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Juvenile temporal arteritis: a clinicopathological multicentric experience

Louis Journeau^a, Marc-Antoine Pistorius^a, Ulrique Michon-Pasturel^b, Marc Lambert^c, Francois-Xavier Lapébie^d, Alessandra Bura-Riviere^d, Philippe de Faucal^e, Patrick Jegou^f, Quentin Didier^a, Cécile Durant^a, Geoffrey Urbanski^g, Baptiste Hervier^h, Claire Toquetⁱ, Christian Agard^a, Olivier Espitia^a ; Groupe d'Étude Français des Artérites des gros Vaisseaux.

^a Department of internal medicine, Hôtel-Dieu, CHU Nantes, Nantes, France.

^b Department of internal medicine, Centre Hospitalier Universitaire de Lille, Lille, France.

^c Department of vascular medicine, Hôpital Saint-Joseph, Paris, France.

^d Department of vascular medicine, Hôpital Rangueil, CHU de Toulouse, Toulouse, France.

^e Department of internal medicine, Hôpital privé du Confluent, Nantes, France.

^f Department of internal medicine, Hôpital Sud, CHU de Rennes, Rennes, France.

^g Department of internal medicine and vascular diseases, CHU d'Angers, Angers, France.

^h Department of internal medicine, Hôpital Pitié-Salpêtrière (AP-HP), Paris, France.

ⁱ Département de pathologie, Hôtel-Dieu, CHU Nantes, Nantes, France.

Corresponding Author:

Dr Olivier Espitia MD, PhD.

Department of Internal Medicine, CHU de Nantes

1 place Alexis Ricordeau 44093 Nantes, France.

Tel : +33 240 083 355 Fax : +33 240 083 379
olivier.espitia@chu-nantes.fr

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Declarations of interest: None

Bullet points

- Juvenile temporal arteritis, in opposition to the classical giant cell arteritis, is an extremely rare, localized, and benign disease affecting the temporal artery in patients aged less than 50 years, and presenting as a lump in the forehead, sometimes painful, without systemic symptoms or biological inflammation, which is most frequently cured by biopsy or excision.
- Kimura disease and angiolymphoid hyperplasia with eosinophilia must be considered as alternate diagnosis.

Abstract

Introduction: Juvenile temporal arteritis (JTA) is a recently-described and little-known inflammatory disease and its etiology is undetermined. Less than forty cases have been published. This paper is aimed at reporting the largest JTA series and to compare it to literature data to better evaluate its characteristics at diagnosis, its evolution and treatment options.

Material and methods: We conducted a retrospective and descriptive multicentric study in France by identifying adult patients under the age of 50 which had a pathological temporal artery biopsy owing to the presence of a temporal arteritis. Patients with temporal arteritis as a manifestation of systemic vasculitis were excluded.

Results: We included 12 patients and the literature review identified 32 cases described in 27 articles, thus a total of 44 patients – 34 men and 10 women – with a median age of 30 and a maximum of 44. All patients presented either a lump in the temporal region or prominent temporal arteries, and 47.7% of patients suffered from headaches. Only 11.4% of patients presented general symptoms and 6.8% a biological inflammatory syndrome; 34.1% had peripheral blood eosinophilia; 83.7% presented a single episode and complete excision without further treatment was documented for 72.7%. Pathology analysis revealed infiltrate of inflammatory cells in the arterial wall in 97.6% of patients but also sparse giant cells for 25% and granuloma for 22.9%, perivascular extension of the inflammation for 82.6%, and presence of lymphoid follicles or germinal centres for 60%. Clinical relapses were present in 16.3% of cases.

Conclusion: JTA is a rare, localized and benign disease. The majority of cases have only one episode which is cured by local surgery.

