Head-and-neck swelling: an under-recognized feature of giant cell arteritis. A report of 37 patients

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ABSTRACT

Objective. To describe the frequency of occurrence and characteristics of head-and-neck swelling (HNS) in temporal (giant cell) arteritis (TA).

Methods. We analyzed the charts of patients with HNS retrieved from a single department series of 260 consecutive patients with TA and reviewed the published French-English literature. Patients with a swelling limited to the temporal fossa were excluded.

Results. A history of HNS was elicited in 17 patients in our series (i.e. 6.5%) and in 20 previously published patients. The swelling was an inaugural feature in most cases and was often transient. Ear-nose-and-throat (ENT) symptoms were observed in 80% of the cases, including jaw claudication or pain upon opening mouth in 22, causing trismus in 10. Two patients had permanent visual impairment and 1 had sudden hearing loss. The temporal artery biopsy yielded giant cell arteritis (GCA) in all the patients but 2. The HNS was often painful and mainly involved mainly the orbital region and face, particularly the lower part of the cheeks and maxillae, less often the neck and, rarely, the forehead and tongue. Concurrent localized limb swellings were also observed in 3 patients. The HNS disappeared in all the patients, either spontaneously or under steroid treatment, and recurred only in 6 patients.

Conclusion. HNS is not exceptional in untreated TA and is strongly associated with ENT symptoms and a positive TAB, but not with visual loss or stroke. Such characteristics imply in these cases a prominent, widespread involvement of the external carotid artery system by giant cell arteritis.

Introduction

Giant cell arteritis (GCA) or temporal arteritis (TA) is a systemic vasculitis of the elderly which affects large and medium-sized arteries, predominantly the branches of external carotid artery. The most common cranial symptoms of TA are headaches, scalp tenderness, visual disturbances and jaw claudication. Other presentations of GCA are much more unusual but are important to recognize so that the potentially catastrophic, but often delayed, ischemic complications of the disease can be avoided. Facial swelling as a prominent feature of TA has seldom been reported (1-4), though mentioned in the past English literature (5, 6). Thus, owing to a relative lack of awareness of this aetiology in the differential diagnosis of facial swelling, delayed or misleading diagnosis, as well as inappropriate therapeutic options, may ensue. In this paper, we report a personal series of 17 patients with head-and-neck swelling (HNS) heralding GCA and survey all similar cases published in the French-English literature, with the aim of reinforcing current knowledge of this feature of TA.
174 items, then transferred to a computerised database. Special efforts were made in evaluating the delay to diagnosis from the onset of symptoms of vasculitis, the presence of constitutional syndrome (defined by a temperature ≥ 38°C for at least one week, severe asthenia, and/or weight loss > 5%), polymyalgia rheumatica (PMR) (9), abnormal temporal artery on examination (absence of pulses on all or part of its course, nodules, thickening, swelling or tenderness on palpation) and upper limb artery involvement (presence of intermittent arm claudication, absent or decreased radial pulse, Raynaud’s phenomenon of recent onset, suggestive findings on selective aortic arch arteriography (10), or at least a murmur heard over subclavian-axillary arteries at admission or within a month). Ear-nose-throat (ENT) symptoms were meticulously recorded and included jaw claudication (11), pain upon opening mouth, trismus, maxillary or dental pain, otalgia, odynophagia, lingual pain or discomfort, throat, dysphagia, dry cough, and carotidynia (12). The occult (or silent) form of TA has been defined previously (31).

Head-and-neck swelling
The finding of HNS, its characteristics and outcome, were recorded in the patient’s clinical history, since the questionnaire did not involve such a pre-established item. Cases with a mentioned HNS were extracted from the whole series and the patient’s chart was thoroughly reviewed. All patients with an unquestionable history of HNS that had lasted at least one day were included in the study, irrespective of the patient’s head and neck appearance at admission. Patients in whom the swelling was confined to the temporal fossa or faced an enlarged salivary gland (14, 15) were excluded.

Literature survey
French or English papers published since 1970 dealing with either GCA, TA or polymyalgia rheumatica (PMR) and facial or neck swelling were reviewed, as well as French congress abstracts. Patient characteristics, clinical presentation, temporal artery biopsy findings and outcome were analyzed. Well-described reports of HNS occurring concurrently with TA were included in the study.

