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Takayasu Disease: Revealed by Pulmonary Involvement

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Abstract **Case Report**

Takayasu's disease is a rare inflammatory arteritis of the large vessels affecting with predilection the aorta and its main branches. The presence of avascular territories, pulmonary infarcts and, in rare cases, pulmonary arterial hypertension, are the main pulmonary manifestations that can be observed, hence the interest of thoracic angioscanner. Positron emission tomography appears to be a highly sensitive tool for determining disease activity. The reference treatment includes corticosteroids associated with methotrexate in case of uncontrolled disease. Percutaneous transluminal angioplasty and sometimes revascularization surgery are necessary in case of critical ischemia or threatening aneurysm.

Keywords: Takayasu disease; Aortitis; Pulmonary involvement.

INTRODUCTION

Takayasu's disease, also called "pulseless women's disease", is a rare arterial disease (the ncidence is estimated at 3/1000 000 inhabitants). The diagnosis of Takayasu's disease is first based on the presence of arterial involvement of particular topography: involvement of the primary carotid arteries, the pulmonary arteries, the suprarenal and interrenal aorta. Other clinical arguments are female gender, young age, ethnic origin, general manifestations (erythema nodosum) or biological (inflammatory syndrome). The anatomopathological aspect of granulomatous vasculitis with a predominantly medial adventitial appearance distinguishes Takayasu's disease from other inflammatory arteriopathies [1].

PATIENT AND OBSERVATION

Mrs R F, 57 years old, of Moroccan origin, without toxic habits, poorly followed for arterial hypertension for 2 years, and who reported a right lumbar pain for one year admitted to the department of respiratory diseases for hemoptysis of small abundance associated with chest pain and dyspnea stage 3 MMRC, and in whom the clinical examination found crepitating rales at the two pulmonary bases and a left hemiplegia. Chest X-ray showed a right paracardial opacity of alveolar type and a bilateral large hilum appearance (Figure 1). The SV was accelerated to 95 mm in the first hour, the brain CT scan showed a left ischemic stroke, the thoracic angioscanner showed a segmental and sub-segmental pulmonary embolism (Figure 2). Abdominal angioscan showed stenosis of the right renal artery and the superior mesenteric artery (figure 3). The PET scan showed hyperfixation of the right subclavian artery, the right superior mesenteric artery, and the right superior lobar artery. The diagnosis of TAKAYASU disease was retained according to the Ischimawa criteria. The patient was put under anticoagulant treatment, inhibitor of the enzyme of conversion, Corticotherapy 30 mg/ day with progressive degression (after 6 weeks by 5 mg, then degression every 15 days by 5 mg) the evolution after 3 months of retrospect, was good, marked by the disappearance of the thoracic pain, the dyspnea and the hemoptysis, regulation of the blood pressure figures the thoracic angioscanner of control showed a disappearance of the pulmonary embolism without sequels.

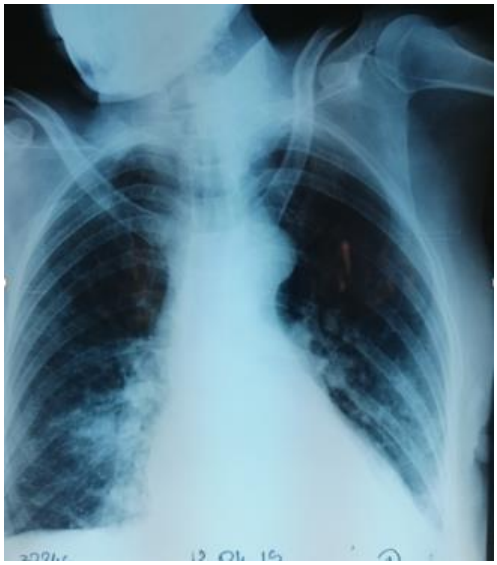

Figure 1: Chest X-ray showed a right paracardial opacity of alveolar type and a bilateral large hilum

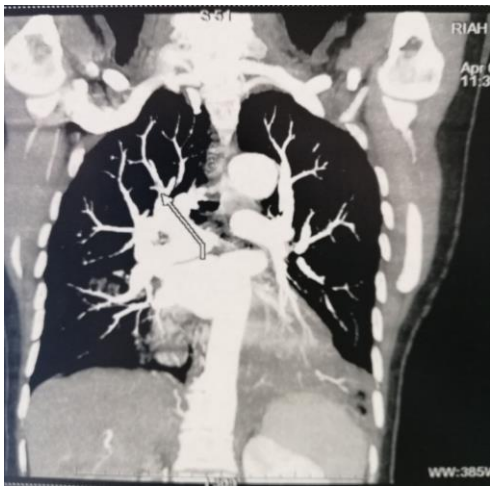

Figure 2: The thoracic angioscanner showed a segmental and sub-segmental pulmonary embolism