1.Introduction

Juvenile temporal arteritis (JTA) is a rare and little-known inflammatory disease of the temporal arteries. Contrary to giant cell arteritis (GCA)[1,2], JTA is a recent entity, initially described by Lie *et al.* in 1975 [3], and its etiology is unknown. Less than forty cases have been published so far, of which only six had bilateral involvement [3–30]. JTA is a localized juvenile temporal arteritis, with most of the time a localized eosinophilic infiltration confined to the temporal arteries which seems unique to this age group. Temporal locations of systemic vasculitis, such as polyarteritis nodosa, or non-eosinophilic vasculitis with clinical and histological features resembling classical GCA are the main differential diagnoses of JTA.

Some diagnostic criteria of JTA have been suggested by Tomlinson *et al.* in 1994 [11] : 1) Age under 40; 2) non-tender or painful palpable lump in the forehead area of the temporal artery; 3) no associated signs such as fever, myalgia, visual symptoms, or anemia; 4) normal erythrocyte sedimentation rate; 5) eosinophilic pan-arteritis with intraluminal thrombus which may be associated with parietal micro-aneurysmal lesions; 6) intimal proliferation with discreet damage to media and extensive cellular infiltrate made of lymphocytes, eosinophils and plasma cells in the perivascular tissue; 7) absence of granulomatous lesions or giant cells.

Moreover, the exact nosology of JTA has yet to be defined: similarities between JTA and Kimura disease have previously been reported [18,23,31]. Kimura disease is characterized by a lymphocytic and eosinophilic infiltrate, which is more pronounced in the perivascular tissue, and the presence of lymphoid follicles and eosinophilic abscesses. The issue is similar regarding JTA and angiolymphoid hyperplasia with eosinophilia (ALHE), which can be summarized as superficial and erythematous nodules or plaques, projecting in the cephalic and cervical region; with intimal hyperplasia, lymphocytic and eosinophilic infiltrate, which can extend to the perivascular tissue.

Due to the rarity of JTA, patients are usually subject to misdiagnosis and multiplication of invasive explorations. The mid- and long-term evolution is unknown. Therapeutic management is not standardized. Contrary to Kimura disease and AHLE, relapses seem rare.

The aim of this study was to report the largest JTA series in order to better evaluate its clinical, biological, imaging and pathological characteristics at diagnosis, to present the different therapeutic management options, to inform about its long-term evolution, and to present a literature review of JTA cases.

2.Material and methods

We conducted a retrospective and descriptive multicentric study in France. Inclusion criteria were adults aged under 50 and the presence of a temporal arteritis, be it clinical (induration and/or pain in the temporal region) or via imaging technique (hypochoic wall thickening and at best the halo sign in Doppler ultrasound;

tortuosity, dilation or aneurysm, wall thickening or occlusion in Doppler ultrasound, angio-CT scan, MRI or even arteriography) and pathological temporal artery in histology study. Exclusion criteria were temporal arteritis as a manifestation of systemic vasculitis or diseases (Takayasu's disease, ANCA-associated vasculitis, polyarteritis nodosa, cryoglobulinemia, Buerger disease, lymphomas).

Cases were first identified as adult patients under the age of 50 which had a pathological temporal artery biopsy in the university hospitals of Angers, Brest, Nantes, Rennes, Poitiers, Lille, Limoges, Toulouse, Dijon, George Pompidou European Hospital in Paris (AP-HP), Ambroise Paré hospital in Boulogne (AP-HP), Louis-Mourier hospital in Colombes (AP-HP), and the Medipath® database (first private network of pathologists in France). We also inquired for potential cases via the French Study Group for Large Vessel Vasculitis (GEFA) and the French Society of Internal Medicine (SNFMI).

Medical records of these cases were reviewed with a standardized grid for clinical, biological, temporal arteritis histology, therapeutic, and outcome data.

The study was approved by the "Groupe Nantais d'Ethique dans le Domaine de la Santé" (GNEDS), the ethics committee of the Nantes university hospital, and complied with the requirements of the "Commission Nationale de l'Informatique et des Libertés", in accordance with current French legislation.

Review of the English and French literature was conducted from the MEDLINE base until December 2017. The terms used in the search with various combinations were juvenile temporal arteritis, temporal artery, temporal arteritis, cranial arteritis, juvenile, young, eosinophilic vasculitis, and temporal vasculitis. We selected articles that described patients younger than 50 and who presented temporal arteritis, excluding those with systemic vasculitis or systemic disease involving the temporal arteries.

