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Case Report

A Bacterial Pneumopathy Fortuitously Revealing a Vasculitis of Takayasu: A Case Report

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Abstract

Takayasu's vasculitis is a rare chronic inflammatory arteritis of unknown origin affecting large arteries in young subjects. We report the observation of a 32-year-old female patient who presented for 6 months with dyspnea stage II mMRC associated with an intermittent cough bringing back yellowish sputum with heaviness of the left upper limb and carotidodynia. The clinical examination revealed a blood pressure of 110/70 mmHg in the right upper limb and an impregnable blood pressure in the left upper limb. The left radial, ulnar, humeral and axillary pulses were abolished. There was pain on palpation of the right jugulocarotid axis and auscultation of the vascular axes revealed a right carotid murmur. Biological assessment showed microcytic hypochromic anemia with elevated ferritin level of 209.8 ng/mg, elevated CRP of 81.9 mg/L, positive procalcitonin of 0.3 ng/mL, and accelerated 1st hour SV of 13 mm. Chest X-ray showed a right paracardial opacity. Chest CT scan showed a middle lobar condensation focus. Thoracic and supra-aortic angioscan showed parietal thickening of the aortic arch, ascending aorta, and left subclavian artery with aneurysmal dilatation of the left carotid bulb and initial portion of the internal carotid artery and occlusion of the left axillary artery in favor of Takayasu type IIa vasculitis. The patient received antibiotic treatment and background treatment with antiplatelet agents, Prednisolone and Methotrexate. The evolution was marked by the negativation of procalcitonin after 72 hours of antibiotic therapy and the disappearance of the middle lobar condensation focus.

INTRODUCTION

Takayasu's vasculitis is a rare chronic inflammatory arteritis of unknown origin in young people that affects large arteries: the aorta, its main branches and the pulmonary arteries [1]. We report the medical observation of a patient with bacterial pneumopathy that fortuitously revealed Takayasu vasculitis.

PATIENT AND OBSERVATION

Mrs. L.Z, 32 years old, 6 months postpartum, treated for COVID-19 15 months ago.

She consulted for a dyspnea stage II mMRC evolving since 6 months associated with an intermittent cough bringing up yellowish sputum with heaviness of the left upper limb and a carotidodynia in a context of apyrexia and a decline of the general state made of asthenia and a weight loss of 18 kg.

The clinical examination revealed a pale patient with a performance status of 0. She was hemodynamically and respiratory stable, lean (BMI: 13.96 kg/m^2). Her blood pressure was unchanging in the left upper limb and 110/70 mmHg in the right upper limb. The left radial, ulnar, humeral and axillary pulses were abolished while the rest of the pulses were present. Pain was elicited on palpation of the right jugulocarotid axis and auscultation of the vascular axes found a right carotid murmur. Pleuropulmonary examination was unremarkable.

Chest radiography showed a sparse and inhomogeneous right paracardial opacity (Figure 1).

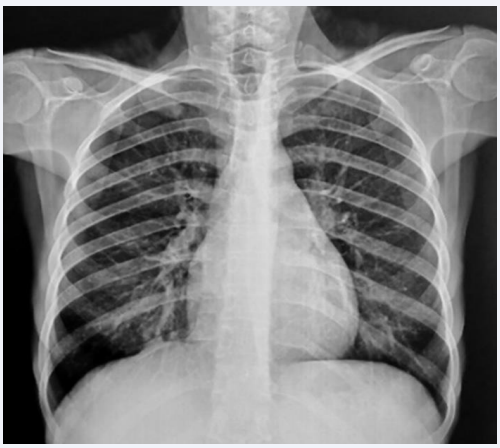

Figure 1 Frontal chest X-ray showing an opacity with right paracardial projection.

Chest CT scan showed a focus of middle lobar condensation (Figure 2).

The biological workup showed a microcytic hypochromic anemia (Hb: 7.2 g/dl, GMV: 70.4 fl, HDAC: 26.8 g/dl) with an elevated ferritinemia of 209.8 ng/mg, an elevated PCR of 81.9 mg/L, a positive procalcitonin of 0.3 ng/mL, and an accelerated sedimentation rate of 13 mm at the first hour.

