

# Temporal arteritis in a large necropsy series

GÖREL ÖSTBERG

From the University Department of Pathology, Malmö General Hospital, S 214 01 Malmö, Sweden

Temporal arteritis is a disease of old age, often manifesting itself clinically with rather vague symptoms of fatigue, general malaise, and weight loss. The characteristic headache or symptoms from the temporal regions may appear late (Harrison and Bevan, 1967) or may pass unobserved. The first report of a case was given by Hutchinson (1890) in London, and series of varying numbers have since been published from different parts of the world. Most have been collected in more or less specialized clinics and there has been no correlation with any defined population. The number of reports shows that the condition is not rare.

The present investigation establishes the prevalence of temporal arteritis in a necropsy series from a defined population, namely the city of Malmö in Southern Sweden. Discussion with Dr. Hamrin\* of Växjö, who is investigating poly-myalgia rheumatica and temporal arteritis in a county of southern Sweden, gave the impression that there may be differences between the urban and rural districts. As Växjö has no regular pathology service, contact was established with the pathology department in the nearby county of Kalmar.

## Materials

The present series included all adult subjects coming to necropsy in Malmö General Hospital (1,097 cases) between December 1, 1962, and November 30, 1963. The city of Malmö has about 250,000 inhabitants. There is only one general hospital, which includes a department for chronic diseases. About 65 per cent. of all deaths in the town occur in hospital and 98 to 99 per cent. of all patients who die there are subjected to *post mortem* examination. The age distribution of the Malmö series is given in Fig. 1. The frequency of necropsies in the town population is somewhat higher for young subjects and lower for older subjects (Berge, 1967).

Cases were also collected from the Kalmar hospital between January 1 and December 31, 1963. The com-

position of the series of 230 cases from Kalmar was less well-defined; this hospital receives patients from a mainly rural district of about 155,000 inhabitants, and fewer *post mortem* examinations are performed there.

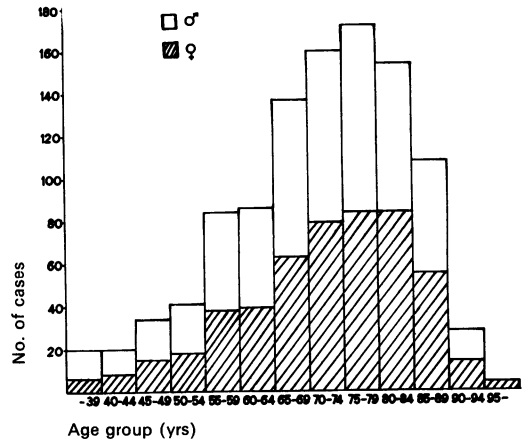


FIG. 1 Age distribution of necropsy material at Malmö General Hospital.

## Methods

About 1 cm. of the temporal artery on each side was removed with scissors from the inner side of the scalp at the time of removal of the vault of the skull by a necropsy attendant. The specimens were fixed in 10 per cent. neutral formalin, cut into three transverse planes, and embedded in paraffin. Sections were stained with haematoxylin and eosin and by the van Gieson method for screening. Arteries with significant changes were then stained for elastic tissue. Owing to the method of collection some specimens contained only subcutaneous tissue and muscle.

In 763 cases (70 per cent.) of the Malmö series specimens of two arteries were obtained, and in 280 (26 per cent.) only one. The corresponding figures for the Kalmar series were 167 (73 per cent.) and 43 (19 per cent.). The 53 (5 per cent.) Malmö cases and twenty (9 per cent.) Kalmar cases in which no arteries could be recognized were not analysed further. A total of 2,183 arteries was examined. The histological preparations did not always

provide three transverse sections. Sometimes only one section was available, and the length of the piece of the artery examined thus varied from the thickness of one microscopic section to about 1 cm. The specimens were not serially sectioned. Atherosclerotic lesions, consisting of fibrous intimal plaques with or without lipid deposits or calcification of the elastica, were not counted. All specimens with marked intimal fibrosis, or pronounced shrinkage of the vessel with irregular folds and reduction of the lumen, were set aside for further analysis. These were stained for elastic tissue and additional sections were made from the blocks. In sixteen cases (twelve from Malmö and four from Kalmar) the vessels showed distinct changes, and in these cases the clinical records and other necropsy findings were studied in more detail. The case histories are presented below and the findings are summarized in the Table (overleaf).

### Case histories

**Case 1, a retired fireman born in 1886,** had Spanish influenza in 1917, renal stone in 1932, varices, symptoms of gallstone, and inguinal hernia in 1955, myocardial infarction in 1959.

He was admitted to the department of internal medicine in March, 1961, because of pain in the right temporal region, with gradually increasing headache on the left side also and, during the last 2 weeks, impairment of vision.

#### Examination

He was in a good general condition with tenderness of both temples, where the vessels were palpated as hard cords. Erythrocyte sedimentation rate 65-73-67-33-38 mm./hr. Biopsy of the temporal artery showed distinct changes of arteritis (Fig. 2). The condition responded favourably to treatment with prednisone. In January, 1963, the patient returned with manifest pulmonary oedema and symptoms of acute myocardial infarction, and died the following day.

