041 PANCREATICOBILIARY VERSUS HEAD AND NECK PRESENTATION OF IGG4-RD: DIFFERENT SIDES OF THE SAME COIN?

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Introduction IgG4 related disease (IgG4-RD) is a rare immune mediated fibroinflammatory condition that can affect nearly any organ. Pancreaticobiliary (PB) and head and neck (HN) are two of the most commonly involved anatomical sites. A recent study has suggested the PB IgG4-RD and HN IgG4-RD have distinct clinical profiles with earlier age of onset in HN compared to PB and a more even gender distribution. Whether response to therapy or need to escalate therapy differs is unknown. In 2016 NHS England approved the use of Rituximab (RTX) in refractory IgG-RD. We aimed to assess differences between PB and HN IgG4-RD in a UK cohort of IgG4 disease managed by a tertiary IgG4-RD multispecialty team.

Methods We performed a retrospective study of a prospectively maintained multidisciplinary IgG4-RD database to identify patients diagnosed with PB and HN IgG4-RD (based on initial presentation) between 2005 and 2019. The electronic patient record was reviewed for details of clinical presentation, organ involvement, investigations and therapy. Use of RTX in cases diagnosed since 2016 was analysed.

Results 60 patients with PB IgG4-RD diagnosed between 2005 and 2019 and 14 with HN IgG4-RD diagnosed between 2013 and 2019 formed the study population. We compared clinical and therapy profiles (table 1) There was a significant difference in mean age at diagnosis, sex, median serum IgG4 level and multi-organ involvement. Persistent elevation of IgG4 following therapy was significantly more common in PB IgG4-RD than HN IgG4-RD. Treatment escalation was significantly more likely in HN IgG4-RD than PB IgG4-RD.

Conclusion In this study we found PB IgG4-RD to be associated with older age at diagnosis, higher serum IgG4 levels and greater multiorgan involvement. HN IgG4-RD was more likely to receive 2nd and 3rd line therapy and for treatment to lead to a reduction in serum IgG4. The findings of this study support the contention that PB IgG4-RD and HN IgG4-RD have different clinical profiles and represent distinct subtypes of IgG4-RD.

Abstract O41 Table 1 Comparison between PB IgG4-RD and HN IgG4-RD

Factor	PB (n=60)	HN (n=14)	P value
Age, mean (SD) years	64.4 (12.5)	51.1 (15.5)	0.001
Sex, male, n (%)	49 (81.7%)	8 (57.1%)	0.023
History of atopy, n (%)	22 (36.6%)	4 (28.5%)	0.76
Serum IgG4 > upper limit of normal (ULN) n (%)	44/59 (74.5%)	7 (50%)	0.10
Serum IgG4 level, median [IQR] multiples of ULN	2 [1–3.75]	1 [1–2]	0.036
Eosinophil count > ULN, n (%)	13 (21.7%)	5 (35.7%)	0.31
>1 organ involvement, n (%)	41 (68.3%)	5 (33.3%)	0.033
Elevated serum IgG4 after therapy, n (%)	37/44 (84.1%)	3 (42.8%)	0.031
Initial steroid therapy, n (%)	44 (73.3%)	14 (100%)	0.031
Second line therapy, n (%)	16 (36.3%)	10 (71.4%)	0.031
RTX therapy (in cases diagnosed since 2016), n (%)	1/29 (3.4%)	6/12 (50%)	0.001

042 INCIDENCE OF NEUROENDOCRINE NEOPLASMS IN ENGLAND 2015–2017

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Introduction Previously published National Cancer Registration and Analysis Service (NCRAS) data showed a higher than expected rise in incidence of Neuroendocrine Neoplasms (NENs) between 2001 and 2015 (Genus et al 2019). We aimed to analyse and report new NCRAS data in England including years 2015 – 2017 as coding (ICD-O-3) quality improves. We hypothesized incidence of NENs would continue to rise and increased accuracy of coding would lead to more accurate tumour classification.

Methods Public Health England data for incidence and prevalence of NEN from NCRAS were analysed, broken down by age group, site, morphology and grade. Statistical analysis was performed using STATA to give age-standardised incidence rates (per 100,000 population) and upper and lower confidence intervals.

Results During 2015 – 2017, 14,138 NENs were diagnosed in England; 7093 (50.17%) female. In 2016, NEN incidence in England was 9.37 per 100,000, an all-time high. Incidence remained the same in 2017. Incidence is trending upward in all agegroups. Incidence is higher in females in 0–54 agegroup, but in males in aged 65+. NEN incidence increased for most sites except lung NET which levelled off after previous yearly rises. Small intestine NET continued to rise sharply with a male preponderance. Pancreatic NET incidence continues to rise steadily. 'Neuroendocrine tumour NOS (not otherwise specified)' incidence reduced whilst 'Carcinoid tumour' and 'Atypical carcinoid tumour' rose, possibly due to recoding. NEC G3 incidence decreased, continuing a trend since 2011, with NET G1/G2 continuing to rise. 23-year prevalence for NENs in England was 26,735 (survival of 108,554 cases). 2017 prevalence was 48 per 100,000 (mid-year population of 55,619,430).

Conclusions Age standardised incidence of NEN has risen above 9 per 100,000 for the first time. Rising incidence of NET remains unexplained, and with increasing survival with chronic symptoms, will become increasingly important. Differing male and female incidence in agegroup and certain sites, for example small intestine NET being greater in males, needs further analysis. Incidence of lung NET reduced for the first time. Improved coding of data may explain the reduction in non-specific 'NOS' numbers. NEC G3 incidence decreased whilst NEC G1/G2 increased, again likely due to coding changes. Prevalence is now higher than many other cancers. As coding continues to improve, more work on NCRAS data is needed to establish a true reflection of the NET landscape.