We present our data by individualizing the cases of our series from those of the literature, and by uniting the two series as global results. In this article, individual data from literature cases were reported only if the data was clearly described in the original article.

3.Results

We included 12 patients (10 men and 2 women). Three men hailed from Maghreb, all others were Caucasians. Thirty-two cases of JTA described in 27 articles were identified by the literature review [3–29]. The case published by Durant et al. described a patient which was cared for at the Nantes University hospital [30] and we therefore went back to the patient’s original medical file to include him in our cohort. Table 1 presents the main clinical and biological characteristics of these 44 cases as well as their treatments and evolution. Eight out of our 12 patients were smokers (75%), vs. 7 out of 32 in the literature (22%). We noted the use of marijuana in two of our patients.

3.1 Clinical data:

With the exception of one of our patients, JTA was described as a dermohypodermic lump or nodule in the temporal region, or as an induration of the temporal artery (Figure 1). For our sole patient without any description of nodule or induration, its file nonetheless mentioned that the temporal arteries were unusually beating. Two cases reported a sensation of temporal “horn growth” and one of our patients had a spontaneously resolving occipital scalp nodule several months before temporal arteritis. Absence of previous trauma was almost constant: only one patient of the literature specified an interval of three months between a trauma and the first appearance of a temporal nodule [21]. JTA was frequently, but not always, associated with mild to moderate headache around the nodules. In the absence of headache, the unsightly aspect of the nodule usually prompted the patients to consult.

Only one of our patients presented general symptoms at the time of diagnosis: asthenia, arthromyalgia, and Raynaud’s phenomenon of the 3rd and 4th digit of the right hand. However, this affected 6 patients of the literature : asthenia and sickness [4] ; weight loss and nocturnal sweating [5] ; arthromyalgia [6] ; fever, arthromyalgia, and digital necrotic ulcer [7].

The median duration of symptom progression prior to JTA diagnosis was 6 months [0.5-60 months] in the literature, and 4 months [1-21 months] in our series.

3.2 Biological data:

Three cases had moderate biological inflammation : one patient of our cohort with a CRP of 10.3 mg/l and two with elevated ESR (24 and 75 mm) [6,23]. There was no case of anemia, and leukocytosis was present in only 4 patients of the literature (mean 13 ± 1 giga/L) [6,8,20,22] among which only one with hypereosinophilia (total leukocyte count 13.9 giga/L and eosinophils 4.7 giga/L) [8]. Three cases of elevated total IgE plasma levels have been reported [8,25,29], but this data is rarely mentioned. Only one patient of the literature had

elevated titers of antinuclear antibodies, without specificity, present since the age of 3, with juvenile arthritis complicated by uveitis and which was quiescent when JTA was diagnosed [20]. There were no patients with anti-DNA antibodies, ANCA or elevated creatine kinase titers.

3.3 Pathological data:

Histological characteristics of JTA (Figure 2) are summarized in Table 2.

In the literature, some pathological reports present minimal lesions with intimal hyperplasia, perivascular lymphocytic infiltration, and the presence of lymphoid follicles [24,28]. This scenario concerns one of our patients who underwent two surgical procedures: the first one consisted in a temporal artery biopsy which showed lymphocytic infiltration of the peri-arterial adipose tissue, associated with a capillary-rich sector. The second intervention consisted in the exeresis of the entire lesion and analysis evidenced a benign capillary angioma. Another patient who presented with bilateral lesions had a first temporal artery biopsy which was negative. A complete excision of the contralateral temporal artery then demonstrated typical JTA and the patient fully recovered.

The presence of intraluminal thrombosis was 63.6% in literature versus 77.8% in our cohort. The nature of the parietal cellular infiltrate was quite variable. The presence of an eosinophilic infiltrate was not systematic.

3.4 Treatment data and evolution:

The majority of patients underwent complete excision of the culprit artery (Table1). There was no difference in the relapse rate between those who had undergone simple biopsy and those with complete excision.