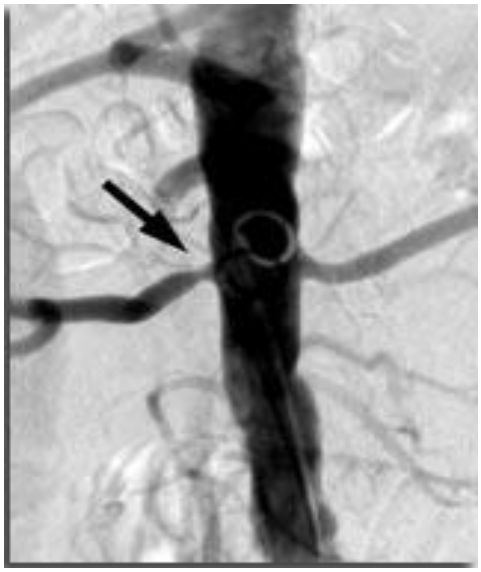

Figure 3: Abdominal angioscan showed stenosis of the right renal artery

DISCUSSION

Takayasu's disease or Takayasu's arteritis, is a rare, inflammatory disease affecting vessels (almost always large-caliber arteries: aorta and its branches, pulmonary arteries). The first case was described by Takayasu in 1905, and its incidence is less than one case per million per year. more common in Southeast Asia, India and South America. Takayasu's arteritis is a disease of the 2nd or 3rd decade. Only 10-20% of patients are older than 40 years at the time of diagnosis [11, 12]. With a female predominance of 62 to 97%. Takayasu's disease is rarely revealed by pulmonary involvement (33 cases reported in the Anglo-Saxon literature in 2001) [15, 16].

The pathophysiology of the disease is still not well understood, but 2 main hypotheses are classically put forward: an infectious origin could trigger an abnormal defensive reaction of the body, or an immunological origin.

An immunological origin is also evoked, given the association with certain HLA antigens in different populations. In Asia, the association with HLA B5, and in particular B52, is more frequent and is accompanied by a poorer prognosis. Takayasu disease is not a transmissible genetic disease or a contagious disease and family cases are more than exceptional. For the symptoms of the disease, it is classic to distinguish the acute period, known as the pre-occlusive phase, and the occlusive phase, characterized by ischemic manifestations [20]:

Pre-Occlusive Period

It associates non-specific general signs including 1 time out of 2 frustrating signs such as fever, arthralgias, fatigue, weight loss. Ocular signs (uveitis, episcleritis) or nodule-like skin ulcerations (erythema nodosum) are more suggestive of the disease. At the time of the outbreaks, other signs such as neck pain along the path of the carotid artery (carotinodynia) are very suggestive of the disease. Patients frequently complain of pain in the upper limbs with weakness or fatigability that is more pronounced from one side to the other [17]. Occlusive period with a time course of 3 years.Over time, some specific signs of the disease may appear. The clinical signs reflect the inflammation of the arteries. The importance of these clinical signs depends on the degree of inflammation of the arteries which can be narrowed (stenosis) or occluded. As the development of arterial narrowing is very slow, many patients are asymptomatic at the time of discovery of the disease. Thus, the diagnosis can be evoked when the blood pressure is taken, which is different between the 2 arms. Palpation of the pulses frequently reveals the absence of some of them [18].

Stroke and myocardial infarction are rare. Cardiac valvular damage should be sought. This is most often aortic insufficiency, which affects 10 to 25% of

