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Case Report

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UNUSUAL OCULAR MANIFESTATION REVEALING TAKAYASU'S ARTERITIS: ABOUT A CASE

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ABSTRACT

Takayasu's arteritis (TA) is a chronic inflammatory granulomatous vasculitis that affects large and medium arterial vessels. The disease involves especially subclavian arteries and aortic branches, but it can affect of any artery. The major pathology is granulomatous panarteritis with intimal proliferation and defects the elastic lamina of the vessels. The diagnosis of TA is a challenge, particularly in its initial phase. We report here the case of a patient with undiagnosed takayasu arteritis associated with pulmonary tuberculosis who presented with chronic unilateral inflammatory exophthalmos, and a subsequent diagnosis of an inflammatory pseudotumor of the orbit. In this case, we believe that extensive blood vessel involvement may be related to pseudotumor formation in Takayasu's arteritis. Because this is an unusual and unreported presentation of the disease, a better estimate of the causal relationship may be possible in the future with additional information. In conclusion, although uncommon, this case highlights that an orbital pseudotumor may be an important finding in Takayasu arteritis. For early diagnosis, better treatment, and a good prognosis, it should be considered in patients presenting with ocular symptoms similar to those of the other vasculitis.

KEYWORDS: Takayasu arteritis; Orbital pseudotumor; Orbital vasculitis; pulmonary tuberculosis.

ABBREVIATIONS

TA: Takayasu's arteritis.
CT: Computer Tomography, tomodensitométrie
MRI: Magnetic Resonance Imaging

INTRODUCTION

Takayasu's arteritis (TA) is a chronic inflammatory granulomatous vasculitis that influences large and medium arterial vessels. It is an unusual and idiopathic disease that affects the aorta and its branches and causes constriction and obliteration.[1,3] The disease involves especially subclavian arteries and aortic branches but can affect any artery. the pathophysiology of the disease remains unknown. The major pa-thology is granulomatous panarteritis with intimal proliferation and defects of the elastic lamina of the vessels.[4] Orbital pseudotumors are a rare disorder of the orbital tissue, which encompasses a wide range of pathological processes and has different etiologies and prognoses. The inflammatory process in the orbit may have variable clinical features such as myositis, dacryoadenitis, perineuritis, episcleritis, and a localized mass. The most frequent diagnoses are infection, vasculitis, thyroid dysfunction, sarcoidosis, and neoplasia.[5] Recently, IgG4-related disease has been increasingly reported as a cause of orbital pseudotumor, due to increased knowledge and awareness.[6] Orbital pseudotumor is not a rare associa-tion in rheumatologic disorders. Among ANCA-associated vasculitis, granulomatosis with polyangiitis is more frequently associated with retro-orbital granulomas, which cause serious complications such as vi- sion loss, orbital, and facial deformity.[7] Here, we report the case of a 40-year-old woman who presented with chronic unilateral inflammatory exophthalmos. She eventually received a diagnosis of Takayasu arte- ritis, which is associated with pulmonary tuberculosis and an orbital pseudotumor.

OBSERVATION

A 35-year-old female presented with a chronic exophthalmos of progressive aggravation that had been evolving for 9 months. The ophthalmological examination found in the right eye: a visual acuity with negative light perception; a non-axial, painful, inflammatory, non-reducible, non- pulsatile exophthalmos; hypotropia; a chemosis; Infectious Keraritis; ocular hypertonia; the rest of the examination was hampered by the state of the media **[figure 1]**. In the left eye, the visual acuity was 10/10, Slit lamp examination and tonometry were normal.

Biological and radiological examinations revealed: a negative inflammatory balance; negative serologies (toxoplasmosis, CMV, rubella, syphilis, HIV); normal thyroid test; a normal converting enzyme; and a negative tuberculin test. The patient underwent an orbital MRI, which showed a right extra conical process suggestive of lymphoma or an inflammatory pseudo tumor **[figure 2]**.