Our sole patient with systemic signs (asthenia, arthromyalgias, unilateral Raynaud's phenomenon, 30 years-pack active smoking history) presented a minor inflammatory biological syndrome (CRP 10.3 mg/L) and he was the only one to receive a short course of steroids. His temporal artery biopsy showed no atypia and he totally cured with the artery excision and the steroid course, with no relapse after 5 years of follow-up.

Two of our patients, both of whom suffered from bilateral lesions, received colchicine as a first-line treatment. In one case, the side that was biopsied entirely healed but the contralateral lump was still present after 15 months of follow-up. For the second patient, the treatment lasted for 3 months, with bilateral healing.

The first of these two patients, who relapsed initially, presented a right temporal artery induration which was excised, and a few months later, a left induration. There were no other symptoms and he received no specific oral treatment. After a 2 months' follow-up, he still presented left temporal artery induration which was stable, with parietal thickening in ultrasound examination but no halo sign. The second patient was biopsied on the right side and twelve months later a Doppler ultrasonography examination revealed bilateral lumps: the right one was the persistence of what was biopsied twelve months before, and the left one was new. No biopsy or systemic treatment was undertaken. Two years later, the patient is symptom-free.

Concerning the 5 patients of the literature who relapsed : in two cases, the relapse was unique and affected the opposite side, with no systemic treatment [14,26]. In the three other cases, there were several relapses : the

first patient went through short courses of steroids for each relapse [5], the second underwent local excision [19], and the third received oral steroids for 5 months for the first relapse and methotrexate for the second [29]. In three cases, relapses occurred on the same side as the initial presentation: in one case, the recurrence was unilateral three months after the initial episode and appeared a few millimeters from the site of the initial biopsy [14], in another case a relapse was also on the initially operated area [19], and in a third case, the patient experienced several recurrences of left temporal headache [5].

No patient developed systemic vasculitis. In one case of the literature [22], one month after excision the patient still complained of headache, malaise and induration at the site of excision, without any kind of systemic treatment, which was more suggestive of lack of initial remission than relapse. One patient presented 24 months later cervical and axillary lymphadenopathies and tuberculous cutaneous infiltrates, even though symptoms and signs of JTA had resolved. Diagnosis of primary cutaneous T-cell-non-Hodgkin-lymphoma (gamma-delta phenotype) was confirmed by histology [25].

Another one of our patients had already been diagnosed with spondylarthropathy (psoriatic arthritis), which was well controlled with methotrexate and abatacept when he developed JTA one year later. Eight years later, the rheumatism was still quiescent when the patient developed asymptomatic chronic hypereosinophilia, suspected of being of a toxic origin (perfume) and which improved spontaneously.

4. Discussion

This study reports the largest series of JTA, an extremely rare variant of localized vasculitis which manifests itself almost exclusively by temporal signs (lumps and headache) in young people. Hypereosinophilia is present in one third of cases. Its evolution is almost always favorable, especially after temporal artery excision, and relapse is uncommon. JTA distinguishes itself from the temporal arteritis of the elderly (giant cell arteritis) by a male predominance, the absence or the discretion of general symptoms or ocular complications, and the scarcity of a biological inflammatory syndrome. As in ACG, Doppler ultrasound of temporal arteries show halo sign in JTA [32]. On the other hand, unlike GCA, there is no sign of vasculitis in other artery territories on TEP-CT or CT scan [33–36]. Two forms seem to stand out: a silent form consisting of an isolated temporal nodule and a more symptomatic form.

JTA may clinically mimics several non-vascular and vascular lesions, such as lipomas, sebaceous cysts, dermoid cysts, aneurysms or arteriovenous malformations, subcutaneous AHLE and classic temporal arteritis. The differential diagnosis include trauma-related thrombosis, thrombosis of a vascular malformation of the temporal artery, or temporal localization of systemic vasculitis (Buerger's disease, ANCA-associated vasculitis, eosinophilic granulomatosis with polyangiitis, periarteritis nodosa) [37–39]. The temporal arteries can also be affected during infectious vasculitis (toxocariasis, syphilis, borreliosis, herpes zoster, AIDS) and during systemic diseases (amyloidosis, systemic sclerosis, sarcoidosis) [38,40].