Results
Features of TA in patients with HNS
A history of HNS was elicited in 17 out of 260 patients, i.e. 6.6% of the series, whilst the survey consisted of 20 case reports (1-4, 16-26). The main pre-treatment characteristics of GCA in these 37 patients are summarized in Table I. The age range was 57 to 90 years (mean 75.4 years) and 69% were women. The time to diagnosis from the first symptoms averaged 7 weeks, though the disease onset was acute in 15 out of 25 reported cases and 72% of the patients developed typical cranial arteritis. At least one ENT symptom was recorded in 80% of the cases, with a maximum of 6 symptoms in the same patient. Ear-and-throat symptoms were, apart from HNS, the only presenting manifestations of GCA in 3 patients. Jaw claudication and/or pain upon opening mouth involved 22 patients, with frank trismus in 10 (27%). Six patients had eye or orbital pain and 12 had transient visual ischemic symptoms but only 2 (6%) developed permanent visual loss. Extra-cranial manifestations were recorded in 32% of the patients. The erythrocyte sedimentation rate exceeded 50 mm/h in 87% of the cases and the temporal artery biopsy (TAB) yielded GCA in 33 out of 35 verified cases.

Characteristics of HNS in patients with TA
The main characteristics of TA-associated HNS are shown in Table II. Facial puffiness was frequently an early or even inaugural sign of TA but was often transient and had subsided on the first admission in half of the patients. The swelling lasted less than 1 one week in 60% of the cases with a maximum duration of 2 months. It was bilateral in nearly three-fourths of the cases and involved predominantly the face and cheeks (23 cases), the eyelids and orbital region (15 cases) and the anterior and lateral aspects of the neck (7 cases), less often the tongue (3 cases), forehead and scalp (2 cases), chin and supra-clavicular space (1 case each). Some patients or their relatives recalled a downward displacement of initial swelling from the cheeks to the maxilla and upper parts of the neck. Three patients had segmental arm and/or leg swellings concurrently with HNS; none of these had acquired angioedema. Patients often complained of pain or discomfort in parts of the face or neck precisely where the swelling developed shortly thereafter, but some felt the HNS painful by itself. On physical examination swelling was discretely pinkish in 8 out of 12 patients and generally not pitting or itchy. The HNS outcome was known in 32 patients. Seven noticed a relapsing or waxing-and-waning course. The swelling disappeared spontaneously (n = 13) or quickly upon corticosteroid treatment (n = 19) but recurred in 6 patients, with multiple recurrences on further attempts to taper the prednisone in one (24).

Discussion
In a patient with peri-orbital, facial and/or neck swelling, there is a lengthy list of diagnostic possibilities to make out, including notably allergic oedema and photosensitive reactions, bacterial, virus or parasitic infections, endocrine disorders, inherited or acquired angio-neurotic oedema, acute onset systemic sclerosis or dermatomyositis, superior vena cava syndrome or jugular vein thrombosis, nephrotic syndrome and other causes of hypoproteinemia. Although swelling in the temporal fossa is a well-recognized feature of GCA, more diffuse swelling of the face as a manifestation of GCA has been described in the English literature only in a handful of cases (1-4). Likewise, available the French literature is scanty with several reports published only in an abstract form (16-26). Therefore, in an elderly person presenting with a swollen face, the degree of suspicion concerning the possibility of GCA is currently low, due to lack of awareness, with the risk of blurring more evoking symptoms. This is illustrated by a rather long average delay to diagnosis in these patients and by the fact that some were first seen by laryngologists with a
presumed diagnosis of sinusitis, since they presented with headaches, facial or periorbital pain and raised ESR. In other patients, the sudden onset of HNS or a concurrent involvement of the tongue, lips or cheek mucosa led to misdiagnose TA as allergic oedema. The appearance of HNS in TA was varied and was in no way pathognomonic. The most common picture was a bilateral (or less often unilateral) painful, slightly pink, non-pitting swelling of the cheeks and maxillae, but other areas could be affected, including the eyelids, orbital region, neck, forehead, cheek mucosa and tongue. Thus, these patients can be first seen by an ear, nose and throat specialist or an ophthalmologist or be sent to the emergency room, before the possibility that a vasculitic process is taken into consideration.