Biopsy was inconclusive, the decision was made to perform a minimally invasive exenteration for diagnostic purposes **[figure 3]**; the anatomopathological results with additional immunohistochemistry were in favor of a pseudo-inflammatory process without any specificity, and without any arguments in favor of a lymphomatous process **[figure 4(A - B)]**. The patient received a bolus of corticoids. A follow-up MRI after 6 months showed a progression of the intra- orbital tissue process in comparison with the initial MRI **[figure 5].** A CT scan performed showed obvious thickening of the walls of the aorta, renal artery, and superior mesenteric artery in favor of Takayasu disease with the presence of pulmonary micronodules related to a specific pulmonary infection (probably tuberculosis) **[figure 6,7(A - B)].**

Fibroaspiration was performed in the presence of pulmonary noduleswhich showed the presence of M. tuberculosis, confirming active tuberculosis in the patient. The presentation was consistent with Takayasu disease revealed during the workup of inflammatory exophthalmos. The patient started antibacillary therapy, declared cured, and then put on oral corticosteroid therapy for her Takayashu disease, with good improvement.

FIGURES

Figure 1: Frontal photographic image showing the clinical aspect of the affected eye.

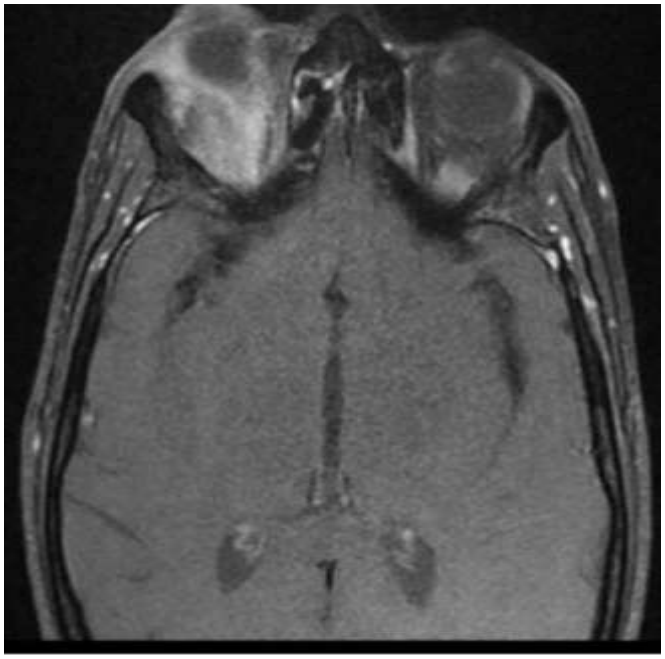

Figure 2: Orbital MRI, axial section, showing a right extra conical process suggesting a lymphoma or an inflammatory pseudo tumor.

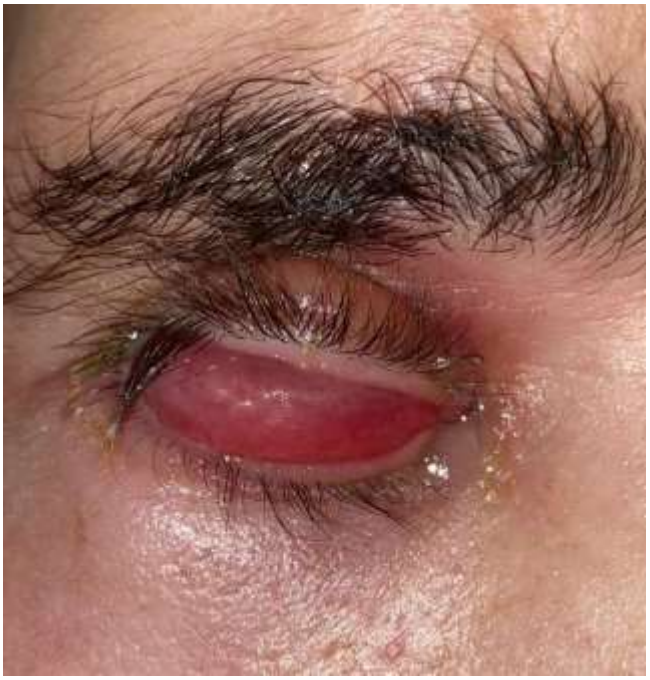

Figure 3: Front view photograph showing the clinical appearance of the affected eye after surgery.

