

Window of Opportunity in Vasculitides

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Abstract

Most of vasculitides are clinical entities upon initiation of pathology, however, early manifestations of most necrotizing vasculitides are rather non-specific and subject to overlooking. Fever, malaise and anorexia are among these constitutional symptoms in these settings.

Small vessel vasculitis (SSV):

Isolated cutaneous vasculitis or leukocytoclastic vasculitis is usually the sole vasculitis of minimal significance and self-limited in most instances. These types of vasculitides are usually diagnosed on time due to characteristic skin manifestations and there is no concern about them [1]. However, other SVVs can be life or organ threatening and early diagnosis and prompt medical intervention is crucial. ANCA associated vasculitides (GPA, EGPA, MPA) can generally be presented with pulmonary, renal, skin or neurologic manifestations with evidence of extravasation of RBCs from the capillary bed. Diffuse alveolar hemorrhage (DAH) is the most devastating vasculitis condition with rapidly progressive course and universally fatal if left untreated. In recent era, DAH is better diagnosed and managed with better outcomes. Upon the diagnosis of DAH is made, intensive immunosuppression and possible plasma exchange is standard of care that dramatically improved disease outcome. DAH could be the single vasculitic condition that needs emergent medical intervention even before establishing the primary cause of it due to its grave behavior and very high chance of emerging death. Differentiating DAH from other more common causes of hemoptysis (infections or neoplasm) is crucial and starting therapy before time-consuming diagnostic approaches is mandatory. Due to wide range of unexpected clinical presentations related to vasculitis, all practitioners should keep vasculitis in any complicated case of rather common and benign conditions. Few physicians if any can imagine an ANCA associated vasculitis as basic mechanism of otitis media, Riedel's thyroiditis or influenza vaccination complications [2-4]. In case of non-systemic vasculitis neuropathy or single organ vasculitis (SOV), a rather new term that may be less familiar to most rheumatologist, systemic signs and symptoms may be absent and high index of suspicion by a clinician could save the organ after proper management.

Medium Size vasculitis:

Polyarteritis nodosa and Kawasaki disease (KD) are main clinical conditions in this category of vessel size. Clinical settings related to medium size vasculitis is aneurismal dilatation and infarction of target tissues. Prolonged unremitting fever and other constitutional symptoms along with muco-cutaneous manifestation and non-exudative conjunctive injection in a child always alerts for KD [5].

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On the other hand mononeuritis multiplex, abdominal vascular event and hemodynamically significant bleeding, bowel infarction, refractory or unexpected case of hypertension, scrotal/ testis pain, cryptogenic abdominal pain, ischemic skin lesions or single organ infarction [6-8].

Large vessel Vasculitis:

Giant cell arteritis (GCA) is the most common idiopathic vasculitis in elderly [9]. It can be challenging with more indolent course that can be misinterpreted for months or even years. Initial non-specific manifestations are sometimes followed by a quiescent clinical but not pathological phase. Thus, attention should be made in potential cases of TAK or TA. Delay in diagnosis may herald major visual and neurologic damage [10]. Along with the non-specific clinical presentation in initial phase of disease, para-clinical diagnostic tools are important in definite diagnosis of GCA. Serum markers (ESR and proinflammatory cytokines and anemia of chronic disease) are usually abnormal in case of GCA on the other hand many cases of GCA have normal blood markers and indolent [11]. Gold standard of diagnosis is based on tissue diagnosis, however ultrasonography and MRI angiography (MRA) has a major role in establishing large vessel vasculitis in the absence of tissue confirmation [12]. In recent years role of PET-CT is grown dramatically especially in early diagnosis of disease in pre-occlusive phase of diseases [13].

Unusual presentations of GCA could be tongue necrosis, TIA, upper arm/leg claudication [14], one and a half syndrome described by Fisher [15], and many other neurologic manifestations in elderly. Dementia in pertinent setting has been attributed to GCA that can be easily cured by appropriate management [16].

For years, the main clue to diagnosis of Takayasu arteritis (TAK) was diminished or absence of radial pulses along with persistently elevated ESR and possible other signs of inflammation within the body. This approach no longer could be a rational one due to long delay between detection and treatment in order to prevent progression of pathology. We suggest in any fever of unknown origin or other constitutional features with persistently elevated ESR in pertinent clinical setting to be “actively” screened for possible TAK in “pre-pulseless” or “pre-occlusive” period. Due to inherent tendency to major vessels, we would suggest

any sensitive imaging technique such as MR angiography or PET-scan to detect early signs for vessel wall inflammation.

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