Discussion – This is the first objective study using well instruments to evaluate pediatric TA.

Conclusion – TA in children needs a close follow up due to high relapse rates & requires aggressive immunosuppression.

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P112

IgG4 in chronic periaortitis

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Introduction – Chronic periaortitis (CP) is a rare disease characterised by a fibro-inflammatory tissue surrounding the abdominal aorta and the iliac arteries. It has been reported that about 50% of CP belong to the spectrum of IgG4-related disease; CP with high IgG4 levels seems to occur almost exclusively in men and to have more frequent extra-retroperitoneal manifestations. The diagnostic role of IgG4 in CP is unknown. The aim of this study was to explore the clinical significance and diagnostic reliability of IgG4 in CP.

Methods – Total IgG, IgA, IgM, IgG1, IgG2, IgG3, IgG4 levels were measured in 66 consecutive patients with active CP, 51 healthy controls, and 58 disease controls (36 with retroperitoneal neoplasms and 22 with active abdominal aortitis secondary to large vessel vasculitis). Normal IgG4 serum levels were 8–140 mg/dL.

Results – High IgG4 (> 140 mg/dL) levels were found in 14 CP patients (21%). Demographic characteristics and clinical features were similar between the IgG4-related and IgG4-unrelated cases; there was no difference in gender distribution (78% vs. 61% were male, \( p = 0.478 \)) or age [median, interquartile range (IQR), 57 (52–63) vs 57.5 years (52–63)]. Of the nine patients with extra-retroperitoneal involvement, only 3 (33%) had high IgG4 levels. The two groups did not differ in CP localisation, prevalence of acute renal failure or thoracic aorta involvement.

High IgG4 levels were found in three healthy (5.8%), two neoplastic (5.5%), and two (9%) aortitis patients. The median (IQR) IgG4 levels were 46 mg/dL (26–122.8) in CP vs 35 (14.4–69.5) in healthy controls \((P = 0.048)\), 40 mg/dL in neoplastic controls (17.5–68.8) \((P = 0.14)\) and 30.5 mg/dL (12.3–63) in aortitis patients \((P = 0.14)\). The area under the ROC curve (IgG4 in CP vs in all control subjects) was 0.597 (95% CI 0.509–0.685).

Conclusion – Only ~20% CP patients have high IgG4 levels. IgG4 do not seem to discriminate different CP subsets and its diagnostic reliability for CP is low.

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P113

Takayasu arteritis (TA) and sacroiliitis. A large vessels vasculitis masquerade from anti-TNF alpha therapy

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Introduction – A 22-year-old woman was admitted to our hospital for inflammatory back pain, abdominal pain and fatigue during the last two years with leucocytosis microcytic anemia elevated platelet count hypergammaglobulinemia and elevated CRP.

Methods – A MRI diagnosed axial spondiloarthritis. Adalimumab 40 mg every two week was started with a good response. Six months later she developed a reduction in left radial pulse associated with numbness of the left hand. Doppler echocardiography showed severe aortic failure with ascendental aortic dilatation and LVD with EF of 45%. A US detected a significative left subclavian, left external carotid and vertebral ste-nosis and aneurismatic dilatation of right subclavian confirmed at MRA.

Results – TA with sacroiliitis was diagnosed. PET was negative for active lesions so aortic valve and ascending aortic replacement was performed. The patient is alive and well.

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P114

ITAS.A suggests persistent disease activity in Takayasu aorto-arteritis (TA) after induction therapy

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Introduction – The original Indian Takayasu Activity Index (ITAS2010) was developed as clinical disease activity tool. In active disease, the score was high. Acute phase resone (ESR or CRP) was added to the Indian Takayasu Activity Index (ITAS2010), by a score of 0 to 3 and ITAS.A was compared to ITAS in response to therapy at two centres.
Methods. – In Vellore, 132 patients with active disease were studied at 0 and 6/12 after therapy with steroids plus mycophenylate. In Lucknow, 46 patients were assessed at 0 and < 12/12 after therapy with steroids plus methotrexate or azathioprine while TADS (Takayasu Damage Score) was also used to assess development of damage.

Results. – In Vellore, ITAS2010 indicated satisfactory suppression of disease activity. However the ITAS.A score indicated continued disease activity. In Lucknow, at follow-up, the ITAS-A was higher than 2 in 79% of cases and only one in four had a value less than 1, even at 12 months. The mean score of TADS was 6 indicating marked damage.

