Abstract IDDF2018-ABS-0262 Figure 1

He had sparse pubic and axillary hairs, small testes and inadequate phallus size. He had adequate cognitive skills with 30/30 mini-mental status, completed secondary-school but had poor scholastic performance compared to peers and siblings. There were extensive onychomycosis in all 4 limbs.

Results Stool microscopy and culture were normal. Blood investigations showed both albumin and globulin <2 mg/dl on repeated occasions. His 24 hour urinary protein excretion was negative. Echocardiography, colonoscopy, skiagrams and abdominal sonogram were normal. Contrast-enhanced CT abdomen showed diffuse gut wall oedema. Histopathology from endoscopic duodenal mucosal biopsy showed multiple dilated lacteals in submucosa containing lymph, suggestive of intestinal lymphangiectasia. The patient was put on a diet containing medium chain fatty acid (coconut oil) and high protein content. He improved and gained weight with remission of diarrhoea, and oedema in subsequent follow up for next six months.

Conclusions In unexplained cases of GI symptoms, anasarca with a decrease in both albumin and globulin, endoscopic biopsy of intestinal mucosa can help in diagnosis.

IDDF2018-ABS-0266

AGGRESSIVE MULTIMODALITY THERAPY IN EXTRA GASTROINTESTINAL STROMAL TUMOUR CAN HAVE PROLONGED SURVIVAL; A CASE REPORT

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Background Extra Gastrointestinal Stromal Tumour (EGIST) is a rare clinical entity with aggressive biological behaviour. Only few case reports have been published in the literature. We present a case of malignant EGIST who have prolonged survival with multimodality therapy involving surgery and targeted therapy.

Methods A 39 year female presented to our outpatient department in May 2016 with complaints of pain in the abdomen. The patient had a history of laparotomy in 2011 for a benign ovarian cyst. Eighteen months later She was diagnosed as a case of GIST and was on treatment with imatinib 400 mg/day. The patient was asymptomatic until in 2016 she developed pain abdomen. On clinical examination, there were palpable lumps involving umbilical and right iliac fossa. CECT showed a heterogeneous complex cystic mass in pelvis along with multiple omental and parietal wall nodules. The core biopsy suggested Malignant GIST positive for CD117, SMA and vimentin. CD34 was negative and Ki 67 was 60%. The patient was given Imatinib at a dose of 800 mg/day in divided doses. After five months of treatment, the patient had a good response but had intermittent lower abdominal cramps for which she was planned for surgery. Total Abdominal Hysterectomy with bilateral salpingo-oophorectomy, omentectomy and peritonectomy was done. Surgical recovery was good, and the patient was given imatinib 800 mg/day.

Results Post surgery histopathology also suggested Malignant Extra intestinal GIST with IHC similar to the core biopsy. The patient was compliant with therapy, and there was no comorbidity till the last follow update in February 2018. Post surgery patient was disease free for fourteen months and overall 61 months from the initial diagnosis.

Conclusions Extra Gastrointestinal Stromal Tumour is a rare disease, and no standard treatment guideline is available. Imatinib forms. The backbone of treatment and surgery is indicated for localised disease. In our case, aggressive surgery after a good response to Imatinib has resulted in longer survival. Although more data is required aggressive surgery with Targeted therapy seems promising.

IDDF2018-ABS-0267 AZATHIOPRINE FOR MAINTENANCE OF STEROID-FREE REMISSION: EVEN A LOW DOSE IS EFFECTIVE IN INDIAN INFLAMMATORY BOWEL DISEASE (IBD) PATIENTS

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Background Azathioprine is widely in the Asia Pacific region for patients with steroid-resistant and dependent IBD. However, the predictors of efficacy and optimal dosing of Azathioprine (AZA) has not been evaluated in this region. We aimed to investigate the optimal dosing and the predictors of efficacy of AZA in maintaining steroid-free remission in a well defined Indian IBD cohort.

Methods Prospectively collected data of patients on AZA from the IBD registry, at Asian Institute of Gastroenterology, Hyderabad was analysed. The relapse rate, time to relapse and reasons for stopping treatment were included. An age and a sex-matched historical cohort of patients not on AZA were used as a control. Median relapse-free interval was estimated using Kaplan-Meier survival analysis. The impact of treatment with azathioprine on the risk of relapse was assessed using Cox proportional hazards model.

Results 396 patients [Mean age 37 years, 59% men, 208 (52%) with CD] on AZA were identified. AZA was effective in preventing relapse in both Crohn's disease (CD) and Ulcerative colitis (UC) compared to patients not on AZA ($p < 0.001$) (table 1). The mean dose of AZA was 1.48 mg/kg (Range 0.59–3.77). Only 23.2% patients had to discontinue AZA for lack of effectiveness, pregnancy and side effects. Patients with CD on AZA took a longer time to their relapse than with UC on AZA (Log-Rank Test- $p = 0.072$) (figure 1). In the multivariate Cox proportional hazards model, UC patients had 80% higher chance of having a relapse compared to CD patients adjusting other variables (Hazard ratio- 1.8, 95% CI-1.2–2.7, $p = 0.007$).

Abstract IDDF2018-ABS-0267 Figure 1

Abstract IDDF2018-ABS-0267 Table 1

	Non-AZA (n=434)	AZA (n=396)	P-value (Fisher's Exact Test)
Number with Relapse	60.4% (262/434)	25.5% (101/ 396)	<0.001
CD	64.9% (98/151)	20.2% (42/208)	<0.001
UC	58.0% (164/283)	31.4% (59/188)	<0.001

Conclusions Low dose AZA appears to be a safe and effective drug for maintaining steroid-free remission in Indian IBD patients. AZA is more efficacious in preventing a relapse in CD compared to UC.

IDDF2018-ABS-0268 FAMILIAL AGGREGATION OF INFLAMMATORY BOWEL DISEASE IN INDIA: PREVALENCE, RISKS AND IMPACT ON DISEASE BEHAVIOUR

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Background Positive family history is considered the strongest risk factor for the development of Inflammatory Bowel Disease (IBD). However, information about familial aggregation of IBD in Asia is limited.

We aimed to analyse the prevalence and risk of familial IBD in an ethnically distinct Indian population and the differences in disease behaviour and severity between familial and sporadic cases.

Methods Familial aggregation was assessed in a large well established and prospectively maintained IBD database of 3553 patients (UC 2053, CD1500). The prevalence and relative risk of IBD in first and second-degree relatives of the index cases were evaluated. The disease behaviour and severity were compared to sporadic cases.

Results 4.13% of CD and 4.34% of UC patients had a family history of IBD. Positive family history was more common in the paediatric age group (<18 years at diagnosis) compared with adults (>18 years) ($p = 0.0002$, OR-2.8, 95% CI 1.6–4.8) (figure 1). Patients with a family history were more likely to have an affected family member on the paternal side (67.4% UC and 70.9% CD). Concordance of disease type in affected relatives was significantly higher in UC (79.7%) compared to CD (37.1%). Pan-colitis was higher in