Pediatric Rheumatology

Poster presentation

Case of vasculitis and sarcoidosis in a child with cystic fibrosis

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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):P272 doi:10.1186/1546-0096-6-S1-P272

This abstract is available from: http://www.ped-rheum.com/content/6/S1/P272

The occurrence of cystic fibrosis (CF) and sarcoidosis is not previously reported in childhood. Hypergammaglobinaemia and immune complex deposition may contribute to granulomata formation in sarcoidosis.

Following diagnosis of cystic fibrosis at 3 months old and despite standard CF management of daily nebulized colomycin, cefaclor, intravenous colistin and tobramycin this female patient became colonized with pseudomonas and by mid childhood had impaired pulmonary function with FEV1 67%, FVC 81% predicted. At 11 yrs, she developed episodes of rash, joint swelling and pyrexia.

Skin biopsy: florid vasculitis affecting small and medium sized vessels. Colchicine 500 mg tds initially helped but aged 12 yrs, her renal function fell acutely with serum creatinine 197, urea 19.3, ACR of 20, and calcium 4.3. 1,25 OH Vitamin D levels high at 120 mmol/l and angiotensin converting enzyme (ACE) raised at 137 mmol/l.

Renal biopsy: extensive interstitial nephritis with multiple foci of non-caseating granulomata and multinucleated giant cells.

Renal angiography: small vessel vasculitis without microaneurysms.

She had episodes of red, sore eyes.

Conjuctival biopsy: non-necrotising epithelial granuloma.

Sarcoidosis was diagnosed.

A high resolution CT scan of the chest showed CF changes only.

The hypercalcaemia and ACE responded rapidly to methylprednisolone. Her lung and renal function improved on maintenance azathioprine and prednisolone. By 6 months she was unable to wean steroids adequately without flaring.

A report of mycophenolate use in a post-transplant adult cystic fibrosis patient encouraged us to try mycophenolate instead and she now remains in remission with improvement of her cystic fibrosis lung disease as well.