patients. It is related to a dilatation of the aortic ring, often associated with a dilatation of the ascending part of the aorta. Retinal involvement is possible. Pulmonary arterial involvement is uncommon and is only exceptionally isolated [16]. It occurs in all types of arterial involvement [4]. Involvement of the pulmonary arteries is frequent (more than 40% of cases) if it is systematically sought by pulmonary scintigraphy (Fig 1) or arteriography [4]. The lesions are stenosis, occlusion or aneurysm of the pulmonary arteries or their branches (Fig 2). These lesions are often asymptomatic. When arterial damage is multiple or diffuse, the progression may be towards pulmonary arterial hypertension (PAH). The prevalence of PAH in Takayasu disease is estimated to be in the range of 7-14% [11, 17]. Exercise dyspnea, although most often of cardiac origin, chest pain and hemoptoic sputum may be the first manifestations [16]. Pulmonary obstruction may be complicated by pulmonary infarction (Fig. 3) with alveolar infiltrate and pleural effusions. Occlusion of a pulmonary artery may progress to parenchymal necrosis, with the appearance of excavations, putting patients at risk for aspergillosis grafting. Shunts between the pulmonary arteries and the systemic circulation (coronary, bronchial) have been frequently described (6% of patients on angiography) [21, 22]. The existence of shunts between the systemic circulation and the pulmonary arteries is consistent with diffuse involvement and proximal occlusion of the pulmonary arteries. These shunts would correspond to the development of bronchial arteries [13]. The richness of this collateral circulation can then be responsible for sometimes massive hemoptysis [25]. Other presentations may be encountered much more rarely: acute or chronic interstitial pneumonia, alveolar hemorrhage. Exceptionally, pulmonary involvement may be initially isolated (11 of 33 cases with initial pulmonary involvement), in which case venous thromboembolic disease is discussed [15, 16]. Only systemic signs (fever, arthralgias, inflammatory syndrome) may then suggest arterial inflammatory disease. However, they are not specific and lead to the discussion of Takayasu's arteritis, Behçet's disease, gigantocellular arteritis or syphilis. The discovery of aortic involvement can then represent a major diagnostic element. However, it may not appear until after several years of follow-up [16]. Finally, pulmonary involvement can be asymptomatic and justifies diagnostic exploration to differentiate genuine Takayasu disease from atherosclerosis, fibromuscular dysplasia, or other inflammatory arteritis. Finally, Takayasu disease may be responsible for hypertension, which is often difficult to control and is related to renal artery stenosis [3]. Neurosensory involvement Headaches (20-42% of cases), visual disturbances (19-28% of cases), malaise (16-47%), more rarely convulsions or ischemic strokes [8, 11, 13], ischemic retinopathy in 37-52% of patients [4, 13]. Associated with these clinical signs, biological abnormalities are observed. Thus, the inflammatory syndrome (elevated sedimentation rate or C-reactive protein) often present in the acute phase of the disease disappears spontaneously over time.Diagnosis is confirmed by imaging. Arterial Doppler ultrasound, angioscanner, MRI and arteriography can show stenoses, arterial occlusions or aneurysms. Above all, these non-invasive exploration techniques are evocative in showing inflammatory thickening of the wall of the aorta and its branches. An arteriography is exceptionally necessary, apart from interventional radiology procedures (see below) used in the first line and tend to supplant arteriography. Inflammation around the arteries is described, especially around the aorta (aortitis). The attacks are most often multiple and can give images called "candle flame or radish tail". Arterial aneurysms can also be observed. The PET-scanner (positron emission tomography) is increasingly performed to establish the diagnosis of the disease. It allows to obtain a cartography of all the affected vessels showing inflammatory zones of intense fixation of a radioactive tracer. Its use could allow to evaluate the activity of the disease and would constitute an interesting tool for follow-up [4]. The most frequently used diagnostic criteria sACR classification criteria [22] Age of onset < 40 years Vascular claudication of the extremities, especially in the upper limbs Decrease in at least one brachial pulse Systolic blood pressure difference > 10 mmHg between the two arms Murmur on a subclavian artery or abdominal aorta On arteriography, stenosis or occlusion of the aorta or its branches or proximal limb arteries; Abnormalities usually focal or segmental, not related to arteriosclerosis or fibromuscular dysplasia The presence of three or more criteria provides a sensitivity of 90.5% and a specificity of 97.8% for the diagnosis of Takayasu diseaseare those of the American College of Rheumatology (ACR) and those of Ishikawa modified by Sharma [5]. Histological diagnosis is less and less necessary for the diagnosis due to the progress in imaging. Symptomatic medical treatment is based on corticosteroid therapy, the primary aim of which is to reduce the inflammation of the arteries. This treatment is effective in more than half of the cases and is usually started at a dosage of 1 mg/kg/day and maintained at this dose for 4 to 6 weeks. Thereafter, depending on the evolution of the disease, the dosage is progressively decreased according to the clinical response. The usual duration of treatment is 12 to 24 months [5]. In rebellious forms of the disease or those requiring very high doses of cortisone, the addition of an immunosuppressive treatment is the rule. These treatments are classically methotrexate (20-25 mg/kg per week), azathioprine (2 mg/kg per day), mycophenolate mofetil (2 g/day) or, more rarely, cyclophosphamide. The use of more recent treatments called biotherapies, primarily anti-TNF (Tumor Necrosis Factor) or anti-interleukin-6 receptors, is possible in the presence of uncontrolled disease. Treatment for hypertension and cortisone-induced osteoporosis is essential [23]. To correct vascular damage (revascularization) and improve symptoms, it is

sometimes necessary to resort to surgical or interventional radiology treatments. Arterial bypass surgery, deobstruction and vascular prosthesis are sometimes proposed [26, 27].

CONCLUSION

TAKAYASU disease is a rare vasculitis whose diagnosis and follow-up are difficult because of the absence of a specific marker, and can be the cause of formidable complications such as neurological damage, hence the importance of early detection. Pulmonary involvement is rarely revealing but should be systematically investigated by pulmonary ventilation-perfusion scintigraphy and angioscanner. However, the use of new imaging techniques such as angiomicrography and PET/CT is promising both in the diagnosis and follow-up of these diseases. Finally, there are no randomized controlled studies concerning the treatment of this vasculitis. Thus, corticosteroids are still essential in combination with another immunosuppressant for TAKAYASU disease.

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