CHARACTERISING THE DISEASE PRESENTATIONS OF LCH OVER 10 YR AT A UK PTC

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Background Langerhans Cell Histiocytosis (LCH) describes a clinical spectrum of disease caused by accumulation of pathologicalangerhan’s cells infiltrating tissues. Further understanding regarding the patho-physiology and natural history of langerhans cell histiocytosis has emerged with the understanding of its status as a clonal BRAF stimulated malignancy.

Objectives To characterise the disease presentations of Langerhans Cell Histiocytosis in a UK Principal treatment centre (PTC) over the past 10 years.

Methods Our PTC serves a population of 3.6 million sharing care with 11 regional shared care centres. A retrospective case review was performed for all patients treated for LCH over the last 10 years. Data were collected on demographics, disease location, treatment modality, intervals of remission free outcome and latest evaluation at follow up.

Results 1041 children have been treated for cancer over the past 10 years (1/1/2006–31/12/2016). We identified 45 cases of LCH from children aged 2 mths to 16 years (median age of 2 years 6 mths) in our database accounting for 4% of our patients. There were 44% females and 58% males. 64% of cases presented with single-system disease at diagnosis and 36% with multi-system disease. 4 children were identified as having diabetes insipidus at diagnosis 2.5%.

33/45 (73%) had a biopsy performed consistent with LCH, 10 had no biopsy performed and 2 had a biopsy which was negative but were treated as per LCH due to typical clinic-radiological correlation. 2 cases of neurodegenerative LCH were identified in this case series.

Multi-system patients have a more challenging course and in our series 10/18 (56%) went on to have a recurrence and of these 2 patients have had 4 relapses and are still undergoing further therapy.

Conclusions Our centres experience over the past 10 years which shows the full spectrum of presentation from the benign self-limiting single system bony disease to that of extensive multi-system disease leading to secondary HLH. Novel strategies to evaluate and monitor BRAF in blood and urine such as those being employed in the latest international LCH IV trial offer the opportunity to better understand the risk factors for disease severity.

ARCHIVE DIS CHILD
A146

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Discussion

The increased number and proportion of patients diagnosed in the 15–24 age bracket who went on to develop mental health problems compared to those aged 0–14 emphasises the unique needs of this cohort. Further work will investigate the risks of mental health contact according to ethnic group, treatment modality and socio-economic status and seek additional mental health consultation data from linked primary care records.

Background

Children with cancer are at increased risk of malnutrition. Early nutrition intervention helps to maintain weight, lean body mass, improved treatment tolerance and QOL. RUTF, with higher recovery rates in pilot studies has brought a paradigm shift in the management of malnutrition. This pioneer trial evaluated the effectiveness of RUTF in prevention of malignancy related weight loss, improvement of micro/macronutrient status, treatment tolerance and QOL.

Methods 70 children (5–15 years) with hematolymphoid and solid tumours were enrolled post the appetite test. Randomization into 1:1 using a computerised table and stratification by type of malignancy. Nutritional status (weight, height, BMI) and calorie intake as well as weight gain at 6 weeks (2.6 kg vs. 2.8 kg) was higher in the RUTF arm compared to controls on standard dietary care. At 6 weeks there was significant reduction of MAM/SAM children ((16 vs. 23, p<0.05)) with increment of lean mass in the RUTF arm. Children on RUTF experienced significant reduction in the episodes of febrile neutropenia (18.9% vs. 30.3%, p=0.06), protocol delays (2.7% vs 30.3%, p<0.05), grade 3/4 neutropenia (40.5% vs. 66.7%, p<0.05), thrombocytopenia (21.6% vs. 30.3%, p<0.05) and anaemia (18.9% vs. 36.36%, p>0.05) beyond 6 weeks. Mean HRQOL scores were better in the RUTF arm at baseline, 6 weeks and 3 months.

Conclusion RUTF is cost-effective in improving nutritional status resulting in higher weight and lean body mass which translates into improved treatment tolerance and QOL.