4.1 Pathophysiology

The pathophysiology of JTA is still unknown. It has been hypothesized that trauma may result in pseudoaneurysm formation and that a subset of these cases may develop reactive capillary proliferation with inflammatory reaction, but in the literature only one case is reported after trauma making this hypothesis unlikely [41]. It is possible that the wall of the temporal arteries provide an antigenic stimulus which may be at the origin of an immune reaction, since artery excision alone heals JTA while the eosinophil count decreases. In the JTA as in the GCA there are no biomarkers for the diagnosis [42,43].

4.2 Clinical presentations

A clear distinction between JTA and GCA is clinically important because high-dose corticosteroid therapy is almost standard management for the latter but is unnecessary in JTA and may be potentially harmful.

Most of the reported cases of the literature presented Tomlinson *et al.* JTA's criteria [11]. However, a few cases showed no-previously described histological pattern and others had minor systemic clinical symptoms which lead us to question the nosology of JTA. The most frequent differences were: presence of systemic clinical signs [4–7,10,22]; presence of sparse giant cells [5–10,14,20] and/or granulomas [3,6,7,14,15] on the

pathological analysis; presence of an inflammatory biological syndrome [6,20,22,23]. Two patients of our cases had giant cells and granuloma on their temporal artery biopsy but apart from that, the clinical description was perfectly compatible with JTA. One of the two relapsed on the contralateral side but received no specific treatment since he was paucisymptomatic. The other one cured and experienced no relapse after three months of colchicine.

Clinically, it is remarkable that temporal artery lesions occurred in children and young adults, and appeared in such an innocuous manner. The clinical features that characterize JTA are non-tender or painful palpable lump in the forehead area of the temporal artery and the absence of associated features such as myalgia, visual disturbance, fever, anemia or increased erythrocyte sedimentation rate, which occur in GCA. The visual symptoms noted in classic temporal arteritis reflect the involvement of the ophthalmic artery, which has not been observed in any case of JTA. JTA is sometimes characterized by panarteritis, occasionally with a prominent eosinophilic infiltrate, intimal proliferation, discrete damage to the media and extensive cellular infiltrate made of lymphocytes, eosinophils and plasma cells in the perivascular tissue; granulomatous lesions or giant cells are rare. On laboratory evaluation, the erythrocyte sedimentation rate is normal, and the peripheral blood shows hypereosinophilia in one third of cases.

4.3 Nosology of JTA, Kimura disease and angiolymphoid hyperplasia with eosinophilia (ALHE)

Considering clinical description and evolution, a nosological overlap between “JTA – ALHE – Kimura” could be discussed.

4.4 Kimura and ALHE

Kimura disease and ALHE are soft-tissue inflammatory reactions, associated with variable degrees of vascular proliferation, often in close proximity to muscular arteries, which are sometimes involved by these diseases. The inflammatory infiltrate often contains numerous eosinophils. Kimura disease and ALHE are conditions that involve the head and neck region, tend to recur despite treatment, and share several histopathologic features, such as lymphoeosinophilic infiltrates of the involved tissue and vascular proliferation [22,44].

Kimura disease is a peculiar angiolymphoid proliferative disorder of soft tissue (with large subcutaneous or salivary gland masses) with eosinophilia and elevated IgE; it occurs almost exclusively in young Oriental men [13,15,45–47]. The early subcutaneous lesions show prominent vascular proliferation with plump histiocytoid endothelial cells and a predominantly lymphocytic infiltrate and abundant eosinophils. In later stages, vascular proliferation is still evident but less conspicuous, and nodular lymphoid hyperplasia and fibrosis become prominent with deep involvement of subcutaneous tissue and muscle. Kimura disease is not known to involve a specific artery, but it has a predilection for the head and neck, especially the periauricular and submandibular regions; there are frequent systemic manifestations, such as lymph node enlargement; involvement of cubital or inguinal regions are also described. A histopathologic pattern of lymphoid nodules with germinal centers

and numerous eosinophils including eosinophilic abscess formation is characteristic of Kimura disease [48]. Vascular proliferation may be present, but the characteristic cytomorphologic alterations of the endothelium seen in ALHE are not identified. It has an excellent prognosis, although it may recur in 30% and wax and wane over time [49].