Facial or neck swelling appears to be an underreported early feature of GCA, since it was observed in a sizeable portion of our patients, nearly 7%. Furthermore, this figure is a minimal estimate since no specifically designed item was devoted to HNS in our prospective questionnaire. Moreover, in many cases the diagnosis of HNS was only based on the patient’s history. In a prospective study of 345 patients with TA, the frequency of facial oedema was 12% (27). The discrepancy existing between the frequency of HNS in our study and the aforementioned study, and the rarity ascribed to this finding in the literature, is best explained by the use of questionnaires and prospective study designs, which allow the recognition of discrete and

<table>
<thead>
<tr>
<th>Author (ref)</th>
<th>Age / sex</th>
<th>Onset</th>
<th>Presenting features of TA</th>
<th>ESR (mm/h)</th>
<th>TAB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cohen (1)</td>
<td>62 / F</td>
<td>acute</td>
<td>glossitis, odynophagia and maxillary pain</td>
<td>73</td>
<td>Pos</td>
</tr>
<tr>
<td>Herreman (16)</td>
<td>87 / M</td>
<td>n.s.</td>
<td>cranial, with PMR and JC</td>
<td>120</td>
<td>Pos</td>
</tr>
<tr>
<td>Acetta (2)</td>
<td>68 / M</td>
<td>n.s.</td>
<td>cranial, with PMR and JC</td>
<td>68</td>
<td>Pos</td>
</tr>
<tr>
<td>Delvigne (17)</td>
<td>71 / F</td>
<td>acute</td>
<td>cranial, with diaphragm and AF</td>
<td>140</td>
<td>Pos</td>
</tr>
<tr>
<td>Manganelli (4)</td>
<td>62 / F</td>
<td>acute</td>
<td>cranial and PMR and PV</td>
<td>94</td>
<td>Pos</td>
</tr>
<tr>
<td>Planerin (18)</td>
<td>84 / F</td>
<td>acute</td>
<td>cranial and PMR and PV</td>
<td>68</td>
<td>n.d.</td>
</tr>
<tr>
<td>Acetta (2)</td>
<td>68 / F</td>
<td>acute</td>
<td>cranial and PMR and PV</td>
<td>95</td>
<td>Pos</td>
</tr>
<tr>
<td>Gaches (21)</td>
<td>79 / M</td>
<td>acute</td>
<td>cranial, with JC and bilateral PVL</td>
<td>n.a.</td>
<td>Pos</td>
</tr>
<tr>
<td>Sarraf (20)</td>
<td>77 / F</td>
<td>acute</td>
<td>cranial and PMR and JC, sore throat, dysphagia and AF</td>
<td>54</td>
<td>Pos</td>
</tr>
<tr>
<td>Paris (23)</td>
<td>88 / F</td>
<td>acute</td>
<td>cranial, with JC and bilateral PVL</td>
<td>n.a.</td>
<td>Pos</td>
</tr>
<tr>
<td>Paulus (24)</td>
<td>85 / F</td>
<td>acute</td>
<td>cranial, with JC and bilateral PVL</td>
<td>n.a.</td>
<td>Pos</td>
</tr>
<tr>
<td>Regnoky (25)</td>
<td>70 / M</td>
<td>acute</td>
<td>cranial, with JC</td>
<td>80</td>
<td>Pos</td>
</tr>
<tr>
<td>Myara (26)</td>
<td>78 / F</td>
<td>acute</td>
<td>cranial, with diaphragm (no headaches)</td>
<td>42</td>
<td>Pos</td>
</tr>
<tr>
<td>Current series</td>
<td>79 / F</td>
<td>progressive</td>
<td>cranial, with JC, sore throat, hoarseness, dry cough, otalgia and AF</td>
<td>92</td>
<td>Pos</td>
</tr>
<tr>
<td>86 / F</td>
<td>progressive</td>
<td>cranial, with JC, PLOM, lingual pain, dysphagia and AF</td>
<td>130</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>78 / F</td>
<td>progressive</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>59</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>81 / F</td>
<td>acute</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>53</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>77 / M</td>
<td>acute</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>72</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>72 / F</td>
<td>acute</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>134</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>78 / M</td>
<td>progressive</td>
<td>occult (silent) form, then eye pain</td>
<td>102</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>76 / M</td>
<td>progressive</td>
<td>cranial, with JC, dry cough and otalgia</td>
<td>42</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>72 / F</td>
<td>progressive</td>
<td>cranial, with JC, PLOM, maxillary pain, dysphagia and AF</td>
<td>46</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>75 / F</td>
<td>acute</td>
<td>cranial and upper limb artery involvement, with maxillary pain</td>
<td>113</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>79 / M</td>
<td>acute</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>60</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>78 / F</td>
<td>acute</td>
<td>cranial, with JC, maxillary pain, otalgia and dysphagia</td>
<td>134</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>81 / F</td>
<td>acute</td>
<td>maxillary pain, without headaches</td>
<td>60</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>76 / F</td>
<td>progressive</td>
<td>cranial, with PLOM, maxillary pain, without headaches</td>
<td>90</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>90 / F</td>
<td>acute</td>
<td>maxillary pain, without headaches</td>
<td>138</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>76 / F</td>
<td>progressive</td>
<td>occult (silent) form, then eye pain</td>
<td>102</td>
<td>Pos</td>
<td></td>
</tr>
<tr>
<td>65 / M</td>
<td>progressive</td>
<td>cranial and PMR, with hoarseness</td>
<td>48</td>
<td>Pos</td>
<td></td>
</tr>
</tbody>
</table>