Direct examination for BK, sputum GeneXpert and sputum cytobacteriological examination were negative.

The diagnosis of pneumonia of probable bacterial origin was retained. The patient was put on antibiotic therapy (Amoxicillin-clavulanic acid 3g/day for 10 days). Given the abolition of the pulses and the impenetrable blood pressure in the left upper limb as well as the carotidodynia, a Takayasu vasculitis was suspected.

In view of the suspicion of this vasculitis, the thoracic angioscanner and the supra-aortic trunks angioscanner showed a circumferential and regular parietal thickening of the aortic arch and the ascending aorta with an aneurysmal dilatation of the left carotid bulb and of the initial portion of the homolateral internal carotid artery parietal thickening of the left subclavian artery reducing its caliber and occlusion of the left axillary artery with persistence of a thin circulatory channel, circumferential parietal thickening of the left subclavian artery and occlusion of the left axillary artery with persistence of a thin circulatory channel.

In view of the suspicion of this vasculitis, the thoracic angioscanner and the supra-aortic trunks showed a circumferential and regular parietal thickening of the aortic arch and the ascending aorta with an aneurysmal dilatation of the left carotid bulb and of the initial portion of the homolateral internal carotid artery parietal thickening of the left subclavian artery reducing its caliber and occlusion of the left axillary artery with persistence of a thin circulating channel, circumferential and regular parietal thickening of the distal portions of the two common carotid arteries and stenosis of the two external carotid arteries, more pronounced on the left, circumferential and regular thickening of the left medio-basal and right postero-basal branches of the pulmonary artery in favor of Takayasu type IIa vasculitis (Figure 3).

In view of the fear of renal artery stenosis, the abdominal angioscanner performed did not show any lesion of the abdominal aorta or renal arteries.

Based on the ACR (1990) diagnostic criteria for Takayasu vasculitis, our patient validated 4 of 6 criteria, while she validated 2 major and 3 minor criteria of the Ishikawa (1988) classification modified by Sharma (1996). Thus, the diagnosis of Takayasu vasculitis was retained.

After consultation with the internists, the patient was put on antiplatelet agents 75 mg/day, Prednisolone 1 mg/kg/day for 1 month and then a decrease in dosage over 1 year and on Methotrexate 15 mg: 1 weekly injection for 4 consecutive weeks with adjuvant treatment (Calcium, Vitamin D, Potassium and Folic Acid).

The evolution was marked by the negativation of procalcitonin after 72 hours of antibiotic therapy and the disappearance of the middle lobar condensation focus (Figure 4).

DISCUSSION

Takayasu's vasculitis is the most frequent inflammatory arteritis affecting young people. It affects large-caliber arteries, mainly the aorta, its main branches and the pulmonary arteries. Its annual incidence varies between 2 and 3 cases per million inhabitants. Its prevalence is estimated at 40 per million inhabitants in Japan, while in the United Kingdom it is 4.7 per million inhabitants. It affects people under the age of 40. It is predominantly female, with an F/H ratio of 8/1 in Japan and 4.8/1 in France [1]. The cause of the disease remains unknown. There are several hypotheses: genetic hypothesis with exceptional cases of familial involvement [2]; infectious hypothesis on the relationship between tuberculosis and Takayasu vasculitis: a fortuitous association or a nonspecific inflammatory reaction triggered by a Mycobacterium tuberculosis antigen [3]; immune hypothesis leading to an autoimmune mechanism with antigenic stimulation [4].

