Pediatric Rheumatology

Poster presentation

Erdheim-Chester disease in children: clinical, radiologic, treatment characteristics of three cases

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Background

Erdheim-Chester disease (ECD) is a rare non-Langerhans cell histiocytosis (NLH), characterized by bilateral metaphyseal sclerosis of long bones and visceral infiltration. The histopathological hallmark is a xanthogranulomatous infiltration of foamy CD68+ CD1a- histiocytes and Touton giant cells. Only five paediatric cases have been reported.

Materials and methods

We contacted all paediatric rheumatology and oncology centers in France. Three cases of ECD have been identified. We describe the clinical, radiological, histological characteristics and treatments.

Results

All patients were female. Mean age at onset was 5 years (patient A: 17 months, B: 7 years, C: 6 years). Mean diagnosis delay was 5 years. Initial presentation included fever, bone pain (A and B), diabetes insipidus and growth hormone deficiency (C) with sinus infiltration. B and C developed retroperitoneal fibrosis with hydronephrosis and renal failure in C. Patient A subsequently developed exophtalmos. ECD was evoked on typical skeletal radiographies (A and B), and on retroperitoneal fibrosis on RMI (B and C). ECD was confirmed histologically on biopsies (retro-orbital/sinusal infiltration and bone). All patients received corticosteroids, alone (B) or combined with chemotherapy (A and C), which was partially and temporally effective. INFα was then used in all patients leading to regression of lesions (B), stabilisation of disease (C) and decrease of bone pains (A).

Conclusion

Although exceptional, paediatric ECD can be evoked in case of NLH with long bone osteosclerosis and/or visceral involvement. Prognosis depends of disease localisations and their consequences. Treatment needs to be considered case by case with special interest in IFNα.