36. ALL HEADACHES ARE NOT GCA

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Introduction: Vasculitis is a heterogeneous group of autoimmune disorders having multisystem involvement. In many cases, the neurological disorders have an atypical clinical course or even an early onset, and the healthcare professionals should be aware of them.

Case description: We report a case of 57-year-old Asian male without any significant past medical history admitted under the urology department. He had presented with abdominal pain and fever and was being treated for presumptive pyelonephritis (on antibiotics). He was referred to rheumatology when he developed severe unilateral headache on right side with significant scalp tenderness. Headache involved temporal and fronto-parietal-occipital area with no jaw claudication or any significant visual disturbance. There was no limb claudication or polymyalgia symptoms. On examination he had new onset hypertension (BP:174/94mmHg), rest of the systemic examination was normal with no bruit. Musculoskeletal and skin examination was normal.

Investigations included CRP of 204, hemoglobin 133, white cell count 15.2 (neutrophils: 12.06), creatinine 118 (eGFR 59). Liver function: normal. Urine dip: Normal and blood/urine culture: negative. Immunological profile and viral screen was negative. Urgent CT head was done for severe headache which was normal.

Patient was referred to rheumatology with provisional diagnosis of giant cell arteritis due to headache and raised inflammatory markers. On reviewing his CT abdomen, the renal parenchyma showed symmetrical well demarcated bilateral involvement which was more in favor of renal infarct rather than pyelonephritis. CT angiography of abdomen was requested which showed multiple vessel involvement including mesenteric artery and bilateral renal arteries with beaded appearance and stenosis.

MR angiogram showed 6mm aneurysm of PICA with stenosis of P1 segment of PCA. Currently he is being screened for ADA2 deficiency. During his stay his renal function deteriorated, not responding previously to antibiotics.

He was started on methylprednisolone pulse and antibiotics stopped in view of worsening liver function. On day 3 of methylprednisolone pulse he had significant improvement of his headache with stabilization of renal and liver function. Our plan is to start him on steroid sparing therapy based on the ADA2 levels.

Discussion: The classification of vasculitis is still unsatisfactory as the patho-genetic mechanisms have not been fully understood. Existing criteria classify based on predominant vessel involvement but there is still some overlap between them. The prevalence of CNS involvement in medium vessel vasculitis ranges as widely as 2-10% and does not usually occur until late in the course of the disease. The most commonly reported CNS manifestation is diffuse encephalopathy, followed in frequency by focal deficits and seizures. Intracranial aneurysms are rare with around 15 reported cases usually multiple and located in supra- as well as infra-tentorial compartments. Most of the cases presented with subarachnoid or parenchymal hemorrhage. Treatment guidelines are still not clear and most are treated conservatively by medical management. Repeat hemorrhages or re-bleed in spite of medical treatment have also been reported

Deficiency of ADA2 (DADA2) has been recently recognized first molecularly described monogenic vasculitis having biallelic hypomorphic mutation in ADA2 gene. CNS involvement is one of the main features of DADA2 which presents with a wide spectrum of clinical manifestations ranging from systemic inflammation to cutaneous or visceral PAN like vasculopathy and early onset stroke.

Our patient had presented with primary CNS and renal involvement with impending rupture aneurysm. Timely control of inflammation is pertinent in such cases with remission induction and maintenance to prevent stroke. Although cyclophosphamide is still used in such cases DADA2 usually require anti-TNF therapy. Our patient had good response to primary steroid therapy.

Key learning points: Acute GCA-like neurological presentation of medium vessel vasculitis with cerebral involvement is rare. If not recognised in a timely fashion, it can lead to sub-arachnoid haemorrhage which is the most common presentation of such cases.

DADA2 is a recently recognised entity that presents with stroke and vasculitis/vasculopathy and responds to anti-TNF therapy.

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