Case study

A Case Report of Takayasu's Arteritis With aortic insufficiency As Initial Presentation.

Abstract :

Takayasu's disease is the first cause of inflammatory aortitis in young subjects.
The disease preferentially affects the women.

Takayasu's disease could be a mode of non-specific reaction of the aorta to various infectious or dysimmune etiological factors.

Aortic insufficiency is present in 7 to 10% of cases. It is usually moderate.
Its mechanism is not unambiguous: direct damage to the aortic valves or dilation of the aortic annulus.

Coronary damage responsible for angina or even myocardial infarction are also possible.

We report the case of a severe aortic insufficiency secondary to takayasu disease complicated by acute coronary syndrome in a 42-year-old woman.

Key words :

Takayasu's disease, severe aortic insufficiency, acute coronary syndrome, case report.

Introduction :

Takayasu's arteritis is a vasculitis of the large arterial trunks.

This disease occurs before the age of 50 and mainly affects young women.

This vascular inflammatory disease is preferentially located in the aorta and the large supra-aortic trunks.

It is complicated by aneurysms or arterial stenosis and rarely (as in our case) moderate or severe aortic insufficiency.
Diagnostic confirmation of Takayasu's arteritis is provided by a beam of arguments based on age and sex, clinical findings, and imaging evidence including damage to the aorta and/or its branches with circumferential thickening of the arterial wall with stenosis and sometimes aneurysmal dilatation, often multifocal.
The inflammatory syndrome is common at the onset of the disease and should be sought by C-reactive protein assay (CRP) and the fibrinogen level or the sedimentation rate, which will then be used for the monitoring of disease activity under treatment.

Case presentation :

We report the case of a 42-year-old woman, with no particular pathological history: in particular, no toxic habits, neither hypertensive nor diabetic nor known dyslipidemia.

The patient presents to the emergency room following the onset of aggravating dyspnoea.
The clinical examination in the emergency room found a patient with NYHA stage 3 dyspnea, with normotensive 130/62 mmHg, heart rate at 70 bpm, normopnea at 18 cycles per minute.
On pulmonary auscultation, crackles were found at the base, with a saturation of 94% in free air.
There was also edema of the lower limbs down to the ankles, as signs of global heart failure.

A peripheral venous line was placed and a bolus of 60mg of furosemide was administered urgently then relayed orally.

Electrocardiogram (ECG) performed in the emergency room showed Left ventricular hypertrophy with secondary repolarization disorders (figure1).

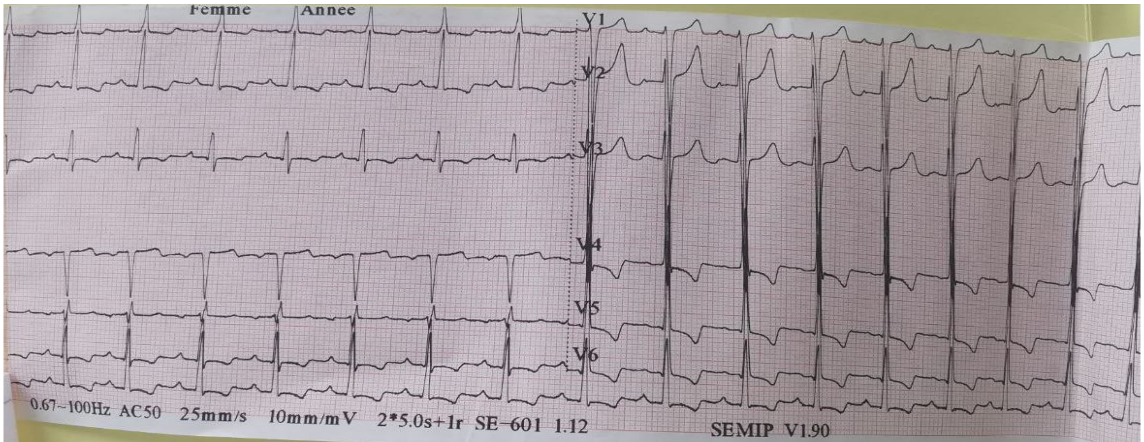

Figure 1 : ECG which showed Left ventricular hypertrophy with secondary repolarization disorders

The patient was hospitalized in the cardiology department of the CHU Ibn Rochd in Casablanca, Morocco.