Discussion. – The incomplete response to active induction therapy with persistent disease activity despite clinical improvement noted for ITAS.A was seen in two centres using different immuno-suppressive plus steroid regimes. Persistent activity would predict development of damage and indeed significantly elevated TADS scores were seen.

Conclusion. – ITAS.A, combining clinical data plus acute phase response, provides new information. The apparent incomplete response to therapy despite clinical improvement has major implications for therapy.

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P115

Chronic asymptomatic aortic dissection (AD) in Takayasu’s arteritis (TA); how to treat?

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Introduction. – AD is extremely rare in TA and only a limited number of reports have been published. Little is known about the management of this rare complication and usually surgery is reserved for Stanford type A. Our patient was managed medically and had a good outcome despite her Stanford type B AD. In our case the diagnosis of chronic intramural haematoma (IMH) was made radiologically using serial MRA in a clinically asymptomatic patient. Despite that we changed patient’s immunosuppressive therapy as we believe IMH was caused by persistent low-grade vasoritis.

Methods. – A young Indian lady was diagnosed with TA based on a history of claudicant legs and left arm with subsequent angiogram revealing complete occlusion of the left subclavian artery and distal abdominal aorta. She underwent aortobifemoral arterial bypass grafting in 2002 with complete resolution of her symptoms. Histology of the abdominal aorta specimen obtained during the operation confirmed TA.

Results. – She stayed on a low dose of prednisolone and methotrexate (MTX) for nearly a decade. HTN required frequent adjustments of antihypertensive therapy and she has always had a mildly elevated CRP 10–25. Serial MRA from 2008 to 2011 showed slowly increasing thickness of the posterior wall from aortic arch down to descending thoracic aorta. Subsequent CT angiogram confirmed IMH starting from left subclavian artery following spiral dissection to the level of abdominal aorta at L3-L4 just before the graft (pic 1–2).

Discussion. – IMH is a variant of AD caused by ruptured vasa vasorum into the media wall. We hypothesise that fragility of the aortic media in TA is caused by vasa vasoritis [1] leading to infiltration of media by inflammatory cytokines thus causing disruption of elastic fibres and facilitating formation of IMH. No doubt HTN also contributes into fragile media.

Conclusion. – Based on the above we opted for switching this patient’s immunosuppressants from MTX to Mycophenolate Mofetil and to consider further escalation to biologic therapy in case of future radiological progression.

Reference

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P116

Contribution of anti-ferritin antibodies to the diagnosis of giant cell arteritis

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Introduction. – The diagnosis of giant cell arteritis (GCA) can only be ascertained by performing temporal artery biopsy (TAB). Recently, Baerlecken et al. [1] reported on the detection of antibodies directed at the human ferritin heavy chain (FTH1) in 92% of patients with GCA vs 1% of healthy controls. We decided to evaluate the diagnostic value of anti-ferritin antibodies in patients undergoing TAB for a suspicion of GCA.

Methods. – We included 122 consecutive patients suspected of GCA. Blood sampling was performed at the time of TAB. Sera from 40 healthy individuals were used as negative controls. We investigated for the presence of IgG directed against 19 FTH1 amino acids by using an ELISA test. Correlations between FTH1 antibodies and clinical manifestations were investigated using non-parametrical tests.

Results. – Anti-FTH1 antibodies were identified in 72.5, 41.3, 31.9 and 2.5% of patients with TAB+ GCA, TAB- GCA, GCA controls and healthy individuals, respectively, with a threshold at the mean of healthy controls + 3 SD, anti-FTH1 antibodies were identified in 60, 34.5, 21.2 and 0% of the patients with TAB+ GCA, TAB- GCA, GCA controls and healthy individuals, respectively.

By grouping TAB+ GCA, TAB- GCA patients with a threshold at 2 SD, the positive and negative predictive value were of 71.9 and 56.9%, respectively. Positive and negative likely ratios were at 1.96 and 0.58, respectively. In addition, in our population, anti-FTH1 antibody titer correlated significantly with CRP and no correlation was found with aortic and/or visual impairment.

Conclusion. – We therefore confirm the presence of anti-FTH1 in 72.5% of patients with histologically proven GCA. However, the detection of anti-FTH1 is not contributory to the diagnosis of GCA in a cohort of patients with suspected ACG. The ability to identify a specific sub-group...