Unusual variants in the presentation of temporal arteritis

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Summary
A retrospective study was made of 96 patients diagnosed as cranial arteritis of whom 32 were accepted using strict clinical criteria or a positive temporal artery biopsy. Unusual presentations of fever, psychiatric illness, headache-free patients and a 'normal' ESR are described. The recognition of these variations is important in the early diagnosis of temporal arteritis.

Introduction
Temporal (syn. giant-cell, cranial) arteritis as characterized by severe headache, malaise, jaw claudication and visual symptoms is a well recognized clinical entity (Horton, Magarth and Brown, 1932; Hutchinson, 1890; Healey, 1977; Harrison and Bevan, 1967. Recently, it has been noted that many non-cranial vessels are also affected. This widespread involvement gives rise to well known clinical variants in this condition including polymyalgia rheumatica and ophthalmoplegia (Barricks et al., 1977). In this paper, 4 less common but clinically important variants will be described, these are: patients with fever, psychiatric illness, headache-free patients and a group with a 'normal' ESR.

Patients and methods
The case notes were obtained of all patients with a diagnosis of temporal arteritis or cranial arteritis, admitted between 1969 and 1979. Of 96 case notes, 32 were accepted for the study. Patients were only accepted if they had a positive temporal artery biopsy (13 out of 32—41%), or classical manifestations of temporal arteritis including a response to steroids and at least 4 of the following:

(i) raised ESR; (ii) severe headache; (iii) polymyalgia rheumatica; (iv) tender, swollen vessels; (v) jaw claudication, or blindness.

Criteria for a positive temporal artery biopsy included intimal thickening with possible thrombosis; a granulomatous replacement of the media and partial destruction of the internal elastic lamina; at least one giant cell and inflammatory cells – mono-

nuclear, polymorphs or plasma cells. The latter were seen in the media or adventitia in all cases.

Of the 32 patients accepted 14 were male. The mean age was 68·8 years, ranging from 59 to 85 years.

Results
Fever
In this study, 7 of 32 patients presented with a non-specific febrile illness in which headache was absent or mild as shown in Table 1. The most commonly associated symptom was sweating which occurred during most nights in 6 out of 7 patients, the remaining patient suffering sweats during the day. One of these patients complained of rashes. These 7 all complained of general malaise and 6 had muscle pain and stiffness affecting the shoulder girdle. The range of ESR was 35 to 130 with a mean of 103 mm/hr. The WCC ranged from 5·2 to 14·9 with a mean of 9·2×10^9/l.

Case I. A 69-year-old housewife presented with a 3-week history of lassitude and weakness with nausea but no vomiting. She had felt feverish and had been given co-trimoxazole and nalidixic acid for a presumed urinary tract infection, but she had no response. For 3 weeks she had several rigors and frequent night sweats. She had been anorexic for one year and lost 3·3 kg in weight. On examination her maximum temperature was 38·5°C. There was an irregular tender mass over the left temple, her temporal arteries were pulsatile. Examination was otherwise normal. Her Hb was 13·0 g/dl, WCC 10·9×10^9/l, ESR 105 mm/hr. Whilst in hospital her temperature varied (Fig. 1). A temporal artery biopsy showed that the mass consisted of inflammatory infiltration with a giant-cell reaction and narrowing of the arterial lumen due to intimal thickening. She was given prednisolone 60 mg/day. Her pyrexia settled and she was discharged taking 15 mg of prednisolone/day. After 6 months she complained of nocturnal sweats with fever and aching in her limbs and trunk. She was taking 15 mg of prednisolone/day and her ESR was 47 mm/hr. Her
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TABLE 1. Patients presenting with fever

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age (years)</th>
<th>Symptoms (sweats)</th>
<th>Max. temp. (°C)</th>
<th>ESR (mm/hr)</th>
<th>WCC (x 10^9/l)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>75</td>
<td>Increasing</td>
<td>37-7</td>
<td>133</td>
<td>10-4</td>
</tr>
<tr>
<td>Female</td>
<td>71</td>
<td>At night</td>
<td>37-7</td>
<td>88</td>
<td>10-2</td>
</tr>
<tr>
<td>Female</td>
<td>69</td>
<td>At night</td>
<td>38-2</td>
<td>105</td>
<td>13-0</td>
</tr>
<tr>
<td>Female</td>
<td>68</td>
<td>At night</td>
<td>37-5</td>
<td>128</td>
<td>11-7</td>
</tr>
<tr>
<td>Female</td>
<td>76</td>
<td>Persistent</td>
<td>38-0</td>
<td>103</td>
<td>5-2</td>
</tr>
<tr>
<td>Male</td>
<td>75</td>
<td>Persistent</td>
<td>37-5</td>
<td>35</td>
<td>10-9</td>
</tr>
<tr>
<td>Female</td>
<td>62</td>
<td>At night, rigors</td>
<td>37-7</td>
<td>130</td>
<td>14-9</td>
</tr>
</tbody>
</table>

FIG. 1. Temperature chart of patient 1.

steroids were increased to 20 mg/day. Two weeks later she felt better but had suddenly lost vision in her left eye. Examination showed a partial branch occlusion of the left retinal artery. She continued on 20 mg of prednisolone/day and was well 5-5 years later, taking 7-5 mg of prednisolone/day.