### Necropsy

There was severe aortic atheromatosis with a small mural thrombus in the abdominal aorta. Prominent coronary sclerosis with an occluding thrombus in the anterior, descending branch and extensive, fresh infarction in the anterior wall and septum of the left ventricle.

#### Microscopic examination

There was an irregular fibrous thickening of the intima of the aorta, subclavian, and carotid arteries, with thin capillaries in the media, surrounded by clusters of round cells. Sections of the aorta contained multinucleate giant cells around degenerating elastic lamellae. In this case, then, there was probably generalized arteritis with relatively active changes in the large vessels.

#### SUMMARY

Histologically verified temporal arteritis 2 years before death, treated with corticosteroids. Died from acute myocardial infarction. Necropsy showed inflammation of aorta, and subclavian and carotid arteries. Temporal artery distinctly changed (Fig. 3, overleaf).

**Case 2, a housewife born in 1890,** had felt well apart from recurrent urinary cystitis in the 1940s, and acute pyelitis in 1950. Abdominal symptoms in 1962 led to the diagnosis of carcinoma of the sigmoid colon, which was treated surgically in February, 1963. The operation was immediately followed by right-sided hemiparesis and the patient died a few days later, apparently from a vascular insult.

### Necropsy

There were changes interpreted as atheromatous lesions in the aorta with partly coalescent plaques. Immediately above the ostium was a large, 1 cm. mural thrombus that had resulted in thromboembolism of the coronary, splenic, renal, and middle cerebral arteries with infarction.

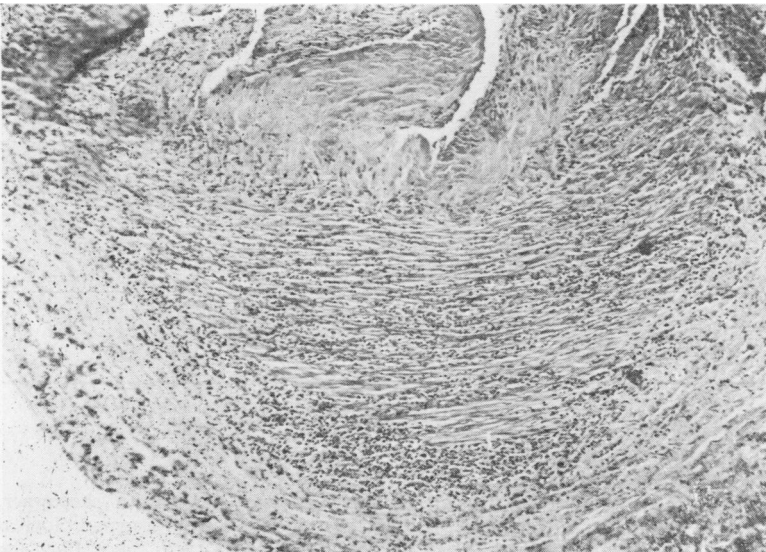


FIG. 2 Case 1. Biopsy specimen, 1961. Slightly tangentially cut artery with thickened intima above. Fibrinoid necrosis of internal elastic lamina. Greatly thickened media with inflammatory cells. No giant cells. Diagnosis: Temporal arteritis. Haematoxylin and eosin  $\times 75$

**Table** *Data on previous illness, main disease at necropsy, and findings in temporal arteries in sixteen cases.*

Case no.	Sex	Age (yrs)	Relevant history	Main disease	Pathological finding			
					Narrowing		Intimal fibrosis	
1	M	77	+1961 Biopsy +	Myocardial infarct	L +	+	+ +	+ +
					R —	No	artery	
2	F	73	—	Aortic thrombosis with embolism Cerebral softening	L —			—
					R —	No	artery	
3	F	82	—	Myocarditis	L —			—
					R —			—
4	M	77	+1961 Biopsy +	Carcinoma of caecum	L +	+ +	+ +	+ +
					R —	No	artery	
5	F	73	+1955	Cerebral softening	L —			+ +
					R —			+ +
6	M	88	+ ?1949	Cirrhosis of liver Pneumonia	L —			—
					R —			—
7	F	92	—	Myocardial infarct Carcinoma of stomach	L —	No	artery	—
					R —			—
8	F	76	+ ?1958	Myocardial infarct	L —			—
					R —			—
9	F	74	+ ?1959	Myocardial infarct	L —			—
					R —			—
10	F	87	—	Cerebral softening	L —			+ +
					R —			+ +
11	F	85	+ ?1955	Aortic stenosis and incompetence	L —			+ +
					R +			+ +
12	F	83	?SR 130 mm./hr 6/63	Congestive heart failure	L —			—
					R —			—
K1	M	74	+ ?1962	Myocardial infarct Carcinoma of bladder	L —			—
					R —			+ +
K2	F	68	—	Carcinoma of breast	L —			+ +
					R —			
K3	F	82	?2 yrs	Bronchopneumonia	L —			+ +
					R —			—
K4	M	82	—	Haematemesis Duodenal ulcer	L —			+ +
					R —			+ +

*Microscopic examination*

Arteritis with destruction of the elastica and abundant lymphocytes and some giant cells, especially in the ascending aorta. Less distinct changes were seen in the distal aorta and iliac artery.

