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Case Report Open Access

Vasculitis of Superficial Temporal Artery in a Pediatric Patient

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Abstract

While superficial temporal artery (STA) vasculitis is typically a disease of the elderly, spontaneous STA aneurysm in children is anecdotic and usually caused by a subjacent vasculitis. Since 1948 around 40 cases have been listed in literature and just 6 of them under the age of 18. Three main forms have been classified: juvenile temporal arteritis, typical giant cells arteritis and temporal artery involvement secondary to systemic vasculitis. We report a rare case of STA aneurysm in an 8 year old patient already known for the surgical treatment of an aortic coarctation. The patient was asymptomatic, clinical exam was unremarkable and no traumatism, fever, arthralgias or recent infective episodes were reported. No residual coarctation at echocardiography. STA echo-colour Doppler suggested juvenile temporal arteritis, showing a dilation of 5.4 x 8.7 mm. Doppler scanning of the other districts was normal. At cerebral MRI a spindle-shaped dilation over the left STA was confirmed without other intracranial anomalies. Thoraco-abdominal angio-TC was normal to the entire aorta and at the visceral arteries level. Blood exams were unremarkable. A biopsy of the left STA was performed under general anesthesia: a true aneurysm of the artery (1.5×8 mm) was resected. At anatomopathologic examination neither epithelioid nor great cells were found but a lymphocytic infiltration was detected around the vasa vasorum. Isolated STA vasculitis in young individuals results in different findings, pathogenic triggers and clinical manifestations from affecting the elderly. Diagnosis is very important to direct the appropriate therapeutic strategy and must include histopathologic evaluation.

Keywords: Superficial temporal artery aneurysm; Temporal vasculitis; Vasculitis in children; Pediatric vasculitis; Pediatric arteritis

Abbreviations

ALHE: Angiolymphoid Hyperplasia with Eosinophili; ANCA: Anti-Neutrophil Cytoplasmic Antibodies; anti-MPO: Anti Myeloperoxidase antibodies; anti-PR3: Anti-Proteinase 3 Antibodies; CSS: Churg-Strauss Syndrome; CRP: C-Reactive Proteine; CT: Computed Tomography; GCA: Great Cells Arteritis; JTA: Juvenile Temporal Arteritis; MRI: Magnetic Resonance Imaging; PAN: Polyarteritis Nodosa; STA: Superficial Temporal Artery; TAO: Thromboangiitis Obliterans

Introduction

Superficial temporal artery (STA) vasculitis is typically a disease of the elderly, being extremely rare in young patients and pediatric population. Overall, around 40 cases have been listed in literature since 1948 [1] of which only seven concern patients younger than eighteen years (Table 1). Three main forms have been classified: juvenile temporal arteritis (JTA), typical giant cells arteritis (GCA) and temporal artery involvement secondary to systemic vasculitis [2]. The latter has been observed more frequently in polyarteritis nodosa (PAN) [3], Churg-Strauss syndrome (CSS) [4] and thromboangiitis obliterans (TAO) [5].

JTA is the more frequent form and confers the best prognosis; all reported cases have been resolved with excision of the affected temporal artery without the need for systemic therapy. On the contrary, GCA and temporal arteritis as a manifestation of a systemic vasculitis require immune modulating therapy to avoid potential end-organ damage [6]. All three disorders described can affect the medium-sized vessels, such as the STA, but their histopathology greatly differs. Clinical manifestation is different too, varying from an almost asymptomatic localized disease to a systemic syndrome with a multi-organ involvement. Histological differences should be firstly taken into account when formulating a diagnosis. Differential diagnosis includes Kimura's disease (an idiopathic angiolymphoid inflammatory disorder which involves subcutaneous tissues and lymph nodes of the head and neck region, characterized by eosinophilic infiltrates and lymphoid follicles - more frequent in oriental population), angiolymphoid hyperplasia with eosinophilia (ALHE), Takayasu's arteritis [7].

Case Report

We present the case of an eight year old patient referred to our Unit for the incidental finding of a throbbing swelling on the left temple, characterized as a superficial temporal artery aneurysm. The patient was completely asymptomatic. No traumatism was mentioned neither fever nor recent infective episodes; no history of allergy nor arthralgias were reported. However, the child was already known since the age of three for having undergone, in March 2007, the surgical treatment of an aortic coarctation using the Crafoord technique. Clinical examination was unremarkable, except for the throbbing swelling on the left temple, with no signs of inflammation in the surrounding skin.

