A CURIOUS CASE OF AUTOIMMUNE HEPATITIS AND ACQUIRED PARTIAL LIPODYSTROPHY

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Background The lipodystrophies are an extremely rare group of metabolic conditions. In acquired partial lipodystrophy (APL), the predominant clinical feature is a progressive, symmetrical reduction of adipose tissue which typically begins during the pre-adolescent period. While the pathogenesis of APL is likely to be complex and heterogeneous, a subset of these patients are reported to develop associated autoimmune conditions, including hepatitis. This has led some to implicate an underlying autoimmune process.

Method We report an ultra-rare case of a 17-month-old boy who presented to hospital with a Henoch-Schönlein Purpura-like illness and subsequently developed anti-LKM-1 positive autoimmune hepatitis (AIH) as well as clinical features of Acquired Partial Lipodystrophy.

Results In this child, the features of lipodystrophy occurred at a very early age and in association with a relatively mild hepatitis. We document his challenging path from first presentation, through to the diagnosis of AIH and APL and discuss his ongoing management. Despite an encouraging reduction in liver transaminases following dual immunosuppression, no improvement was observed in his adipose tissue distribution.

Conclusions Cases of autoimmune hepatitis occurring in association with acquired partial lipodystrophy are extremely rare. The primary aim of this report is therefore to familiarise the PGHAN community with this uncommon association. While much is still unknown about the link between these conditions, we use this case to discuss the current evidence for their shared pathogenesis.

A DIAGNOSTIC DILEMMA: CASE REPORT OF A YOUNG BOY WITH ABDOMINAL TUBERCULOSIS WHO WAS INITIALLY THOUGHT TO HAVE CROHN’S DISEASE

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Introduction Tuberculosis (TB) like Crohn’s disease can affect any part of the gastro-intestinal (GI) tract including anus, peritoneum and hepatobiliary system. The clinical manifestations of abdominal tuberculosis are non-specific and can mimic various GI disorders especially Crohn’s disease which can cause delay in diagnosis and management.

History and Presentation A 14-year-old boy was diagnosed with small bowel ileal Crohn’s disease in 2016 based on clinical symptoms of abdominal pain and weight loss, biochemical features of a raised ESR but normal CRP at presentation and a distorted ileocaecal valve (ICV) with inflammatory changes seen both macroscopically and microscopically at colonoscopy with radiological confirmation of short segment ileal disease on MRI. He was treated with exclusive enteral nutrition for induction of remission, however his ESR remained elevated and he required escalation to Azathioprine within 3 months of diagnosis for continued symptoms of abdominal pain and ongoing weight loss. His clinical course over the next 2 years remained unchanged with a persistently raised ESR and continued disease around ICV and distal ileum in spite of immunomodulator therapy.

Treatment and Investigation Prior to commencing biologic treatment for active Crohn’s disease, he had an Elispot and was found to be positive. This was felt to be consistent with latent TB infection for which he had 3 months of chemoprophylaxis with Rifampicin and Pyridoxine. Following this, his symptoms of abdominal pain resolved, and he gained 5 kg for the first time since his diagnosis of CD. Moreover, his ESR completely normalised. His repeat mri showed a significant improvement of the inflammation in the ileum as well as around the ICV. This was also confirmed with repeat colonoscopy which was markedly improved from previously although still had abnormal distortion of the ICV. His clinical response to the TB treatment and radiological and endoscopic improvement following the TB chemoprophylaxis led to the suspicion of intestinal TB as the correct diagnosis.

Clinical Background and Progress

He was born in the UK. He had a BCG scar. His grandmother was diagnosed with TB in India in 2010. She had visited the UK prior to the diagnosis and stayed with the family for 6 months. She was unwell with cough and weight loss at that time. Both his mother and father had been exposed to her also and his father was also receiving treatment for latent TB now. Based on the history of TB exposure and the clinical, biochemical, endoscopic and radiological improvement following latent TB treatment, he went on to complete a full 6-month course with 4 drug initiation for abdominal TB.

Summary and Conclusion Abdominal tuberculosis should be considered as a differential diagnosis in patients with Crohn’s disease. Careful evaluation of clinical, biochemical, radiological and histological findings can aid in distinguishing between the two conditions, leading to early diagnosis and management.

A SERVICE EVALUATION OF A DIETETIC-LED CLINIC FOR THE MANAGEMENT OF PAEDIATRIC PATIENTS WITH COELIAC DISEASE

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Introduction The management of Coeliac disease (CD) is through a gluten free diet (GFD). The current BSPGHAN guidelines advise that annual follow-up with both a paediatric dietician and paediatric gastroenterologist is necessary to ensure continued adherence to a GFD.

However there are many NHS trusts where paediatric patients with CD are only reviewed in a dietetic-led clinic (DLC) annually.

Aims and Objectives To evaluate a DLC for the management of paediatric patients with CD two years after its establishment. In the current service patients are reviewed on an annual basis by the dietitians after an initial post-diagnosis appointment with a paediatric gastroenterologist and if identified as requiring a medical review through the ‘red flag’ criterion they will be referred back to the paediatric...