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MORPHOLOGICAL DIAGNOSTICS ISSUES OF GRANULOMATOSIS WITH POLYANGIITIS

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Introduction. Granulomatosis with polyangitis (GPA) is a rare autoimmune disease marked by necrotizing vasculitis of small vessels, pauci-immune granuloma formation, and tissue damage, predominantly affecting the respiratory tract and kidneys, often progressing to chronic renal failure. GPA has an annual incidence of 3-14 cases per million people per year. It slightly favors males (M:F ~1-1.5:1) and is most common in Caucasians, northern United states of America.

Aim: to evaluate renal histopathological features in GPA across biopsy samples & their association with clinical data.

Material and methods. 11 kidney biopsies from patients diagnosed with GPA, including one patient with an additional nasal biopsy were obtained via ultrasound guided percutaneous needle biopsy, fixed in 10% formalin, paraffin embedded and sectioned at 3µm. Sections were stained with hematoxylin & eosin (H&E), periodic acid schiff (PAS), Masson's trichrome and Jones stain for Histopathological evaluation. Slides were examined under light microscopy. While Immunofluorescence assessed immune deposits (IgA, IgG, IgM, C3, C1q). Clinical data were obtained from referral charts.

Results and discussion. There were 6 (54.5%) females and 5 (45.5%) males. (M:F = 1:1.2). The age of patients ranged from 30 to 62 years. The mean age was 47 ± 11.2 years. The diagnosis of GPA in all patients were confirmed by the presence of pauci-immune pattern, focal & segmental necrotizing glomerulonephritis with crescent formation, most consistent with GPA in the setting of ANCA positivity. On histopathology examination, all the patients presented with crescentic glomerulonephritis with crescent formation in glomeruli. 6 (54.5%) of the case presented with diffuse crescentic glomerulonephritis & 5 (45%) with focal crescent formation.

On serology examination, 6 (54.5%) presented with C-ANCA & 2 (18.2%) with P-ANCA positivity. 3 (27.3%) cases, the specific type of ANCA could not be determined. On clinical examination, 5 (45.5%) cases presented with "nephritic syndrome". Out of which 3 (60%) have high blood pressure & all have proteinuria, none of them with nephrotic syndrome. In all cases there was functional damage of kidneys. Out of which 2 (18.2%) progressed to acute renal failure & 2 (18.2%) progressed to chronic renal failure. 6 (54.5%) of patients with diffuse crescentic glomerulonephritis developed rapidly progressive glomerulonephritis.All cases presented with systemic damage of organs like kidney, lungs, nose, maxillary sinus. 4 (36.4%) cases presented with lungs damage with hemoptysis & 4 (36.4%) presented with pneumonia with tuberculosis ruled out.Nasal mucosa shows necrotizing vasculitis, granulomas, and ulceration.

Conclusion. To diagnose GPA, a comprehensive approach is required with a thorough analysis of clinical and laboratory data, the results of instrumental examination of the patient and morphological assessment of changes in the affected organs.