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ARP Case Report Nº 17: Takayasu Arteritis

Caso Clínico ARP Nº17: Arterite de Takayasu

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Correct Answers - Clinical Case Nº 17

- Alcinda Reis

Abstract

39 year old female, presenting at ER with aphasia and right hemiparesis. A few hours later, develops mild hemoptysis. Thoracic CT performed to exclude pulmonary embolism. Patient taking immunosupressants for suspected demyelinating disease (multiple sclerosis?), due to relapsing-remitting but progressive episodes of limb weakness and numbness and impaired and blurred vision.

Keywords

Takayasu arteritis (TAK); Vasculitis; Diffuse alveolar hemorrhage (DAH); Viric (CMV) pneumonitis; Imunossupression.

Resumo

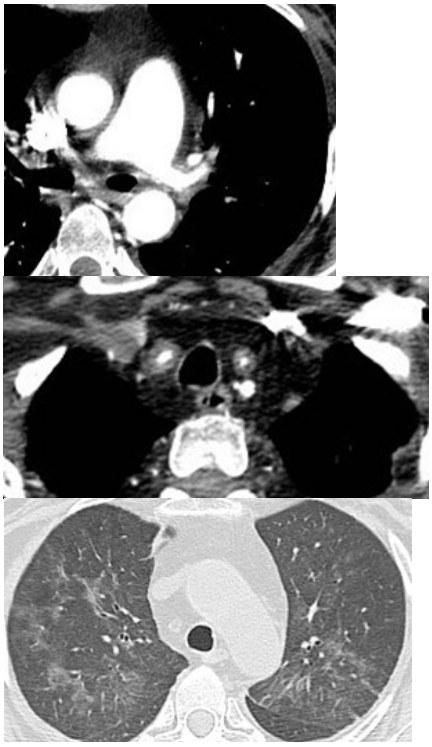
Doente sexo feminino, 39anos, admitida no serviço de Urgência por quadro de afasia e hemiparésia direita. Algumas horas depois, inicia episódio de hemoptises moderadas, tendo sido efectuada TC torácica, para exclusão de tromboembolia pulmonar.
A doente estava sob terapêutica imuno-supressora, por suspeita de doença desmielinizante do SNC (eventual esclerese múltipla), por episódios de imuno-suprimida, por episódios auto-limitados e intermitentes, mas progressivos, de parestesias e perda de força muscular nos membros, bem como de visão turva.

Palavras-chave

Arterite de takayasu (TAK); Vasculite; Hemorragia alveolar difusa (DAH); Pneumonite vírica por CMV; Imunossupressão.

Presentation of the case

39 year-old female presenting at ER with aphasia and right hemiparesis, and a few hours later develops mild hemoptysis. She was taking immunosuppressants for suspected demyelinating disease (multiple sclerosis?), due to relapsing-remitting but progressive episodes of limb weakness and numbness and impaired and blurred vision.
Chest CT was performed to exclude pulmonary embolism. The CT-angiography excluded thromboembolic pulmonary disease, but showed marked stenosis of left main pulmonary artey, and also the marked concentric thickening wall of the supra-aortic vessels, mainly the right brachiocephalic artery and the left subclavian artery, with a stenotic lumen. The thoracic aorta seemed disease-free.
The brain-CT performed on admission had showed a left hemisphere total anterior circulation ischemic stroke.
With these imaging findings, combined with the clinical symptoms, physical signs (arterial hypertension and absent radial pulses), and laboratory evaluation (serum antibodies), we made a diagnosis of Takayasu Arteritis.
The chest-CT also revealed parenchymal alterations, with widespread ground-glass infiltrates/opacities, which we attributed to diffuse alveolar hemorrhage, the falling hemoglobin (drop of 2.9g/dl hemoglobin level) also supportive of the presence of hemorrhage. However, testing for infection was mandatory, given the pharmacologically induced imunossupression (due to the hypothesis of demyelinating disease that had been considered a few weeks earlier).



Bronchoscopy with bronchoalveolar lavage was therefore performed, not only to confirm pulmonary alveolar hemorrhage, but also to rule out other entities, such as infection (fluid was tested for bacteria, viruses and some fungi, namely Peumocystis jirovecii and Citomegalovirus), and came back positive por CMV.
Hence, the final diagnosis was Takayasu arteritis, with diffuse alveolar hemorrhage due to viric (CMV) pneumonitis in an imunossupressed patient.