ALHE is also a dermal and subcutaneous vascular proliferative lesion with prominent lymphofollicular inflammation, occurring most frequently in the head and neck. Patients are mostly Caucasian, between 20 and 50 years of age, and are equally male and female. In about 50% of ALHE, an arterial or arteriolar structure can be observed in close association with – or is the central site of – histiocytoid endothelial cell proliferation.

In addition to direct lesional morbidity, both ALHE and Kimura disease have been reported in association with renal disorders [50–53]. Both diseases have also been associated with the concurrent or subsequent development of T-cell lymphoma [54–62].

Histologically JTA, ALHE, and Kimura disease show eosinophilic infiltrates, and some degree of vascular proliferation. However, in JTA those inflammatory infiltrates are mostly confined to the vessel wall, while in Kimura disease and ALHE, the inflammatory infiltrates are mainly perivascular, and the vascular proliferation is often conspicuous.

It is not clear whether JTA is an independent entity or a presentation of Kimura disease or ALHE. This debate is clinically significant as the conclusion may affect clinical follow-up. To better understand the relationship between JTA and Kimura disease, we reviewed all previous reports of isolated JTA (i.e. without reported or histologically demonstrated signs for concomitant Kimura disease) and of JTA associated with Kimura disease. The following characteristics could be found: Firstly, isolated JTA is an inflammation of the temporal artery with numerous eosinophil infiltrates. It usually shows marked intimal thickening with narrowing of the vascular lumen. Secondly, in addition to these findings, JTA combined with Kimura disease is characterized by periadvential lymphovascular hyperplasia with lymphoid follicles.

JTA seems, from an etiopathogenic point of view, to be a disease distinct from Kimura disease and ALHE. JTA may be primitively a vascular disease where the inflammatory process initially affects the artery and can, by extension, affect the neighboring tissue and lead to the development of lymphoid follicles. Considering their histological description, we hypothesize that the inflammatory process in Kimura disease and ALHE first affects the dermis (ALHE) [63] or the subcutaneous tissue (Kimura disease) [45], be it of a vascular nature (with capillary proliferation) or not. Incidentally, Kimura disease is synonym for sub-cutaneous ALHE [3,11]. In both cases, the cranial region is most usually affected but the lesions can be more diffuse. Fukunaga *et al.* could not evidence vasculitis in twelve cases of Kimura disease [18]. These two entities – Kimura disease and ALHE – could, by extension, generate inflammation on the neighboring arteries; the temporal artery is the main neighboring artery for these two diseases when they affect the head. In this respect, Watanabe *et al.* reported a case of a temporal arteritis which emanated at its origin in the parotid space and was thus satellite of

a Kimura disease primitively affecting the parotid gland [31]. Therefore we could distinguish between JTA which is an authentic primitive vasculitis affecting the temporal from temporal arteritis which is secondary to angiolymphoid hyperplasia with eosinophilia, whose origin may be sub-cutaneous (Kimura disease) or cutaneous (ALHE).

4.5 Follow-up /relapse

In the 24-month follow-up period after the lesions were excised, most patients remained well and reported no recurrence. Most of the systemic treatments (steroids, NSAID, colchicine) were given from the onset, and not because of an unfavorable evolution. Unlike GCA, immunosuppressive therapies are rarely administered [64,65]. Only one case with multiple relapses required the use of methotrexate [29]. No case developed systemic vasculitis during follow-up. One case presented two years after JTA a T-cell lymphoma [25]. Several cases of ALHE and Kimura disease with concurrent or subsequent development of peripheral T-cell lymphoma have been reported [22]. Moreover, clonal T-cell receptor gene rearrangement was demonstrated in the tissue lesions of ALHE patients [55,59].