No adenomegaly, hepatomegaly or splenomegaly were found, nor signs of arthritis. An echocardiography was performed to exclude a residual coarctation or an aortic dilation (diameter 20.6 mm) and a good flow through the aortic isthmus with a maximum gradient of 15 mmHg was found. At the echo-color doppler (Figure 1A) the STA appeared dysplastic over a length of 10 mm and dilated (short axis diameter of 5 mm). The artery's wall was thickened to 1.5 mm with no edematous infiltration surrounding the aneurysm, with a circulating lumen of 2.3 mm without hemodynamic perturbation. Echographic pattern was suggestive of JTA. Doppler scanning of the other districts was otherwise normal. A magnetic resonance imaging (MRI) was then preformed to better characterize the STA and to screen the intracranial arterial tree, which excluded the presence of intracranial aneurysms (Figure 1B-C). At the thoracoabdominal angio-CT a mild enlargement on the sub-isthmic thoracic aorta was described with no pathologic thickening of the wall or aneurysms over the entire aorta and at the visceral arteries level (Figure 1D).

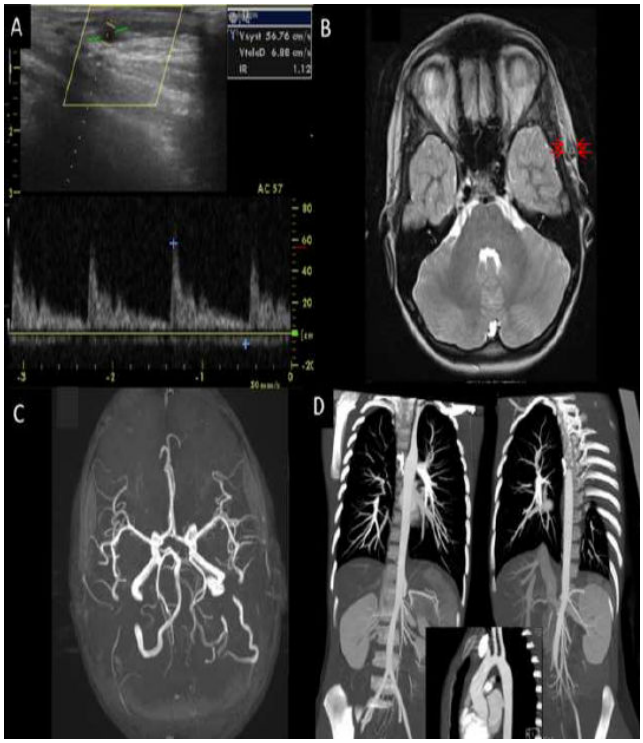

Figure 1: A) Echo-color Doppler showing STA aneurysm and related flow acceleration; B-C) Encephalic MRI showing a dilation of spindle-shaped appearance over the left STA at the temporal region's level (Arrow in Figure B). No images of malformation at the intra-cranial arteries level (Figure C), nor ischemic or haemorrhagic signs, cerebral edema or mass syndrome; D) Thoracoabdominal angio-CT scan showing a normal aortic pattern with a mild enlargement on the sub-isthmic thoracic aorta in the absence of a pathologic thickening of the wall or aneurysms over the entire vessel and at the visceral arteries level.

Blood exams resulted within physiologic limits with normal white blood cells count and no increase in acute phase protein levels (CRP, fibrinogen, and orosomucoid). Immunological markers such as ANCA, anti-MPO and anti-PR3 antibodies were negative and so were the tests for hepatitis B and C viruses. After completion of clinical and instrumental evaluation, a biopsy of the left STA was performed in the operating room under general anaesthesia: a true aneurysm of the artery, approximately 1.5 cm in length and 8 mm in width, was eventually found and resected with a ligature of the remaining branches. Anatomopathologic examination (Figure 2A-B-C-D) showed an extremely re-handled arterial wall with an endoluminal obstruction by a fibrin clot. Neither epithelioid nor giant cells were found but a lymphocytic infiltration around the vasa vasorum (Figure 2B) was observed. This aspect was suggestive of a subjacent vasculitis.