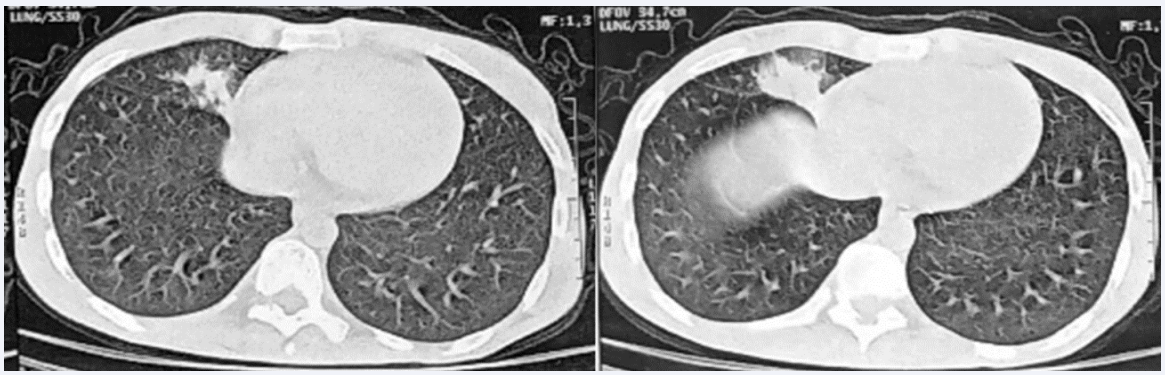

Figure 2 Chest CT scan showing a focus of middle lobar condensation.

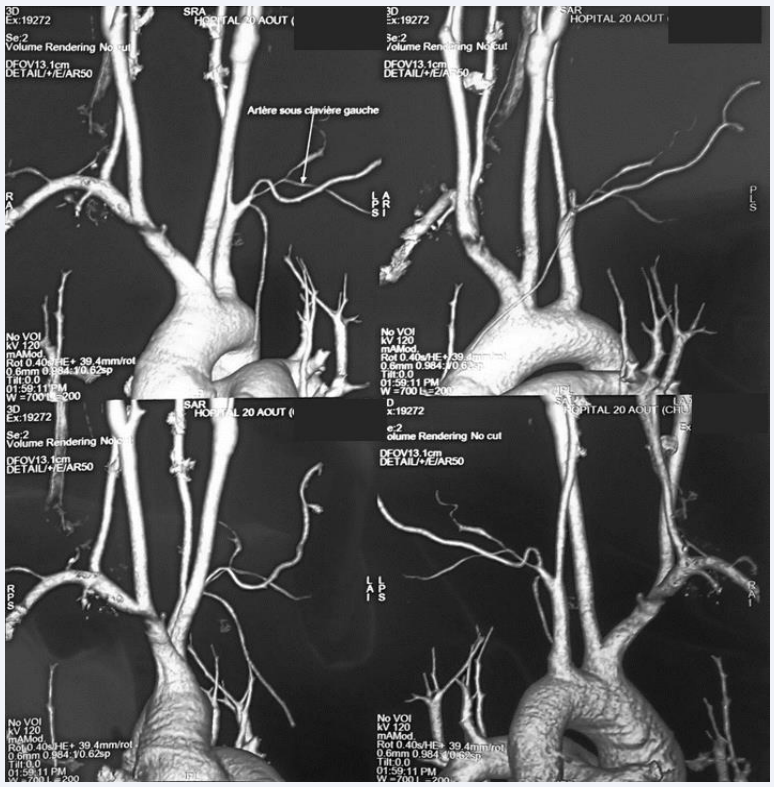

Figure 3 Thoracic and supra-aortic angioscan showing parietal thickening of the aortic arch, ascending aorta, left subclavian artery with aneurysmal dilatation of the left carotid bulb and initial portion of the internal carotid artery and occlusion of the left axillary artery in favor of Takayasu vasculitis type IIa