4.6 Strengths and limits

The main limit of our study is its retrospective nature and the literature review with non-exhaustive data. The main strength of this study is its multicentric nature which confirms the rarity of JTA.

JTA characteristics can be summarized as follows:

- JTA is a benign and local disease affecting patients up to 50 years of age.
- It may be associated with minor systemic clinical symptoms such as asthenia, headache or visual blur.
- Clinical examination is normal apart from a lump in the temporal region.
- Eosinophilia may be present.
- A moderate biological inflammatory syndrome is possible.
- Pathological analysis classically shows arteritis predominating in the intima rather than the media, with a possible extension to the perivascular tissue.
- The presence of granuloma, giant cells or fibrinoid necrosis is rare.
- The clinical context without general impairment or marked biological inflammation, combined with an age inferior to 50, makes the diagnosis of GCA very unlikely.
- The evolution is usually favorable either spontaneously or after local treatment (complete excision). The diagnosis is confirmed after several months of follow-up with no symptoms of systemic vasculitis.

- There is most of the time only one episode, but relapses are possible. Their repetition must lead physicians to reconsider the diagnosis, especially if they need systemic treatment (steroids or immunomodulatory drugs).

5. Conclusion

JTA is a rare, localized and benign disease, which affects up to middle-aged adults – but under 50 year-old – and men more than women. Headaches may be present in half of cases. The presence of clinical signs – local (other than the presence of a lump and headaches) or systemic – was uncommon, as was the case for inflammatory biological syndrome. Eosinophilia concerned one third of patients. More than 80% of cases present only one episode and require only local treatment by excision. Oral steroids were prescribed in 20.9% of cases, usually straightaway and not because of the failure of local treatment. Pathology studies showed arteritis in 97.6% of cases with pan-arteritis only in 78.9%, with lesions predominating on the intima rather than the media, a cellular infiltrate of a variable nature, and the presence of granuloma in 22.9% and of giant cells in 25%. Perivascular extension and/or the presence of lymphoid follicles or germinal centres should prompt the physician to discuss the possibility of ALHE or Kimura disease as alternate diagnosis, which appear to us as being quite distinct pathologies.

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	French cohort (n=12)	Literature (n=32) Literature references [3–29]	Total
Clinical data	n/n (%)	n/n (%)	n/n (%)
Median age [min-max]	38.5 [22-44]	25.5 [7-39] [3–29]	30 [7-44]
Male	10/12 (83.3%)	24/32 (75.0%) [3,5,7–21,25–27,29]	34/44 (77.3%)
Temporal headache	6/12 (50.0%)	15/32 (46.9%) [4–6,8–11,14,16,19,21,22,25–27]	21/44 (47.7%)
Temporal lump	11/12 (91.7%)	21/32 (65.6%) [8–10,12,13,15,17,18,20,23,24,28,29]	32/44 (72.7%)
Initial bilateral temporal involvement	4/12 (33.3%)	7/32 (21.9%) [12,13,16,25,27,29]	11/44 (25.0%)
General symptoms	1/12 (8.3%)	4/32 (12.5%) [4–7]	5/44 (11.4%)
Temporal itching	2/12 (16.7%)	2/23 (8.7%) [9,24]	4/35 (11.4%)
Visual blurring or phosphenes	1/12 (8.3%)	3/29 (10.3%) [4,6,11]	4/41 (9.8%)
Temporo-mandibular pain	0/11 (0%)	2/24 (8.3%) [5,6]	2/35 (5.7%)
Biological data			
Inflammatory syndrome	1/12 (8.3%)	2/32 (6.3%) [6,23]	3/44 (6.8%)
Eosinophilia > 500/mm ³	5/12 (41.7%)	9/29 (31.0%) [8,9,13,15,18,19,23,27,29]	14/41 (34.1%)
Initial treatment			
Complete excision without further treatment	9/12 (75.0%)	23/32 (71.9%) [3,4,8–10,12,14,15,17–19,21–24,26,28,29]	32/44 (72.7%)
Corticosteroids	2/12 (16.7%)	6/31 (19.3%) [5–7,20,25,27]	8/43 (18.6%)
Colchicine	2/12 (16.7%)	0/31 (0%)	2/43 (4.7%)
NSAID	0/12 (0%)	1/31 (3.2%) [14]	1/43 (2.3%)
Evolution			
Median follow-up in months [min-max]	26 [1-219]	24 [1-60]	24 [1-219]
Local relapse	2/12 (16.7%)	5/31 (16.1%) [5,14,19,26,29]	7/43 (16.3%)
Progression into systemic vasculitis	0/12 (0%)	0/31 (0%)	0/43 (0%)