TA: temporal arteritis; PMR: polymyalgia rheumatica; TAB: temporal artery biopsy; JC: jaw claudication; AF: amaurosis fugax; PVL: permanent visual loss; PLOM: pain upon opening mouth; VH: visual hallucinations; Ns: not specified; Nd: not determined.
**Table II. Characteristics of head-and-neck swelling in patients with concurrent temporal arteritis.**

<table>
<thead>
<tr>
<th>Author (Ref)</th>
<th>Location</th>
<th>Characteristics</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cohen (1)</td>
<td>neck, face, cheek, periorbital (bilat)</td>
<td>n.s.</td>
<td>settled within 2 days with CS</td>
</tr>
<tr>
<td>Herreman (16)</td>
<td>periorbital</td>
<td>n.s.</td>
<td>n.s.</td>
</tr>
<tr>
<td></td>
<td>eyelid, lips, limbs</td>
<td>pinkish, migratory, discretely tender</td>
<td>n.s.</td>
</tr>
<tr>
<td>Averett (2)</td>
<td>forehead, face, maxillae (bilat)</td>
<td>painful, fluctuating</td>
<td>n.s.</td>
</tr>
<tr>
<td>Deligny (17)</td>
<td>periorbital (bilat)</td>
<td>thickened, tender maxillary arteries</td>
<td>settled without CS, relapsed and subsided with CS</td>
</tr>
<tr>
<td>Mangone (4)</td>
<td>lower part of the face (bilat)</td>
<td>n.s.</td>
<td>settled within 3 days with CS (80 mg/d)</td>
</tr>
<tr>
<td>Chevalier (19)</td>
<td>face, cheek</td>
<td>skin tenderness facing swelling</td>
<td>settled without treatment</td>
</tr>
<tr>
<td>Ghinelli (3)</td>
<td>face, periorbital</td>
<td>“heavy” feeling after swelling had settled</td>
<td>settled without treatment</td>
</tr>
<tr>
<td>Plaut (18)</td>
<td>face, periorbital (bilat)</td>
<td>pinkish, more prominent in the morning</td>
<td>settled with CS (15 mg)</td>
</tr>
<tr>
<td></td>
<td>periorbital (bilat)</td>
<td>pinkish, tender</td>
<td>settled within a few days with CS (25 mg)</td>
</tr>
<tr>
<td></td>
<td>lower part of the face, scalp</td>
<td>n.s.</td>
<td>settled after a short CS cure, multiple relapses then subsided with continuous CS (20 mg/d)</td>
</tr>
<tr>
<td>Gaches (21)</td>
<td>check, eyelid (bilat)</td>
<td>n.s.</td>
<td>settled rapidly with CS</td>
</tr>
<tr>
<td></td>
<td>check (bilat)</td>
<td>relapsing course</td>
<td>settled rapidly with CS</td>
</tr>
<tr>
<td>Safai (20)</td>
<td>face, suprACLavicular space (bilat)</td>
<td>n.s.</td>
<td>settled rapidly with CS but relapsed</td>
</tr>
<tr>
<td></td>
<td>lower part of the face</td>
<td>pinkish</td>
<td>settled rapidly</td>
</tr>
<tr>
<td>Sarrot-Rey (22)</td>
<td>neck, face, cheek (bilat)</td>
<td>“inflammatory” appearance</td>
<td>settled within 10 days with CS (1 mg/kg)</td>
</tr>
<tr>
<td>Paris (23)</td>
<td>check, periorbital, neck</td>
<td>pinkish, sensible</td>
<td>settled rapidly with CS</td>
</tr>
<tr>
<td>Paulus (24)</td>
<td>periorbital, malar region</td>
<td>pinkish, pitting, painless</td>