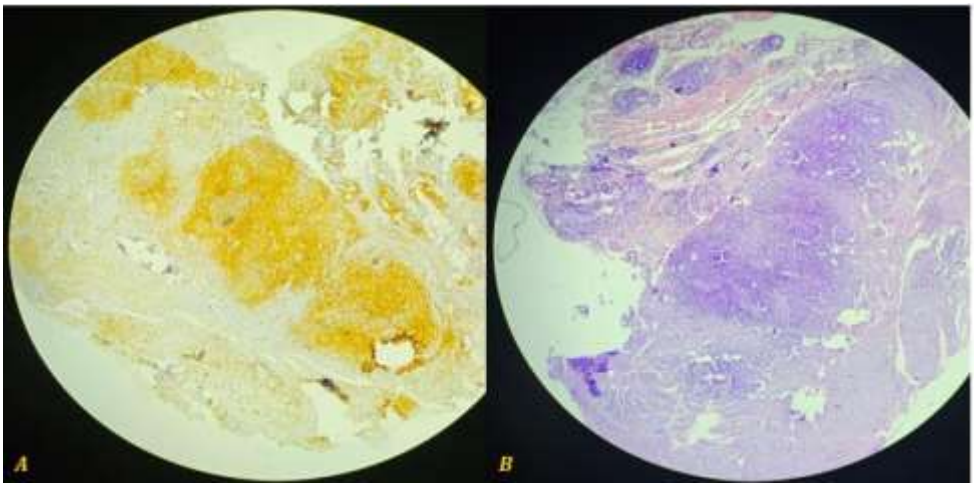

Figure 4 (A and B): Histological section showing a pseudo-inflammatory process with no specificity, and no arguments in favor of a lymphomatous process.

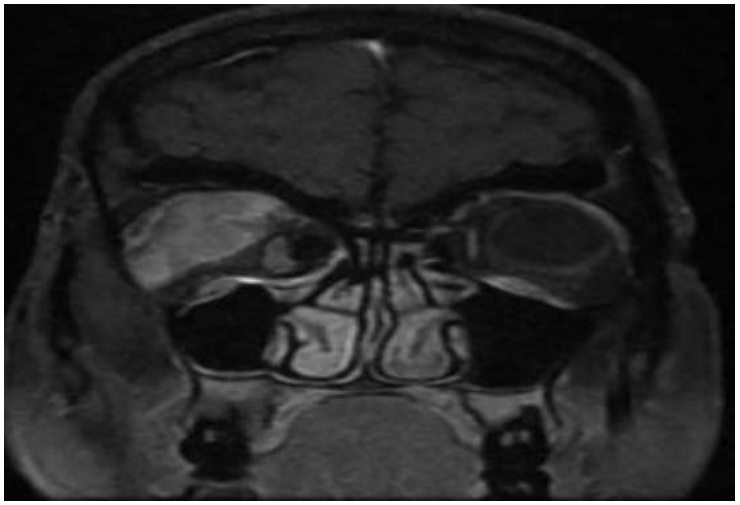

Figure 5: MRI of the orbit, frontal section showing the progression of the intra-orbital tissue process in comparison with the initial MRI.

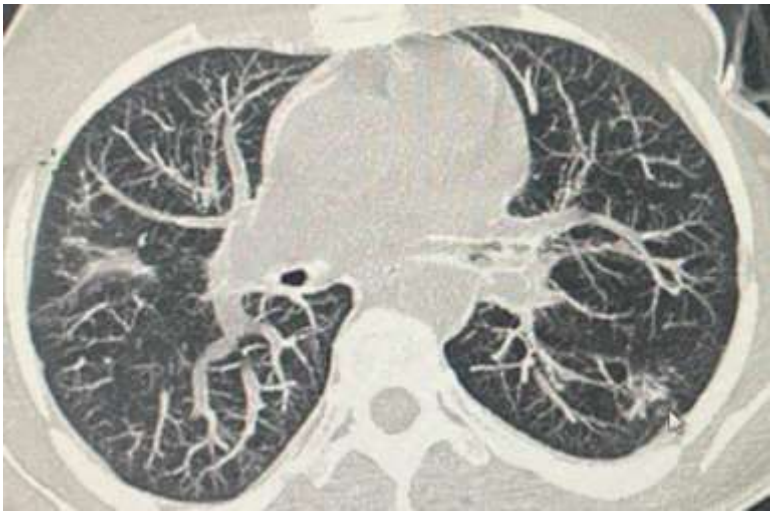

Figure 6: Parenchymal axial scan of the lung showing obvious thickening of the walls of the aorta, renal artery and superior mesenteric artery in favor of Takayasu's disease with the presence of pul- monary micronodules related to a specific pulmonary infection.