Table 1: Clinical, biological characteristics and outcome of JTA.

	French cohort (n=12)	Literature (n=32) Literature references [3–29]	Total
Histology n/n (%)			
Arteritis	11/12 (91.7%)	30/30 (100%) [3–23,25–27,29]	41/42 (97.6%)
Pan-arteritis	5/9 (55.6%)	10/10 (100%) [3,4,13,17,18,20,21,27,29]	15/19 (78.9%)
Intra-luminal thrombosis	7/9 (77.8%)	14/22 (63.6%) [3,5,6,8,9,11,16,19–21,25,26]	21/31 (67.7%)
Sparse giant cell	2/11 (18.0%)	8/29 (27.6%) [5–10,14,20]	10/40 (25.0%)
Granuloma	2/10 (20.0%)	6/25 (24.0%) [3,6,7,14,15]	8/35 (22.9%)
Fibrinoid necrosis	0/11 (0%)	2/25 (8.0%) [5,8]	2/36 (5.6%)
Intima n/n (%)			
Hyperplasia or intimal thickening (*) / endothelial proliferation	9/10 (90.0%)	23/23 (100%) [3–5,7,9–12,15–18,20–24,26,27,29]	32/33 (97.0%)
Media n/n (%)			
Disruption of the internal elastic lamina	5/7 (71.4%)	20/21 (95.2%) [3,4,7–11,13,18,20–23,26,27,29]	25/28 (89.3%)
Disorganization or destruction of the media	3/7 (42.9%)	8/8 (100%) [3,5,10,18–20]	11/15 (73.3%)

Nature of the cellular infiltrate n/n (%)						
Eosinophilic	7/8	(87.5%)	22/25	(88.0%)	[3,4,6,8,10–13,15,17–19,21–23,25,26,28,29]	29/33 (87.9%)
Lymphocytic	7/9	(77.8%)	15/18	(83.3%)	[3,4,6,9,10,12,13,18,20,22,26–29]	22/27 (81.5%)
Mononuclear	3/7	(42.9%)	12/21	(57.1%)	[3,6,7,9,10,12,16,21,27,29]	15/28 (53.6%)
Plasmocytic	1/6	(16.7%)	5/21	(23.8%)	[3,4,18,20]	6/27 (22.2%)
Perivascular n/n (%)						
Perivascular extension of the inflammation	7/8	(87.5%)	12/15	(80.0%)	[3,4,8,11,17,18,20,22–24,29]	19/23 (82.6%)
Lymphoid follicles / germinal centres	1/3	(33.3%)	8/12	(66.7%)	[3,4,17,18,23,24,27]	9/15 (60.0%)

Table 2: Pathological characteristics of JTA.

Figure 1: Clinical presentation of bilateral JTA. Notice the prominent aspect of the temporal arteries.

Figure 2: Sections of temporal arteries showing different histopathological patterns : 1/ a non granulomatous obliterant panarteritis with a prominent intima (A, B), 2/ a non granulomatous panarteritis with mononuclear cells and eosinophils, newly formed capillaries (*) lined by plump endothelial cells in the media and the adventitia and an intimal hyperplasia (C,D) and 3/ a thrombosis with numerous capillaries and numerous eosinophils (E,F) (HES ; original magnifications x 40 and 200 ; blue arrow: media ; black arrow : intima).