Psychiatric changes

Three patients out of 32 presented with mental illness, as shown in Table 2. All 3 patients complained of headache and none had muscle aches or stiffness. The average ESR was 79 mm/hr (range 70-96 mm/hr).

Case 2. A 66-year-old night watchman with a history of loss of vision and headaches was transferred by a psychiatrist. Five months previously his vision had partially deteriorated, but one week ago he had become blind. He had suffered from severe generalized headaches for 5 weeks. He thought his family were all plotting against him and that his wife was being unfaithful. He had visual hallucinations, seeing crocodiles and the R.A.F. attacking him. He did not complain of joint or muscle pain. On examination he was deaf, and blind in both eyes. His fundi appeared normal. A space-occupying lesion was suspected. The investigations were: Hb 14-2 g/dl, WCC 12-3 x 10^9/l; ESR 45; X-rays of chest and skull and isotope brain scan were normal. A psychotic episode was witnessed on the ward and the ESR was 70 mm/hr. He was treated with 60 mg prednisolone/day – a presumptive diagnosis of temporal arteritis being made. A temporal artery biopsy was negative. His behaviour improved and his ESR quickly returned to normal. After 3 months he was symptom-free taking prednisolone 10 mg/day.

The mental changes were attributed to cerebral infarction caused by cranial arteritis; although which vessels were involved is not known since angiography was not justified.

Headache-free patients

In some patients headache is insignificant, and some other sensation in the head may predominate.
Five out of the 32 patients did not have vascular headache as shown in Table 3. Four of these 5 complained of muscle aches and stiffness with associated malaise. The average ESR was 59 mm/hr, the range being 13 to 105.

**Table 3.** Headache-free patients

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age (years)</th>
<th>Symptom</th>
<th>ESR (mm/hr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>75</td>
<td>No complaint of</td>
<td>35</td>
</tr>
<tr>
<td>Female</td>
<td>69</td>
<td>headache</td>
<td>105</td>
</tr>
<tr>
<td>Male</td>
<td>72</td>
<td></td>
<td>50</td>
</tr>
<tr>
<td>Male</td>
<td>60</td>
<td>'Tension headache'</td>
<td>92</td>
</tr>
<tr>
<td>Female</td>
<td>76</td>
<td>Burning round eyes</td>
<td>13</td>
</tr>
</tbody>
</table>

**Case 3.** A 72-year-old retired teacher presented with a 3-month history of ‘touchiness’ of the scalp and temples. He had never experienced pain, and he slept well. For 3 days he noticed swelling and redness over his left temple but there was no tenderness. He had no malaise or myalgia. On examination his temporal arteries were thickened, discoloured but pulsatile, otherwise nothing abnormal was found. Investigations: Hb 13-9 g/dl, WCC 7.5 × 10⁹/l; ESR 47 mm/hr. He was commenced on 60 mg of prednisolone/day and a temporal artery biopsy was positive. The ‘touchiness’ resolved with steroids with were gradually reduced.

**Case 4.** A 60-year-old warehouseman complained of headache in the occipital region, vertex and forehead, this was of 3 months’ duration and worse in the morning. His neck had felt stiff and he complained of horizontal double vision for one week. Examination was normal. Investigations: X-rays of chest and skull, ECG, isotope brain scan and lumbar puncture were all normal. A diagnosis of tension headache was made. He returned 2 years later complaining that his head felt too heavy for his neck and that it was as if he had a knot in the back of his neck; this was relieved by simple analgesics. He was given diazepam. Two months later his headache had settled but he complained of fatigue and anxiety. His ESR was 125 mm/hr. He was admitted to Hull Royal Infirmary and a temporal artery biopsy was positive. Prednisolone 40 mg/day was given with relief of his headache. Four years later he was well, steroids had been tailed off and the ESR was normal.

The initial phase was of tension headache. The late development of diplopia suggested a vertebro-basilar lesion and the high ESR and biopsy confirmed arteritis.

**Normal ESR**

It should be remembered that in the elderly, an ESR of up to 30 mm/hr can be considered normal. In this series, 5 patients out of the 32 presented with a ‘normal’ ESR. Four of these satisfied the criteria given above although they had a negative temporal biopsy. One patient presented with an ESR of 9 mm/hr on the same day as a positive temporal artery biopsy. It is of interest that her ESR the following day was 82 mm/hr.