<td>settled within 4 days with CS (1 mg/kg), numerous relapses</td>
</tr>
<tr>
<td>Regent (25)</td>
<td>lower part of the cheek (bilat)</td>
<td>pinkish, pitting, tender, enhancing overnight</td>
<td>settled rapidly with CS (60 mg/d) but relapsed</td>
</tr>
<tr>
<td>Myara (26)</td>
<td>face, eyelids (bilat)</td>
<td>white, pitting, painless</td>
<td>settled within a few days with CS (0.7 mg/kg/d)</td>
</tr>
<tr>
<td>Current series</td>
<td>face, maxillae, chin, neck (bilat)</td>
<td>painless, “non inflammatory”, lasted 1 month</td>
<td>settled without treatment</td>
</tr>
<tr>
<td></td>
<td>face, neck, suprACLavicular, tongue</td>
<td>associated with segmental limb swellings, lasted 2 weeks</td>
<td>settled without treatment</td>
</tr>
<tr>
<td></td>
<td>periorbital, eyelids (bilat)</td>
<td>preceded by orbital and facial pain</td>
<td>settled quickly without treatment</td>
</tr>
<tr>
<td></td>
<td>periorbital, eyelids (bilat)</td>
<td>preceded by eye and orbital pain</td>
<td>settled quickly without treatment</td>
</tr>
<tr>
<td></td>
<td>face, temporal fossa (bilat)</td>
<td>short-lived (3 days)</td>
<td>settled without treatment</td>
</tr>
<tr>
<td>neck (anterior-lateral aspects) (bilat)</td>
<td>n.s.</td>
<td>treated rapidly with intravenous CS</td>
<td></td>
</tr>
<tr>
<td>periorbital, eyelid</td>
<td>waning and waxing course</td>
<td>settled within 1 month without treatment</td>
<td></td>
</tr>
<tr>
<td>face, check and neck (bilat)</td>
<td>n.s.</td>
<td>settled rapidly with CS (20 mg/d)</td>
<td></td>
</tr>
<tr>
<td>face, cheek</td>
<td>n.s.</td>
<td>settled rapidly with CS, relapsed (6th week), subsided definitively (6th mo)</td>
<td></td>
</tr>
<tr>
<td>neck (anterior-lateral aspects) (bilat)</td>
<td>waning and waxing course</td>
<td>settled within 2 weeks without treatment</td>
<td></td>
</tr>
<tr>
<td>periorbital, eyelids (bilat), root of nose</td>
<td>short-lived (5 days)</td>
<td>settled quickly with CS (0.7 mg/kg)</td>
<td></td>
</tr>
<tr>
<td>face, check</td>
<td>short-lived (7 days)</td>
<td>settled without treatment</td>
<td></td>
</tr>
<tr>
<td>face</td>
<td>painless, short-lived (a few days)</td>
<td>settled without treatment</td>
<td></td>
</tr>
<tr>
<td>eyelids, neck (submaxillary), tongue</td>
<td>painless, short-lived</td>
<td>settled quickly with CS (20 mg/d)</td>
<td></td>
</tr>
<tr>
<td>face, check, lateral aspect of the neck (bilat)</td>
<td>“non inflammatory”, painless</td>
<td>settled quickly with CS (25 mg/d)</td>
<td></td>
</tr>
<tr>
<td>face, check (bilat)</td>
<td>short-lived (two episodes)</td>
<td>settled without treatment</td>
<td></td>
</tr>
<tr>
<td>current series</td>
<td>maxillae, neck (lateral aspect)</td>
<td>“non inflammatory”, painless</td>
<td>settled without treatment</td>
</tr>
</tbody>
</table>