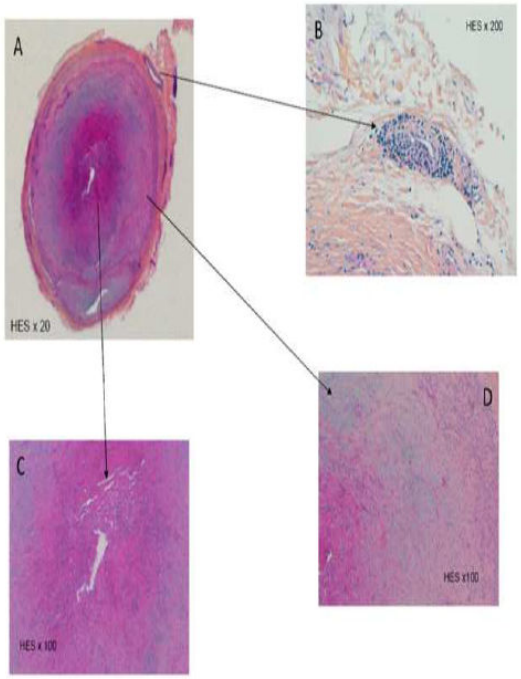

Figure 2: A) Transversal section of the aneurysmatic vessel; B) STA's vasa-vasorum sourrounded by a circumferential lymphocytic cuff; C) Endoluminal obstruction by a fibrinous clot with focal aspects of re-permeability; D) Artery's wall re-handling with myo-fibroblastic proliferation over a myxoide pseudo-chondroid field with destruction of the elastic limitant and rarefaction of the smooth muscular fibres of tunica media.

Discussion

Spontaneous STA aneurysm in children is anecdotal. While post-traumatic psuedoaneurysms are more frequent and typically present in younger patients as the consequence of blunt trauma related to sport injuries, falls, accidents, and altercations, true STA aneurysms are rare pathologies associated with aging and mainly caused by atherosclerosis processes, congenital vulnerabilities of the arterial wall and arteritis. [8]. As the first cause cited is extremely unlikely in paediatric population we consider the former as the main cause especially in association with a constitutive congenital arterial vulnerability. STA vasculitis in the young are listed as above.

Year / Author	Age / Gender	Symptoms	Diagnosis
Meyers (1948) [1]	22 F	Headache	JTA
Bethlenfalvay (1964) [12]	35 M	Headache, swollen TA	JTA
Lie (1975) [10]	21 M, 22 F – 7 M, 8 M	Swollen TA	JTA
de Faire (1977) [13]	23 F	Headache, visual symptoms	GCA
Conn (1982) [14]	49 F	Headache, vision loss,	CSS
Villalta (1985) [15]	32 M	Swollen TA, arthralgia	GCA
Ferguson (1985) [16]	47 M	Swollen TA	TAO
Bollinger (1986) [17]	23 M	Swollen TA	JTA
Lie (1988) [5]	38 M, 32 M, 36 F	Swollen TA	TAO
Amato (1989) [4]	25 M	Multi	CSS
Genereau (1992) [18]	19-32 (5M;1F)	Only JTA	Only 3 are JTA
Vidal (1992) [19]	41 M	Asthma, jaw claudication, headache, swollen TA	CSS
Thomlison (1994) [20]	8 M	Headache, swollen TA	JTA
Fielding (1994) [21]	30 M	Swollen TA	JTA
Lie (1994) [22]	48 M	Swollen TA, cough dyspnea	CSS
Grishman (1995) [23]	34 M	Swollen TA, ischemia of extremities	PAN
Lie (1995) [24]	45 F	Lung AdenoCa, headache swollen TA	GCA
Lie (1995) [25]	21 M	Bilateral TA swelling	JTA
Fujimoto (1996) [26]	39 M	Bilateral TA swelling	JTA
Bert (1999) [3]	9 F	Multi	PAN localized
Endo (2000) [27]	27 M	Bilateral TA swelling, Raynaud phenomenon	CSS
Andonopoulus (2004) [28]	31 M	Swollen TA	JTA
Granel (2004) [7]	34 M	Headache, bilateral TA swelling	JTA
Wu (2004) [29]	30 F	Dizziness, headache, bilateral sensorineural hearing impairment	GCA vs Primary Angiitis of Central Nervous System
Fukunaga (2005) [11]	23 M	Swollen TA	JTA
Pipinos (2006) [30]	17	Asthma, reumatoid arthritis, corneal transplant; TA aneurysm,	GCA
Nesher (2008) [6]	18 M	Swollen post-traumatic TA	JTA
Dinesh (2010) [31]	42 M, 45 M	HIV, Blurred vision and swollen TA; swollen TA	???
Kolmann (2010) [32]	36 F	Headache, swollen TA	JTA
Paparo (2011) [33]	35 F	Swollen TA	Kimura Disease
Kim (2011) [34]	24 F	Swollen TA	JTA vs Kimura
Durant (2011) [35]	44 F	Headache, swollen TA	JTA
McGoech (2012) [2]	31 M, 40M	Symptoms	GCA vs Primary Angiitis of Central Nervous System