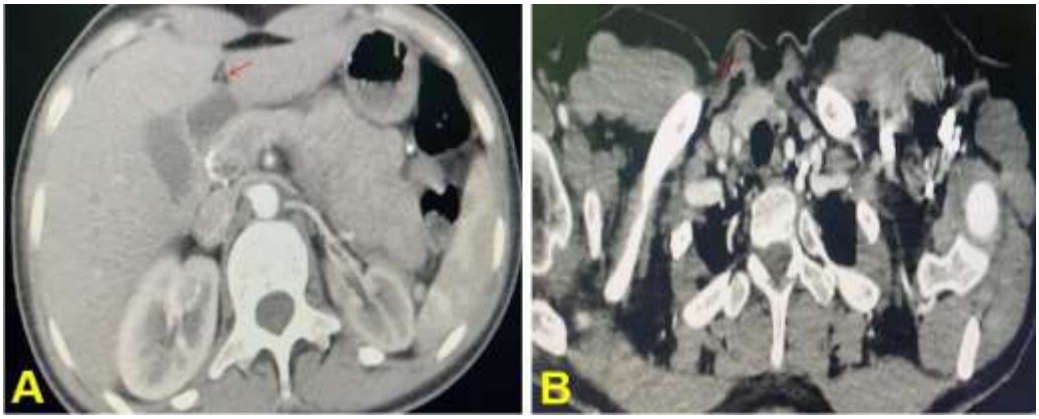

Figure 7(A and B): Axial sections of the Angioscanner (passing through the abdomen): various non-obstructive stepped stenoses concerning the arteries mentioned, thus confirming the diagnosis of Takayasu disease.

DISCUSSION

Takayasu´s arteritis is an inflammatory vascular disease affecting more females than males. It usually begins in the second or third decade of life and involves primarily the large arteries. It was first reported in 1908 by Takayasu, a Japanese ophthalmologist.[8]

The diagnosis of TA is a challenge, particularly in its initial phase. In fact, systemic symptoms occurred in only a third of patients, the erythrocyte sedimentation rate lacks sensitivity, and there is no diagnostic serologic test.[9] According to the American College of Rheumatology[10], three of the following criteria are mandatory to retain the diagnosis of TA: onset before 40 years of age, decrease in the brachial pulse in one or both arms, claudication of the extremities, audible bruit on auscultation of the aorta or subclavian artery, a difference of 10-mmHg or more in blood pressure measured in both arms, and narrowing at the aorta or its primary branches on an arteriogram. Ocular changes in TA were the consequence of the progressive narrowing of major vessels of the neck and resultant ocular hypoperfusion. The most frequent manifestation was Takayasu´s retinopathy, which had been classified into four stages by Uyama and Asayma.[11]

The originality of our case lies in this particular association. To our knowledge, no case has been reported in the literature associating both a unilateral pseudo-inflammatory tumor with takayasu's disease and pulmonary tuberculosis, hence the particularity of our clinical case. Several explanations for how tuberculosis triggers Takayasu disease have been proposed, but the exact mechanism and relationship between Takayasu and tuberculosis remain to be elucidated.

A Korean study showed that the incidence of tuberculosis in Takayasu patients (17%) was higher than that of the general population (5.5% to 5.8%).[12] Some studies have reported the concomitance of Takayasu arteritis and tuberculosis, and have suggested that anti-inflammatory therapy should be initiated cautiously after the control of active tuberculosis.[13,15]

As this is a very rare presentation of Takayasu's arteritis, we do not know precisely which features of the disease

can predict the development of orbital pseudotumors. Temporarily, in the presented case, prevalent alterations of blood vessels, including the pulmonary arteries, are suggestive of extensive involvement and a relatively late phase of the disease. In addition to post- contrast enhancement of the vessel walls, MRI confirmation of an active disease state may be related to orbital inflammation. Further case reports may help to understand this relationship more precisely.

Treatment of TA is based on the use of anti-inflammatory therapy like steroids and/or methotrexate to decrease or to stop the inflammatory activity. Recently, Tocilizumab, an interleukin-6 receptor monoclonal antibody, has proven its effectiveness in the treatment of TA and may reduce the incidence of relapses.[16] Both surgical and endovascular treatments may be risky and achieve poorer outcomes if they are undertaken during the period of inflammatory activity.[17]

CONCLUSION

Although uncommon, this case highlights that an orbital pseudotumor may be an important finding in Takayasu arteritis. It should be considered in patients with inflammatory exophthalmos and other nonspecific ocular symptoms in Takayasu arteritis. Tuberculosis has been implicated in the etiopathogeny of this vasculitis, but without tangible proof. Our observation showed the interest of a rigorous etiological assessment in front of any inflammatory exophthalmos and also disclosed this correlation between tuberculosis and Takayasu disease.

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