Faced with this presentation and after the stabilization of the decompensation, the patient benefited from a complete clinical examination, and biological assessment as well as echocardiography to label the factors of decompensation.

The clinical examination found a notion of arthralgia in the left shoulder and elbow, with no other associated signs.

In addition, the biological assessment objectified an inflammatory syndrome made of inflammatory anemia at 8.4 g / dl with decrease in serum iron level at 0.17mg / l, and ferritin at 8mg / l, with increase in CRP level at 38 mg /l and VS at 116 mm.

The echocardiographic probe objectified a dilated left ventricle, seat of global hypokinesia with ejection fraction at 40%; thin valves in the tricuspid aortic valve, seat of severe aortic insufficiency (PISA=16mm; VR: 137; SOR: 30; PHT: 100 ms) (figure 2), with notion of dilation of the ascending aorta (21mm/m²).

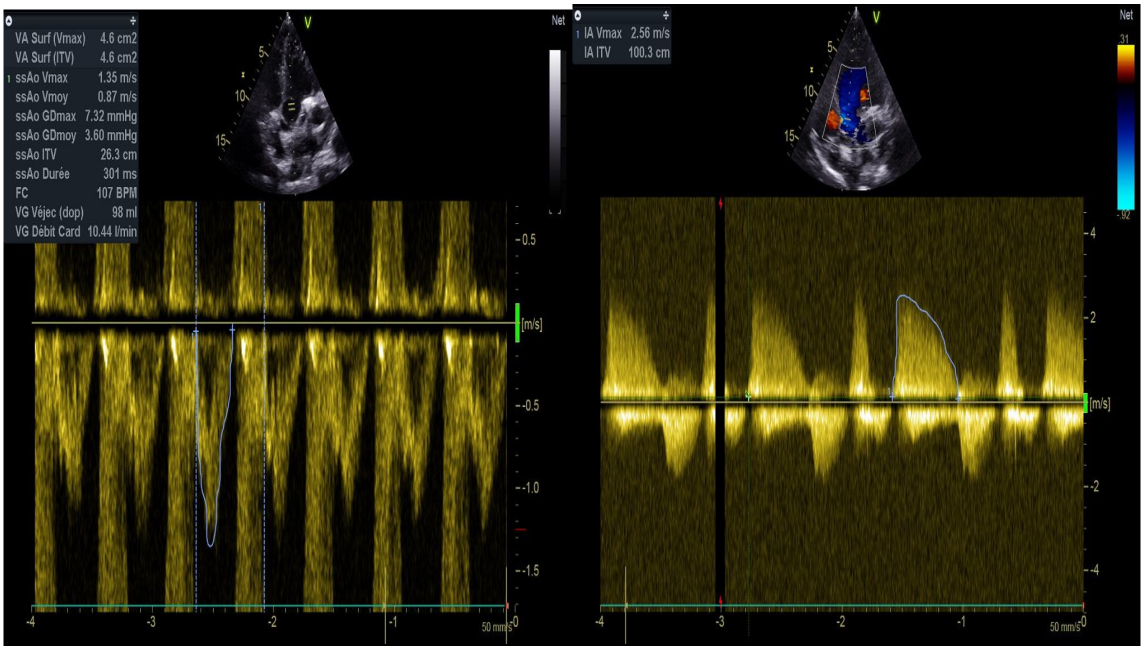

Figure 2 : Severe aortic insufficiency

The action to be taken was completed by thoracic CT angiography, objectifying the appearance of an aortic ectasia of segments 0 and 1, without detectable aneurysmal image, with inflammatory parietal thickening (figure 3).

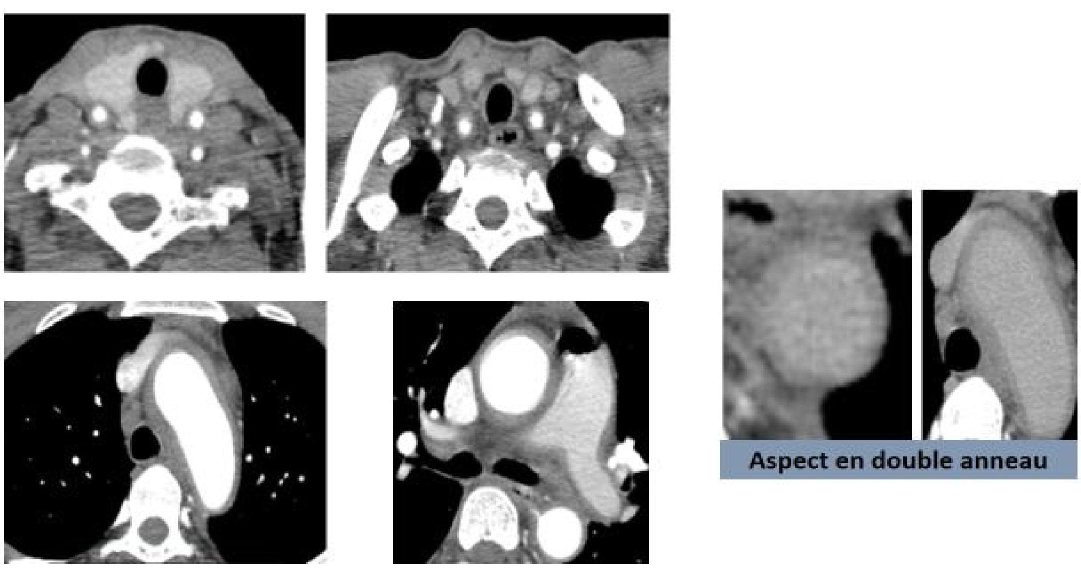

Figure 3 : Thoracic CT angiography showing parietal thickening of the aortic wall with a double ring appearance.

According to this scannographic aspect, clinical data (sex and age and a particularly normal clinical examination) and ultrasound (fine valve); aortic insufficiency of very probable inflammatory origin was concluded (1).

The immunological assessment and the viral and syphilitic serologies are negative.
An ultrasound of the supra-aortic trunks was made showing an aspect of diffuse circumferential parietal thickening of the supra-aortic trunks in a halo aspect (figure 4).

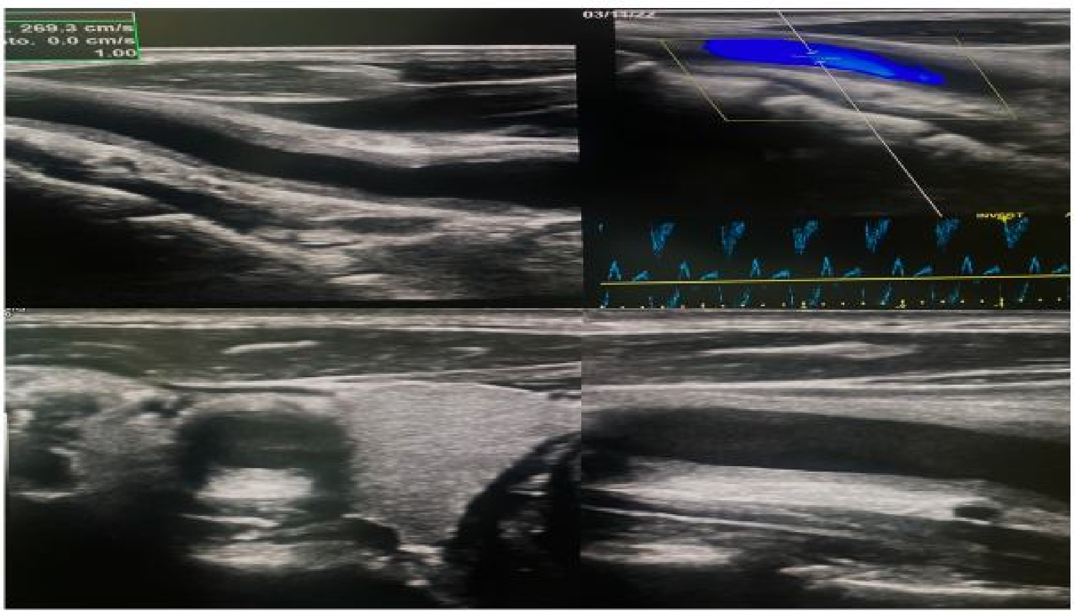

Figure 4 : Ultrasound of the supra aortic trunks showing a halo aspect.

In view of these investigations, the diagnosis of very probable takayasu disease was retained; due to the following criteria:

- Female sex.
- Young age of 42 years old.
- Absence of Cardiovascular Factors (atherosclerosis)
- Aortic location
- Aspect of halo at the level of the aorta and supra-aortic trunks.

• Immunological assessment and viral and syphilitic serologies: negative.

Subsequently, we completed a PET scan showing hyperactivity of the arterial wall of the arch of the aorta, the ascending thoracic aorta and abdominal aorta (figure 5).

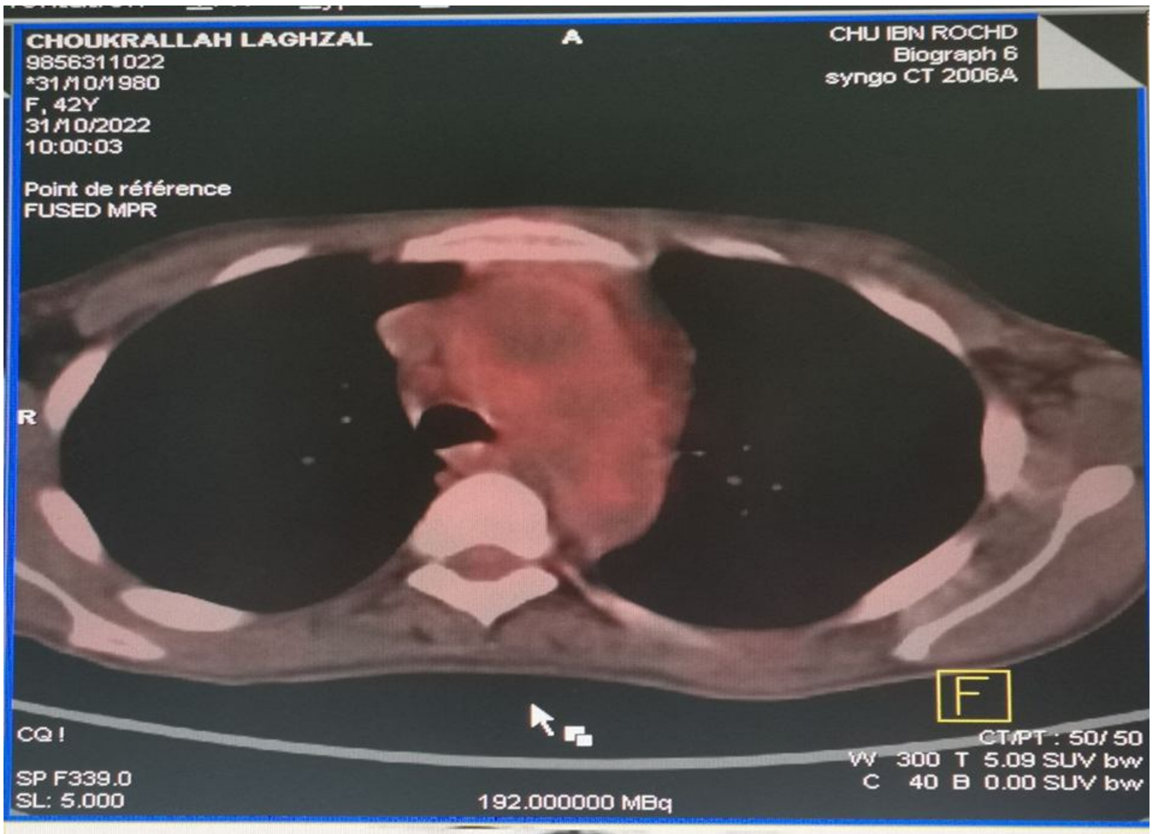

Figure 5 : A PET scan showing a hyperactivity of the arterial wall of the arch of the aorta and the ascending thoracic aorta.

Therapeutically, the patient benefited from corticosteroid therapy based on: 1mg/kg/d associated with adjuvant treatment with anti-platelet aggregation : aspirin 100 mg per day.

The follow-up at 7 days showed under this treatment a biological improvement of the inflammatory syndrome:

*CRP: 24.2mg (11/2/2022)
*CRP: 14.7mg (11/4/2022)
*CRP: 3mg (11/7/2022)

The evolution of the patient was marked by the typical chest pain on 04/11/2022 (after 15 days of his diagnosis).

- On the electrocradiogram level (figure 6): No electrical modification except negative T wave in V3.

- Biologically: Troponins at H3 8.6 Vs 693.5 at H12.

- <u>Echocardiogram</u> check: non-hypertrophied dilated LV, seat of global hypokinesia, LVEF= 40.

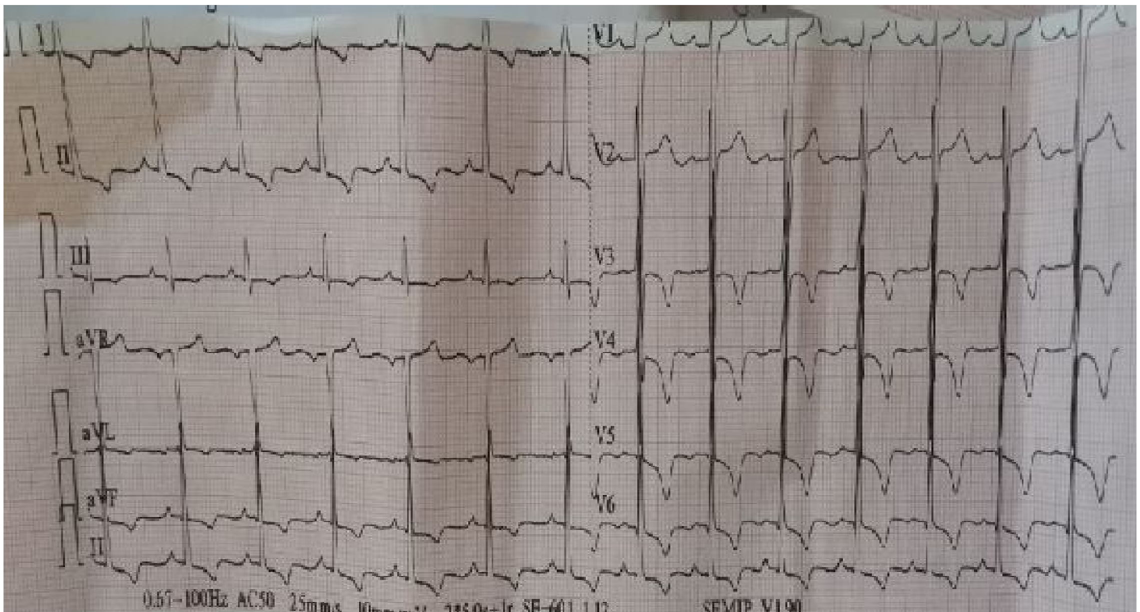

Figure 6 : ECG with No electrical modification except negative T wave in V3.

Therefore The diagnosis of high-risk NSTEMI was retained.

Patient received loading dose and started treatment for Ischemic Cardiomyopathy.

The coronary angiography was done and did not reveal any anomalies (figure7).

Figure 7 : normal coronary angiography

DISCUSSION :

Takayasu's arteritis is a chronic inflammation of the large arteries such as the aorta and its primary branches, causing progressive arterial occlusion. This leads to reduced blood flow in the limbs and organs, resulting in arm or leg claudication, diminished or absent peripheral pulses, and end-organ ischemia.

It affects women more than men, typically presenting between 10 and 40 years(2), as seen in our patient.

It has a variable clinical presentation, ranging from asymptomatic disease and constitutional features to symptoms of end-organ damage.

On the cardiac side : Takayasu's arteritis can result in miscellaneous damage. The frequency of the cardiac damage during the disease of takayasu is 40%, and constitutes a criterion of severity (3).

Isolated aortitis is a rare form of vasculitis that is associated with significant morbidity and mortality through the development of unstable aneurysms, aortic rupture, dissection, and thrombotic luminal occlusions (4).

The frequency of coronary involvement varies according to the series (5 to 45%) (5). It mainly results from ostial damage. (6).

And we can see coronary involvement without an angiographically detected lesion (as in our case) (7).
Coronary involvement sometimes poses a therapeutic problem in terms of revascularization, especially during an inflammatory flare-up(8).

It may manifest itself as valvular damage, including aortic insufficiency; secondary either to annular dilation following an aneurysm of the ascending aorta, or by retraction of the aortic cusps(9).

Discovery by angiography of the ascending aorta (4.2%), or by echocardiography (27.3%). (10)

Positron emission tomography/computed tomography (PET/CT) is a useful imaging modality to detect early lesions and active inflammation in the vessels. (11)

The American College of Rheumatology criteria 2022 is commonly used for the diagnosis of Takayasu's Arteritis (12): Age ≤60 years at diagnosis and imaging evidence of large-vessel vasculitis were absolute requirements to classify a patient as having Takayasu arteritis.

The final criteria items and weights were as follows: female sex (+1), angina (+2), limb claudication (+2), arterial bruit (+2), reduced upper extremity pulse (+2), reduced pulse or tenderness of a carotid artery (+2), blood pressure difference between arms of ≥20 mm Hg (+1), number of affected arterial territories (+1 to +3), paired artery involvement (+1) and abdominal aorta plus renal or mesenteric involvement (+3). A patient could be classified as having Takayasu arteritis with a cumulative score of ≥5 points.

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CLASSIFICATION CRITERIA FOR **TAKAYASU ARTERITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify the patient as having Takayasu arteritis when a diagnosis of medium-vessel or large-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

ABSOLUTE REQUIREMENTS	
Age ≤ 60 years at time of diagnosis	
Evidence of vasculitis on imaging[1]	
ADDITIONAL CLINICAL CRITERIA	
Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit[2]	+2
Reduced pulse in upper extremity[3]	+2
Carotid artery abnormality[4]	+2
Systolic blood pressure difference in arms ≥ 20 mm Hg	+1
ADDITIONAL IMAGING CRITERIA	
Number of affected arterial territories (select one)[5]	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Symmetric involvement of paired arteries[6]	+1
Abdominal aorta involvement with renal or mesenteric involvement[7]	+3

Sum the scores for 10 items, if present. A score of ≥ 5 points is needed for the classification of TAKAYASU ARTERITIS.

1. Evidence of vasculitis in the aorta or branch arteries must be confirmed by vascular imaging (e.g., computed tomographic/catheter-based/magnetic resonance angiography, ultrasound, positron emission tomography).
2. Bruit detected by auscultation of a large artery, including the aorta, carotid, subclavian, axillary, brachial, renal, or iliofemoral arteries.
3. Reduction or absence of pulse by physical examination of the axillary, brachial, or radial arteries.
4. Reduction or absence of pulse of the carotid artery or tenderness of the carotid artery.
5. Number of arterial territories with luminal damage (e.g., stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography from the following nine territories: thoracic aorta, abdominal aorta, mesenteric, left or right carotid, left or right subclavian, left or right renal arteries.
6. Bilateral luminal damage (stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography in any of the following paired vascular territories: carotid, subclavian, or renal arteries.
7. Luminal damage (stenosis, occlusion, aneurysm) detected by angiography or ultrasonography involving the abdominal aorta and either the renal or mesenteric arteries.

Chart 1 : Classification criteria for Takayasu Arteritis

and biologically, anemia as part of an inflammatory syndrome is a complication observed in 48 percent of patients with takayasu disease (13) (as in our case).

Initial treatment strategies should include a combination of high-dose Prednisone (initial dose of 0.5-1 mg/kg/day) and aspirin at an anti-platelet aggregation dose: 100mg per day (14).

CONCLUSION:

Takayasu's disease represents a very interesting clinical entity from a diagnostic, therapeutic and prognostic point of view.

Takayasu's disease can cause various cardiac manifestations through different mechanisms, as mentioned above.

This case is one of the rare cases of cardiac involvement secondary to Takayasu's disease, which should lead to a better diagnostic strategy and therapeutic management.

Declarations :

Consent for publication :

Written informed consent was obtained from the patients for publication of this case report and any accompanying images.

Availability of data and material :

All data generated or analysed during this study are included in this published article.

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