Clinical cases

Case presentation: A 77 year old female patient was referred to our rheumatological outpatient department because of localized pain in the left lateral part of the neck, which persisted over two days. Swallowing of fluids or solid food was painful, but unimpaired. The patient mentioned no recent trauma, but he reported symptoms of an upper respiratory tract infection, without fever, two weeks before. Clinical investigation showed no signs of focal swelling or palpable lymph nodes. ENT consultation ruled out inflammatory processes causing the symptoms including tonsillitis, pharyngitis or laryngitis. Analgesic treatment with tramadol was not effective. Laboratory investigations revealed slightly increased CRP and Erythrocyte Sedimentation Rate (ESR) values. ANA and subsets, as well as ANCA were negative. An ultrasound of the region showed a localized swelling of the media and adventitia of the left common carotid artery, involving half of the circumference over a length of 4 mm. Multiple lymph nodes with signs of inflammation were present on both sides of the vessel. No dissection or stenosis was detectable. Due to a typical clinical presentation and ultrasound appearance without carotid stenosis, typical localisation at the distal part of the carotid communis artery the diagnosis of carotidynie or TIPIC was made. Treatment with nonsteroidal antiinflammatory drugs was initiated without any effect. Therefore, we initiated a treatment with steroids (0.3mg/kg body weight). 3 days after the beginning of the therapy the unbearable pain nearly resolved. An MRI angiography of the vessel performed 5 days after the initiation of the steroid treatment showed an eccentric wall thickening with diameter of 5mm near the bifurcation of the common carotid artery, with a slightly increased enhancement of the carotid wall, but no further inflammatory structures. Awareness of this syndrome and its excellent visualization for diagnosis, exclusion of other pathologies and follow up by high resolution ultrasound could be useful to reassure patients about the benign nature of this syndrome and help the clinician to avoid more invasive imaging modalities.

Conclusion: The disease is first reported by Fay in 1927. Despite only a few case reports are available in the last 20 years and mostly one case is reported. Usually an inflammatory process is suspected. Although a rare event sonography is capable to diagnose such vessel abnormality with high precision. Treatment with NSAIDs and, if not sufficient, eventually steroids are required. REFERENCES: NIL. Acknowledgements: NIL. Disclosure of Interests: None Declared. DOI: 10.1136/annrheumdis-2023-eular.4353

Case presentation: A 77 year old female with a history of hypertension, diabetes, uterine and breast cancer in remission presented to the hospital with right eye vision loss. She reported two weeks of blurry vision in her right eye, bilateral temporal headache, jaw claudication, weight loss, and loss of appetite. She denied prior similar episodes. Exam revealed scalp and right temporal artery tenderness, right eye vision loss. She had equal radial and brachial pulses, and 5/5 strength in bilateral upper and lower extremities. Laboratory work-up showed ESR 120 (ESR 0-38mm/hr) and C-reactive protein 62 (CRP 0-5mg/L). Patient was started on intravenous (IV) methylprednisolone 1g daily for presumed GCA, then tapered and discharged on oral methylprednisolone 48mg (1mg/kg) daily. Temporal artery biopsy confirmed GCA showing patchy moderate chronic inflammation in the intima and media with small lymphocytes, macrophages, and rare giant cells. On follow up, she continued to have right eye vision loss but improvement of other GCA symptoms. Within one month of her initial presentation, she returns to the hospital for left sided weakness, facial droop, and dysarthria.

Results: She endorsed compliance with methylprednisolone, aspirin, and atorvastatin. Computerized tomography angiography (CTA) showed interval worsening of the severe stenosis of the clinoïd segment of the right internal carotid artery (ICA) now with near occlusion. Labs with ESR 33, CRP <0.2. Exam with 4/5 strength in the left upper extremity (LUE) and left lower extremity (LLE). On hospital day 3, she had fluctuating weakness in the LUE and methylprednisolone was increased to 1g daily. Repeat CTA with stable severe focal stenosis of the right clinoid ICA. Cerebral angiogram showed multiple vessels in the anterior and posterior circulation with evidence of irregularity in the lumen suggestive of diffuse vasculitis but no evidence of atherosclerosis in the right ICA. On hospital day 6, weakness worsened in the proximal and distal LUE to 1/5 strength and LLE to 3/5 strength. Given concern for GCA progression she was started on IV tocilizumab 6mg/kg and had emergent angiography with right ICA angioplasty. Patient completed 5 days of IV methylprednisolone 1g and was tapered and discharged on methylprednisolone 48mg daily and tocilizumab 162mg subcutaneous weekly. On follow up she continued to improve; exam without facial droop, 5/5 upper extremity strength, and 3/5 lower extremity strength. This highlights a rare case of GCA related stroke within the ICA territory despite being on high dose steroids. There was a diagnostic dilemma and the initial suspicion for GCA related stroke was low because the patient had been on high dose steroids for 3 weeks with normal inflammatory markers, but cerebral angiography confirmed the evidence of vasculitis. Progression of symptoms despite high dose steroids called for aggressive therapy with angioplasty until therapeutic effect of tocilizumab was achieved. Patient’s response to therapy suggests that tocilizumab is effective for GCA related stroke.

REFERENCES:

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