Ns: not specified; CS: corticosteroids.
short-lived forms of HNS. Patients with HNS in the setting of TA shared some important characteristics. Though the swelling was often an early or even the first manifestation of the disease, it remained very rarely the sole finding for prolonged periods (12). Moreover, less than 10% of the patients presented without headaches or jaw claudication and in none of them was HNS the only cranial symptom of GCA. The most striking finding in our study was that 80% of the patients had ENT symptoms and nearly 80% developed trismus. Patients with GCA complain of varied ear-jaw-mouth-threat symptoms in up to 57% of the cases (13). These symptoms are probably caused by an inflammatory involvement of branches of the external carotid artery such as facial, internal maxillary and ascending pharyngeal arteries, although this has been rarely documented (28, 32). Trismus has been only occasionally reported in TA (29, 33-35), where it may be considered as the greatest degree of jaw claudication (35). Although such an association of HNS with trismus may be overemphasized due to publication biases, this is unlikely in our unselected series, where 3 out of 4 patients who presented with frank trismus, also had HNS. The cause of HNS in TA is unclear, but in most cases its almost simultaneous appearance with facial or periorbital pain, jaw claudication or trismus as well as other ENT symptoms and a positive TAB result, and its prompt regression with corticosteroid treatment suggest that it may be due to a widespread inflammatory involvement of the external carotid tree. Carotid tenderness, which points to an inflammatory involvement of the external carotid artery itself in patients with TA (36), might also rely on HNS. Unfortunately, this feature was not included as a specific item in our questionnaire and examination of the external carotid artery was generally left aside in Doppler studies. Alternatively, abnormal release of vascular endothelial growth factor, a protein that both induces hyperpermeability and stimulates endothelial cell growth (37), could play a role in the development of local or distant swellings in some patients with GCA (38, 39). Contrary to previous reports (3, 40), our study showed no association of facial swelling with sight-threatening complications, although one-half of the patients reported either transient ischamic visual symptoms or eye pain. Friedman and Friedman described the sensation of puffiness of the face without altered facial appearance as a predictor of visual disturbance in TA (40). Similarly, Ghanchi et al. described a patient in whom such a sensation preceded objective facial swelling and visual impairment (3). Visual loss in GCA could be associated, therefore, with subjective rather than true facial puffiness. Likewise, no patients with HNS developed stroke, a manifestation that has been strongly associated with permanent visual loss (41). Why patients with TA and HNS seldom develop irreversible visual loss is unclear. However, most patients described in this study presented with an acute disease onset and an otherwise typical picture of cranial arteritis, which may have prompted caregivers to act preventively before the development of irreversible complications. Finally, variations in the clinical presentation of GCA were found to be correlated with cytokine mRNA expression, the risk of visual ischemic complications being higher in patients whose temporal artery biopsy express high amounts of interferon-γ mRNA (42, 43). One can therefore speculate about the possibility of different cytokine profiles in GCA patients with cranial arteritis and HNS and without.

HNS associated with TA ran a benign course but recurred in some patients in attempts to taper corticosteroids, pointing to a poorly controlled underlying process. In these patients the recurring swelling could not be attributed solely to a corticosteroid side-effect because it responded promptly to further increases in the prednisone dose. Similarly, sinustitis with facial swelling (44) was reasonably ruled out, since none of the patients were receiving antibiotics during the swelling relapse or subsequently. In conclusion, HNS appears to be an under-recognized, early feature of TA. Objective puffiness of the face and/or neck is strongly associated with ENT symptoms, particularly jaw claudication or trismus and a positive TAB, but not visual loss or stroke. Such characteristics imply in these cases a widespread involvement of the external carotid artery system by GCA, with relative sparing of the internal carotid artery system. This case series of patients with HNS underlines the preferential attack of the external carotid tree by GCA for reasons that remain to be elucidated.

References
Head-and-neck swelling in GCA / E. Liozon et al.