Table 1: Temporal artery vasculitis in young as reported in literature

Differences between each other are mainly based on the clinical presentation along with the histopathologic findings. However, diagnosis is not always univocal due to the frequent overlapping of the manifestations. GCA in the elderly is a well-known disease, characterized by the criteria established by the American College of Rheumatology set up in 1990. These include age greater than 50, new onset of localized headache, STA tenderness, elevated erythrocyte sedimentation rate and a necrotizing arteritis in the presence of mononuclear cells or a granulomatous process with multinucleated giant cells [9]. Much less is known about the form affecting the young. Its histological features usually fall into three patterns: granuloma replete with giant cells and with a fragmented internal elastic lamina, nonspecific white cell infiltrate throughout the arterial wall and intimal fibrosis without disruption of the internal elastic membrane. Giant cells are present in only half to two thirds of the cases [3]. Nesher et al. [6] characterize GCA in the young as a non-eosinophilic "Elderly-Type" temporal arteritis, gathering the few cases where histology showed intimal hyperplasia together with the presence of giant cells and mononuclear cells infiltrates in the absence of eosinophils, irrespectively from the symptoms. On the opposite, in JTA they find an arteritis with inflammatory infiltrates containing eosinophils but no trace of giant cells are found. The term JTA was coined by Lie and co-workers in 1975 [10]. It is considered a benign course condition that occurs in children and adults under 40 years, characterized by (a) pain in temporal region, with swelling or artery induration; (b) no associated inflammatory symptoms; (c) objective ophthalmic symptoms; (e) possible blood eosinophilia with eosinophilic infiltrate in biopsy; (f) no need for steroid treatment [7]. It can present unilaterally or bilaterally. Patients with JTA are often asymptomatic, without a history of preceding or concurrent systemic illness. When the initial presentation involves temporal arteritis, the clinical course of the disease helps to distinguish JTA from more aggressive entities. GCA juvenile form is typically more symptomatic and has systemic involvement (even if localized cases are reported [6]) and eventually need high-dose corticosteroid management.

Temporal artery involvement secondary to systemic vasculitis is normally a more complex condition with multi-organ involvement. It has been observed in PAN, CSS, TAO, in primary angiitis of the central nervous system and in eosinophilic granulomatous polyangiitis [2]. It has also been hypothesized that JTA could be a localized form of PAN [10]. The range of manifestation is large but in the majority of cases it is possible to differentiate each form by means of its clinical manifestations, immunological testing, radiological and pathological findings, although some cases of overlapping syndromes are listed. [2,6]. Concerning differential diagnosis with Kimura's disease and ALHE, Fukunaga et al. [11] suggested the possibility that the lesion of the temporal artery in JTA could be secondary to one of these conditions, being effectively an expression of the same disease. However, no temporal artery involvement of any of these two forms has yet been reported in childhood. Our case was finally diagnosed as a JTA, having five of the six features described by Lie et al. [10] and having excluded a local involvement of STA from other conditions. In contrast with the typical histopathologic findings in JTA no eosinophilic infiltrate was found but signs of a subjacent vasculitis and absence of giant cells were strongly suggestive of it.

In conclusion, isolated STA vasculitis is a very rare pathology in the young, being almost anecdotal in children. It has different findings from the form affecting the elderly and can be determined by different types of pathogenic triggers with different clinical manifestations. Diagnosis is very important to direct the best therapeutic strategy and it should take into account laboratory screening, imaging and histopathologic evaluation. No association between aortic coarctation and STA aneurysm or STA vasculitis in children have been described in literature and their coexistence in our case is probably merely accidental.

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