**Discussion**

This study correlates with the typical findings of temporal arteritis for patients over the age of 60 years (mean 69 years) with a raised ESR (mean 71 mm/hr). There are reports of temporal arteritis with a normal ESR (Kansu *et al*., 1977), and this is described in this paper. It is important to remember that a single normal ESR does not exclude the condition and that in an elderly person with doubtful symptoms, possibly of temporal arteritis, at least 3 ESRS should be taken and a temporal artery biopsy performed.

Temporal arteritis is essentially an inflammatory condition affecting a number of different vessels; this accounts for the variety of its manifestations. The most commonly involved vessels are superficial temporal, vertebral, ophthalmic and posterior ciliary arteries, which is reflected in the common presentations of scalp tenderness and blindness (Huston *et al*., 1978). The fact that any vessel may be involved in the disease process is seen in the clinical variants described in the literature (Huston *et al*., 1978; Healey and Wilske, 1977; Swinson, Goodwill and Talbot, 1976; Medical Practice, 1979; Wilkinson and Ross-Russell, 1972).

Giant cell arteritis involving the intracranial vessels is described in cases 2 and 4, the first was manifest as a psychotitic state, this being only one of the well recognized mental changes (Medical Practice, 1979). In this study, 3 patients out of 32 (10%) presented with mental changes but Ross-Russell found that 7 of 35 patients (20%) with temporal arteritis were depressed and 4 of 35 (11%) presented with confusion (Ross-Russell, 1959). One can understand how malaise and a very severe headache may cause misery but psychotitic changes are probably caused by ischaemia of the brain due to cerebral or extracranial vessel involvement. Mental disturbances of affect due to the giant-cell process may render the headache insignificant by changing the pain threshold.

The prodromal state which overlaps polymyalgia rheumatica is of importance. Biopsy specimens from the temporal arteries in both conditions have demonstrated the same giant-cell infiltration (Harri-son and Bevan, 1967). Evidence of arteritis in the temporal arteries was demonstrated in 16 of 21 biopsies in patients presenting with polymyalgia rheumatica (Hamrin, Jonsson and Landberg, 1964).
Case 1 shows the interrelationship between cranial arteritis and polymyalgia rheumatica.

Temporal arteritis is a well known cause of pyrexia, temperatures of up to 39°C accompanied by night sweats have been reported in the literature (Healey, 1977; Huston et al., 1978; Healey et al., 1977). The incidence of pyrexia varied from 29 of 35 patients (Healey, 1977) to 5 of 50 patients (Healey and Wilskey, 1977) and 2 of 42 patients (Huston et al., 1978). Of the 7 patients (21%) in this study, presenting with fever, all complained of sweats, which in 6 of them occurred at night. Six of the 7 complained of symptoms of polymyalgia rheumatica, i.e. malaise, muscle pain and stiffness particularly affecting the shoulder girdle. It is interesting that not one of the patients with fever complained of headache. The ESR for this group of patients appears significantly higher than that of the whole group, i.e. 103 as compared with 71 although the WCC (mean 9.2 x 10⁹/l) was normal. It appears from this study that fever in temporal arteritis is associated with a higher than average ESR polymyalgia rheumatica.

The headache of temporal arteritis described in the text-books is severe and persistent, prevents sleep and is associated with scalp tenderness. Several other types of headache are described including a generalized ache associated with pain in the neck (Healey and Wilskey, 1977), spasms of shooting pain (Wilkinson and Ross-Russell, 1972) and a constant occipital headache (Medical Practice, 1979). It should be remembered that headache may be a minor complaint or, as seen in cases 1 and 3, patients may deny headache. In this study 4 of the 5 patients with atypical cranial sensations complained of muscle aches and stiffness associated with malaise. This indicates that polymyalgia rheumatica is a very helpful symptom in diagnosis of temporal arteritis. The average ESR of these 5 patients was 59 mm/hr as compared to 71 mm/hr for the whole group. It is interesting to speculate that those patients with atypical cranial sensations have a milder form of the condition as indicated by a lower than average ESR.

Looking at the ESRs of the patients in this study there appear to be 3 groups:

(i) a slight raised ESR (<50 mm/hr) which is associated with a long prodromal illness preceding an acute episode of classical temporal arteritis with a sudden rise in the ESR;

(ii) a moderately raised ESR (<70 mm/hr) with a mild form in which headache is an insignificant complaint;

(iii) a very high ESR (up to 100 mm/hr) with a severe illness associated with fever, systemic upset and polymyalgia rheumatica.

Because corticosteroids greatly reduce the risk of loss of vision associated with temporal arteritis, early diagnosis is of importance. There is little difficulty in the classical case but this study shows that the condition may occur in the absence of headache. Variants such as polymyalgia rheumatica, fever, ophthalmoplegia and psychiatric states are not uncommon and, if recognized, blindness and other arterial occlusions may be prevented.

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