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Case series: Presence of sacroiliitis and their associated HLAB27 positivity and vitamin D deficiency in cohort IgG4-RD Egyptian patients

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Background: IgG4 related disease is an immune mediated condition that can affect different organs. Multiple organ involvement is seen in 60-90% of patients. Many organs have been described to be involved, the pancreas, salivary glands, lacrimal glands retroperitoneal fibrosis, periaortitis, thyroiditis, and lung fibrosis. IgG4 related sacroilitis has not been reported previously except in two occasions.

Objectives: To report in a retrospective analysis, the presence of sacroilitis, possible associated vitamin D deficiency and HLAB27 positivity in cohort of Egyptian patients diagnosed as IgG4-RD who were seen in our center at Kobri El-Kobba military medical complex in addition to Air force specialized hospital in Cairo, Egypt during the period of Jan. 2015 till July 2017.

Methods: Five patients with biopsy proven IgG4-RD were included. The diagnosis was made based on clinical manifestations, detecting elevated circulating plasmablasts, imaging studies, flowcytometry of peripheral blood with gating to CD38, CD19, CD20 and CD27 and their appropriate tissue biopsy characteristic to the disease. Two patients (40%) had retroperitoneal fibrosis while the two patients (40%) had interstitial lung fibrosis and the last patient (20%) had an IgG4 related sclerosing cholangitis. Three of the five patients (60%) were males between the age of 50 and 60 years old. The other two patients (40%) were middle aged females. For all patients, MRI sacroiliac joints were done for their common complaint of low back pain related anatomically to the sacroiliac joints. Also, HLAB27 was done for all and an assay for vitamin D level was done during the bone assessment for the patient's due to their complaint of generalised bone pain and myalgia. A bone biopsy was not done, although the patients were diagnosed as IgG4-RD, yet, bone biopsy was essential to confirm whether sacroilitis was due to an IgG4-RD pathology or any other possible etiology.

Results: All patients showed bilateral sacroilitis as proved by MRI to the sacroiliac joints, with marrow edema in three patients (60%), erosions in one patient (25%) and vacum air bubbles in the sacroiliac joint of one patient (25%). Vitamin D assay proved deficiency in all patients (100%) with varying degrees in the deficient titer. HLAB27 was positive in three patients (60%) and it was negative in two patients (40%) of the included cases.

Conclusion: This retrospective case study reported the presence of sacroiliitis, low vitamin D level in a case series of IgG4-RD cohort of Egyptian patients as well as the coexistance of a positive HLAB27 in (60%) of the included patients. Further studies are essential to elucidate the etiopathogenesis of the sacroilitis associated with the IgG4-RD cases.

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