disease was complicated by neuropathy and ischemic lesions of several digits. Thalidomide was started in December 2003; since then, the disease is stable and laboratory work up normal. First clinical features of the younger sister occurred at three years of age. She had an acute stroke. Following hypertension, renal disease and neuropathy, PAN was diagnosed. Steroids, azathioprine and cyclophosphamide were given with scarce benefit. After another ischemic cerebrovascular event, thalidomide was effective in controlling the disease activity.

No side effects have been observed.

Conclusion

Both our patients refractory to steroids and immunosuppressants had a significant improvement when thalidomide was introduced. No flares have been observed during a 5-year follow-up. Although the exact mechanism of action is not fully understood, thalidomide mainly prescribed in connective diseases, could be considered an alternative therapy in severe PAN.

References

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