with iRPF from four tertiary medical units in Greece and b) to evaluate factors potentially associated with disease relapse.

Methods: Medical records of patients diagnosed with RPF from 2000–2018 in four rheumatology units (Laiko, Eυροclinic, Sismanoglion and Sotiria Hospitals) were retrospectively evaluated. Sixty-seven patients with iRPF were included in the study.

Results: The median age at diagnosis was 56 years (IQR:52.0–60.0), with median disease duration 8.0 years (IQR:3.0–11.0), 58% were smokers and 73% males. Patients more often presented with constitutional symptoms (57%), low back pain (63%), raised inflammatory markers (78%), anemia (43%) and compromised renal function (15%). Commonest imaging findings were periarteric-perilliac mass (46%), periarteric mass (33%), periarteritis (25%) and hydronephrosis (36%) with envelopment of one (31%) or both ureters (16%). Tissue biopsy was requested in all patients. Biopsy was performed in only 10% with 3 having marked numbers of IgG4-positive plasma cells. Serum IgG4 was measured in 36/67 and 36% had elevated levels at diagnosis (median 224 mg/dl, IQR:174-328). Clinical/laboratory/radiological presentation did not differ between patients with elevated and normal serum IgG4 levels. Steroids were first-line treatment in 93% of patients. Other immunosuppressives used as steroid-sparing agents were azathioprine (70%), cyclophosphamide (19%), mycophenolate-mofetil (18%), D-penicillamine (12%) and methotrexate (8%). Relapse occurred in 19% of patients at a median of 36 months (IQR:18-66) after diagnosis with 69% of them being under therapy. Relapse did not correlate to initial imaging findings or to any treatment modality, yet patients with increased serum IgG4 tended to have higher relapse rate (26% vs 11%, p=0.071).

Conclusion: Diagnosis of iRPF was mostly based on imaging studies in our cohort. Steroids were used as first-line treatment. Relapse occurred in one-fifth of patients independently of initial clinical/radiographic presentation or treatment modality used. RPF patients with initially elevated serum IgG4 levels tended to have a higher relapse rate.

REFERENCES:
