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Bilateral ocular ischemic syndrome as a manifestation of Takayasu arteritis in children

Takayasu arteritis (TA) is a chronic, autoimmune, granulomatous, inflammatory disease of the aorta and its major branches at their origin, which results in dilatation, occlusion, stenosis, and (or) aneurysm formation of the affected arteries.[1,2]

TA could be seen in different races, but the incidence is higher in the Southeast Asian population. The onset of disease is most frequent under 40 years old, generally in the third and fourth decade of life, and there is a predilection for women.[3,4]

TA in childhood could affect young infants to late adolescents, with the youngest patient diagnosed at 6 months old. It is reported as the third most common cause of vasculitis in the pediatric patients and is the only large vessel vasculitis in this group.[3,4]

The incidence of TA in the pediatric population has been estimated at 1.2 to 2.6 million per year.[4,5] Around 5% of patients with TA are children and adolescents. The majority of children are diagnosed between 8 and 13 years old, and as well as adults, there is a predilection for female patients, with a ratio of 3:1.[4]

We present a case of a pediatric patient who was diagnosed with bilateral ocular ischemic syndrome (OIS), as an initial manifestation of childhood Takayasu arteritis (c-TA).

Case report

A 12-year-old Peruvian female complained of headache, several episodes of left eye amaurosis fugax, right hemiparesis, with spontaneous recovery, and bradylalia for 15 days. On examination, visual acuity was 6/6 in both eyes (BE). Anterior segment and gonioscopy were within normal limits. Dilated retinal examination revealed venous dilation, some arteriolar narrowing, cotton wool spots, and widespread microaneurysm formation in BE (Fig. 1). Fluorescein angiography (FFA) showed patchy choroidal filling, delayed arm-to-retina circulation time, prolong arteriovenous time, staining of retina vessels, and nonperfusion areas in BE (Fig. 2). The diagnosis of bilateral OIS was done. A complete work-up was required in order to establish the cause of OIS. Angiogram revealed a severe stenosis in both subclavian arteries, stenosis of the right common carotid artery, occlusion of the left common carotid artery, severe stenosis of the right and left vertebral arteries (Fig. 3), and she had also abdominal aorta involved. Laboratory studies: C-reactive protein was 54, erythrocyte sedimentation rate of 84, and infection causes were ruled out. The diagnosis of TA was established. She started on corticosteroids and mycofenolate with good response, with a partial reperfusion of carotids and good perfusion of both eyes.

Discussion

The pathologic course of TA starts in the adventitia, which progresses to the intima with marked proliferation and fibrosis, eventually causing vascular narrowing, occlusion, with or without thrombosis, and as a result, compromise of the blood flow occurs through the involved vessels. Aneurismal formation can be seen later.[5]

c-TA starts with a nonspecific and acute inflammatory phase (anorexia, fever, night sweats, weight loss, arthralgia, and skin rash).[3,4] After that, one-third of children experience effects of tissue ischemia with significant vascular sequelae. The average time between the symptoms until the final diagnosis is 19 months, almost four times longer than adults.[3–5]

The spectrum of clinical features varies in pediatric and adult patients, being hypertension the most common symptom in both groups (73%), followed by headache (53%), constitutional symptoms (53%), and fever (45%). Bruit and claudication pain are uncommon in c-TA.

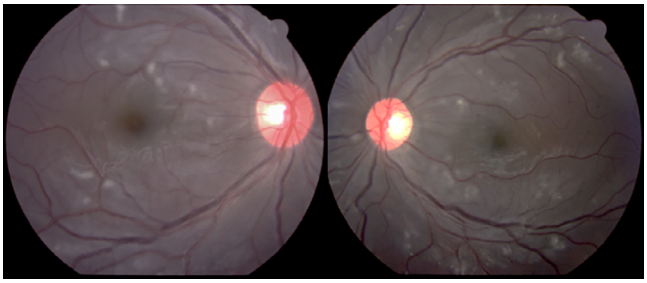

Fig. 1—Bilateral color fundus pictures showing venous dilatation, arteriolar narrowing, cotton wool spots and widespread microaneurysm formation.

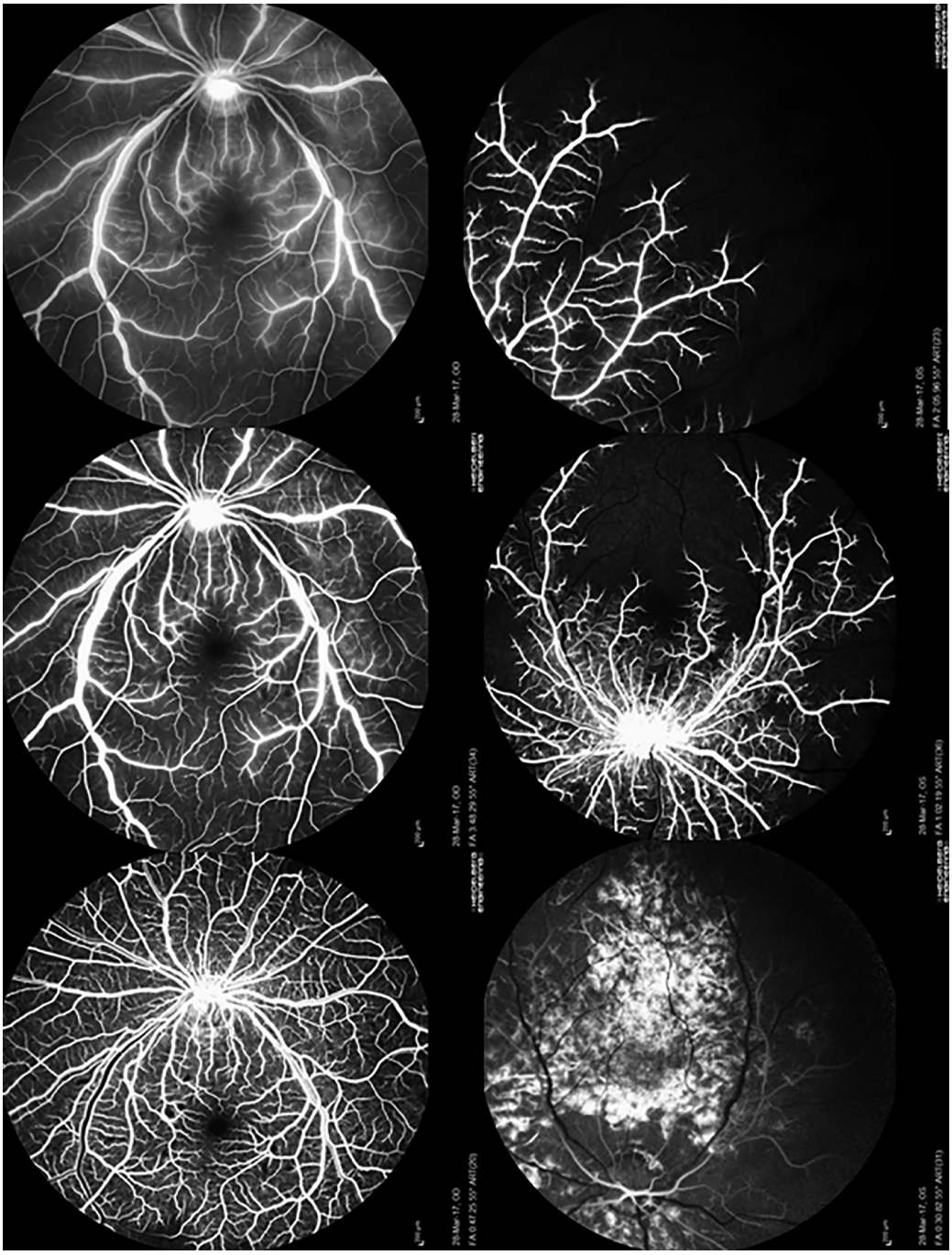

Fig. 2—FFA revealed patchy choroidal filling, delayed arm to retina circulation time, prolong arteriovenous time, staining of retina vessels and non-perfusion areas in both eyes.

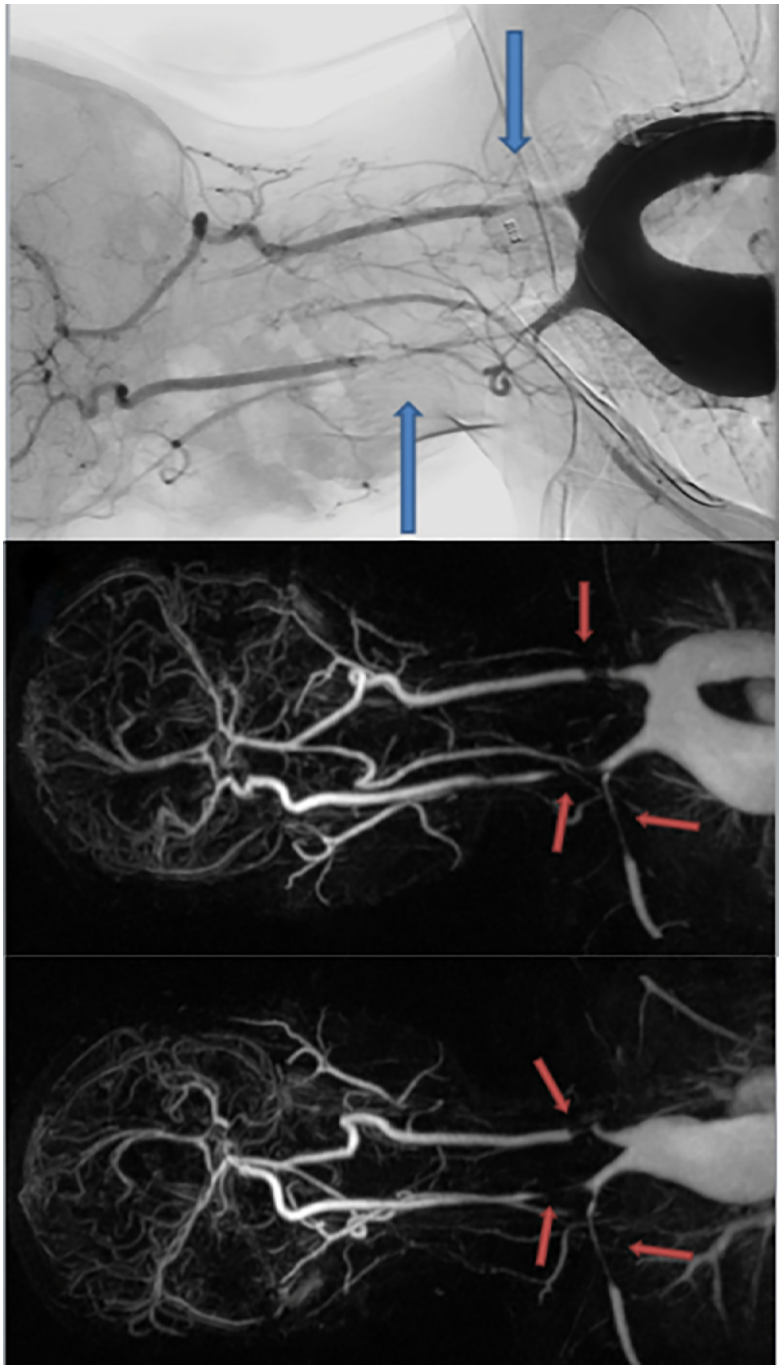

Fig. 3—Angiogram showed a severe stenosis in both subclavian arteries, stenosis of the right common carotid artery, occlusion of the left common carotid artery and severe stenosis of the right and left vertebral arteries.

Eye involvement is rare in c-TA. Most children have inferior aortic involvement with renal complications, but no ocular ischemia.[1–3]

Chun et al. reported amaurosis fugax as the most common ocular symptom in their study of 78 patients with TA (25.6%). Other includes transient, progressive, and acute loss of vision, sometimes associated with retro-orbital pain.[6]

The manifestations in Takayasu disease are classified in hypertensive or hypoperfusive manifestations. The hypertensive manifestations occur as result of involvement of the renal artery or suprarenal aorta, leading to severe and uncontrolled hypertension. On the other hand, hypoperfusive manifestations are secondary to an occlusive arteritis of the aortic arch and its branches, leading to the ischemic ocular manifestations. It usually develops after occlusion or severe stenosis of the carotid arteries.[7]

Most retinal findings in Takayasu disease result from hypoperfusion and resemble those seen in the ocular ischemic syndrome caused by atheromatous carotid artery occlusion.[1] The best described ischemic ocular manifestation in TA is Takayasu retinopathy. Initially, affected patients develop generalized tortuosity and vasodilatation of retinal veins with arteriovenous anastomosis, capillary dropout, and microaneurysms. Eventually, complications of ocular ischemia, including vitreous hemorrhage, neovascular glaucoma, retinal detachment, and optic atrophy may lead to blindness.[1]

Because early eye changes may be subtle and asymptomatic, all patients with TA should be routinely evaluated to rule out eye manifestations to institute early treatment to prevent blindness.[2] Therefore, it is important to know the clinical and ocular manifestations, natural course, and appropriate treatment methods before the development of severe complications.

Treatment

The primary objective of treatment in c-TA is to prevent irreversible vessel damage and to preserve vital organs.

Corticosteroids remain the mainstay of treatment in c-TA. Second-line agents like methotrexate, azathioprine, mycophenolate mofetil, and cyclophosphamide have been tried along with steroids. Biological therapy, mainly antitumour necrosis factor agents, has also been tried in management, with good results.

Currently there is no evidence-based data proving the superiority of a single agent over another in the treatment of c-TA.

Percutaneous transluminal angioplasty with stenting and surgical interventions has been described alongside medical therapy to alleviate end organ ischemia after vascular damage.

Prognosis

Death occurs as a result of complications, such as congestive heart failure, strokes, or myocardial infarction. The mortality rate in c-TA has been reported around 35% by earlier series. Extent of vessel involvement and severity of hypertension are the crucial factors deciding the outcome in most of these series. However, early diagnosis and integrative management principles have resulted in a favourable change in long-term survival rates over the past decade.[1]

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Cerebral venous sinus thrombosis and iron deficiency anemia presenting as bilateral disc edema in a child

Case presentation

A 30-month-old female presented upon referral from her pediatrician with new-onset esotropia. History was significant for a recent viral illness with associated vomiting, headache, and weight loss without irritability or change in behaviour. Sudden-onset esodeviation of the right eye (OD) was noted 2 weeks before presentation. Past medical history was significant for iron deficiency anemia diagnosed 2 months before for which she had been started on oral iron replacement therapy (ferrous sulfate 112.5 mg twice daily). External examination demonstrated a right head turn of 15 to 20 degrees. The child was able to fix and follow in both eyes (OU) with motility demonstrating an abduction deficit OD and full motility in the left eye (OS). Pupillary examination was normal without afferent defect. Dilated examination revealed 4+ disc edema in both eyes (OU).

With significant disc edema and concern for a right sixth nerve palsy, the patient was sent for emergent imaging to rule