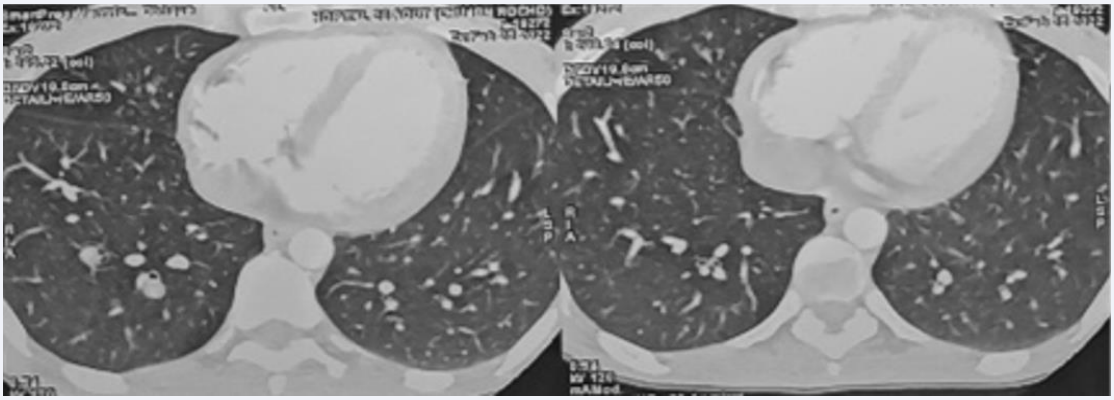

Figure 4 Control chest CT scan showing disappearance of the middle lobar condensation focus.

The evolution of the disease is in 2 phases: a systemic phase and an occlusive phase. In the systemic phase, general non-specific signs such as fever, night sweats, weight loss and arthromyalgia are prominent. Sometimes the diagnosis can be guided by ophthalmologic signs such as episcleritis and uveitis or cutaneous signs such as erythema nodosum, nodular hypodermitis and skin ulcerations. During the occlusive phase, the progression of vascular inflammation leads to the development of arterial aneurysms or stenoses [5].

To date, there are no specific biological markers or antibodies for diagnosis. The inflammatory syndrome is inconsistent, and there is no correlation between its absence and the absence of disease activity. Arterial imaging is the major element that allows for a positive diagnosis and monitoring of the evolution of the disease. Arterial Doppler ultrasound, CT angiography and MRI angiography are used to search for damage to the aorta and its branches. Positron emission tomography has been proposed to study the inflammatory activity of the arterial wall. Histology is

not systematically performed, but it allows the demonstration of a giganto-cellular panarteritis with a predominantly medial-adventitial pattern.

The diagnosis of Takayasu's vasculitis is made on the basis of a combination of clinical, biological, radiological and sometimes histological findings.

There are several classification criteria, the most frequently used being the American College of Rheumatology (ACR) criteria and the Ishikawa criteria modified by Sharma [6].

The location, elevational extension of the disease and severity of the aortic lesions are variable from patient to patient [7].

Treatment is based on corticosteroids: Prednisone (1 mg/kg/d for 1 month and then decreasing over 1 year); immunosuppressants such as Methotrexate (0.3 mg/kg/week), Azathioprine (2 to 3 mg/kg/day) or more rarely Cyclophosphamide; Biotherapies have also shown their effectiveness [3,6,8]. Surgical treatment is reserved for tight stenoses of the renal and pulmonary arteries, for aneurysmal lesions, in case of failure of angioplasty, or for large aortic replacements and major aortic valve insufficiency [7].

In France, survival is 95% at 5 years and 91% at 10 years [3]. The main causes of death are heart failure (46%), renal failure (11%), stroke (10%) and postoperative complications (6.4%). Arterial aneurysm ruptures can be fatal and result in death [1,3].

The particularity of our observation is the fortuitous discovery of Takayasu's vasculitis in its vascular phase following the occurrence of a bacterial pneumopathy.

CONCLUSION

Takayasu's vasculitis is a rare pathology, the detection by a simple clinical examination can delay or prevent the evolution towards dreadful complications such as arterial stenosis or aneurysm.

Management requires a rigorous clinical, biological and imaging evaluation. The treatment is mainly based on the combination of corticosteroids and immunosuppressants.

Contributions of the authors

Hajar Arfaoui: Responsible professor and 1st author; Soukaina Hallouli: Attending doctor and 2nd author; Nabil Bougteb: Resident doctor; Hasna Jabri: Responsible professor; Wiam Elkhattabi: Responsible professor; Hicham Afif: Head professor. All authors contributed to this work. They have read and approved the final version of the manuscript.

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