Discussion

Takayasu Arteritis (TAK) is a rare, chronic, idiopathic (it is thought to be an autoimmune disease) inflammatory vasculitis that primarily affects large vessels, mainly involving the thoracoabdominal aorta and its branches and the pulmonary arteries.
It is much more common in women than men (9:1), and most often starts in young adults. The non-specific inflammation causes intimal fibroproliferation, which usually leads to concentric wall thickening, fibrosis and thrombus formation and ultimately leads to segmental stenosis, occlusion, dilatation, and aneurismal formation in the vessels involved.
Manifestations range from asymptomatic disease, found as a result of reduced or absent peripheral pulses (hence the name «pulseless disease»), to catastrophic neurologic or cardiac involvement. Many of the symptoms may result from vessel stenosis or thrombus formation and reflect involvement of the aortic arch and its main branches: limb claudication; weakness and fatigue; dizziness, headaches or fainting; high blood pressure; chest pain; heart attack; transient amaurosis, blurred vision, syncope; stroke.
Also, the disease is so rare that doctors may not easily recognize it. Thus, there is often a delay in detecting it, sometimes several years.
Because of its considerable morbidity and mortality, an accurate and early diagnosis plays a crucial role in improving the outcome for patients with suspected TAK, and since large-artery biopsies cannot easily be done, imaging is considered the cornerstone of its diagnosis.
CT/MR angiography is a reliable tool in non-invasively depicting both luminal and mural lesions in the aorta and its main branches. CT and MRI findings of TAK include vascular wall thickening (it can be several milimitres) and enhancement in the active acute phase, with transmural calcifications identified as the disease progresses, and arterial stenosis, occlusions and aneurysms later in the disease.
Large arteries can also be inflamed in a few other diseases (differential diagnosis). Examples include other types of vasculitis, mainly giant cell arteritis and polyarteritis nodosa; but also relapsing polychondriti, Cogan´s syndrome and Behçet`s Disease. Some infections can also cause inflammation in large arteries. The main CT differential diagnosis should also include common diseases such as atherosclerosis.
It is not an easy task to differentiate aortic calcification in TAK from that in atherosclerosis. Atherosclerotic plaques are more common in patients aged 45 years and above, and not usually associated with long segment luminal stenosis. Calcification in ascending aorta can be observed in some TAK patients, but it is rare in atherosclerosis. Giant cell arteritis shares similar pathogenesis and imaging features with TAK; however, giant cell arteritis commonly affects patients older than 50 years. In giant cell arteritis, branches of the external and internal carotid arteries are most frequently diseased. Polyarteritis nodosa frequently occurs in adults who are 30–50 years old, affecting males more than females, and it also more commonly affects patients with hepatitis B. Gastrointestinal and renal arteries are the primary sites diseased. Multiple small aneurysm formation in the involved artery is the characteristic manifestation on CTA images in polyarteritis nodosa.
Diffuse alveolar hemorrhage (DAH) is a severe and potentially fatal medical condition defined by the accumulation of red blood cells into the alveolar space,
In contrast to simple extravasation of erythrocytes facilitated by impaired hemostasis or hemodynamic causes, DAH in vasculitis is due to capillaritis (vasculitis in the lung microvasculature).
Pulmonary capillaritis may be primary as in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis or secondary to drugs (especially antithyroid drugs such as propylthiouracil), infections or connective tissue diseases, especially systemic lupus erythematosus.
The diagnosis of DAH is considered in patients who develop progressive dyspnea with patchy bilateral alveolar opacities on chest imaging (with density ranging from ground glass to consolidation) that cannot be explained otherwise.
DAH may manifest with hemoptysis, a valuable sign, that can be severe, and a drop in hemoglobin(HB) level (these cases are easier to diagnose). However, ~1/3 of patients don't have hemoptysis upon presentation, despite having blood in the lungs, and falling HB is supportive of DAH, but nonspecific. DHA may also manifest solely with pulmonary infiltrates and hypoxemic respiratory failure (it may mimic pneumonia, and theses cases are harder to diagnose).
Bronchoscopy with bronchoalveolar lavage is required, if the clinical situation of the patient allows, as it helps establish the diagnosis and rule out other entities. Bronchoscopy with serial lavage will confirm diagnosis of DAH (it even allows the diagnosis of pulmonary alveolar hemorrhage in those cases with subclinical presentation, as the presence of more than 20% of hemosiderophages in the bronchoalveolar lavage is diagnostic).The presence of hemosiderophages in the bronchoalveolar lavage is very typical, as the alveolar macrophages phagocyte the erythrocytes.
The lavage is also important to exclude infection, so fluid should be tested for viruses, bacteria, and possibly fungi, depending on presentation.
Once DAH is diagnosed and hemodynamic as well as infectious causes have been excluded, it is mandatory to search for anti-neutrophil cytoplasmic autoantibodies (ANCAs), as the pulmonary hemorrhage may be the first manifestation in ANCA-associated vasculitis (most commoly seen in small-vessel vasculitis, specifically microscopic polyangiitis (MPA) or Wegener granulomatosis (WG)).
Other syndromes exceptionally associated with DAH include Goodpasture syndrome, Churg-Strauss syndrome, Henoch-Schönlein purpura, Takayasu's arteritis, giant-cell arteritis, cryoglobulinemia, polyarteritisnodosa or Behçet's disease and systemic lupus erythematosus. Less commonly, DAH may be secondary to infection or drugs/toxins.

The diagnosis of DAH in our patient (with Takayasu arteritis) prompted us to search for other entities, rather than the vasculitis itself, as the causative agent. In fact, as discussed above, when associated with vasculitis, DAH is almost specifically seen in the small-vessel type (MPA and WG). The incidence of rupture and bleeding complications in TAK is very low, its typical finding the stenosis of the vessel (in 90% of patients). And as expected, in our patient with Takayasu arteritis, the DAH was due to infection(CMV pneumonia).