## SUMMARY

No previously known disease. Aortic arteritis and

changes in temporal artery found at necropsy.

**Case 3, an unmarried woman, formerly a shopowner, born in 1881,** had felt well, apart from a gynaecological operation of unknown nature in 1916. Since 1949 she had had spells of dizziness and fatigue, and had been

Vascularization		Fibrosis of adventitia		Round cells in adventitia		Comments
Media	Adventitia					
+	+	+	+	+	+	Biopsy + 1961 Arteritis (aorta and subclavian and carotid arteries)
+	+	+	+	+	+	Arteritis (aorta and iliac artery)
—		+	+	+	+	Atherosclerosis No microscopy
+		+		+		Biopsy + 1961 Atherosclerosis No microscopy
—		+		+		Arteritis (aorta and carotid artery)
+		+	+	+		Atherosclerosis No microscopy
—		+	+	+		Atherosclerosis No microscopy
—		+	+	+		Atherosclerosis No microscopy
+		+	+	+	+	Atherosclerosis No microscopy
—		+		+		Atherosclerosis No microscopy
—		+	+	+		Atherosclerosis No microscopy
+		+	+	+	+	Atherosclerosis No microscopy
—		+		+		Atherosclerosis No microscopy
+		+	+	+		Atherosclerosis
+		+	+	+	+	No microscopy
+		+	+	+		Atherosclerosis
+		+	+	+	+	No microscopy
—		+		+	—	Atherosclerosis No microscopy
—		+		+	+	Moderate atherosclerosis No microscopy
—		+		+	+	Aorta widened with thin wall; thrombosis No microscopy
—		+	+	+		Dilated aortic arch; atherosclerosis No microscopy
—		+		+	—	

treated for high blood pressure. In the autumn of 1962 she had shortness of breath and occasionally palpitations. She had no chest-pain, headache, or nausea.

She was admitted to the department of internal medicine in 1963, after an attack of irregular cardiac activity. The electrocardiogram showed rapid fibrillation and left bundle branch block. She died some hours later before any laboratory studies could be performed.

#### Necropsy

There was enlargement of the left side of the heart with diffuse, non-specific myocarditis. Signs of cardiac failure were present with general stasis and oedema of the lower legs. The aorta showed moderate atheromatosis, as did the coronary arteries. Fairly marked atherosclerotic changes in the basal cerebral arteries. The vessels were not examined histologically.

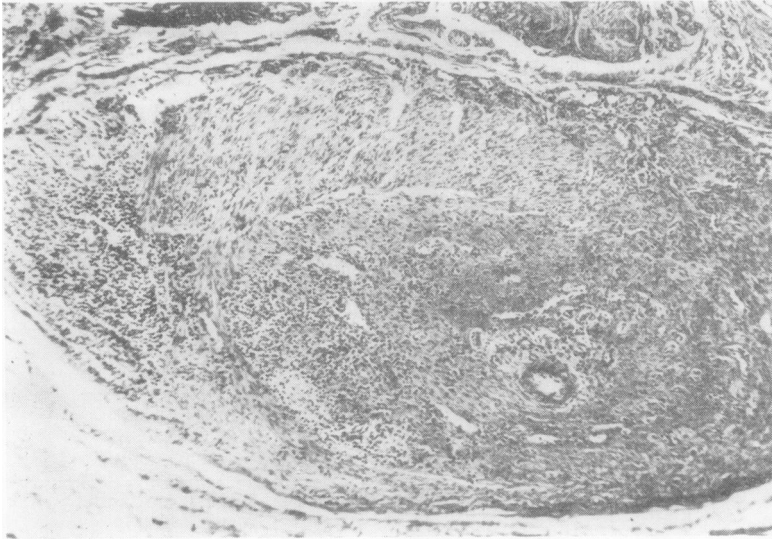


FIG. 3 *Case 1. Necropsy specimen. Thin artery with narrowed lumen. Fibrous thickening of all layers of the wall with many thin capillaries and lymphocytic infiltration. Haematoxylin and eosin  $\times 75$*

**SUMMARY**

No known previous disease. Acute cardiac failure. Non-specific diffuse and relatively mild myocarditis. Changes in temporal artery.

**Case 4, a labourer born in 1886, who retired in 1953. He had previous symptoms of duodenal ulcer.**

He was admitted to the department of internal medicine in 1961 for investigation of increasing headache of 1 month's duration. The headache was localized to the forehead, temple, and back of the head. Vision had also become somewhat impaired. Mild reduction of body weight had resulted from loss of appetite.

**Examination**

The temporal arteries were found to be tender and erythrocyte sedimentation rate 105 mm./hr. Biopsy showed active arteritis (Fig. 4). The patient re-

sponded well to phenylbutazone but soon afterwards double vision supervened and prednisone was substituted. His vision again improved and the erythrocyte sedimentation rate in November was 60 mm./hr. Medication was withdrawn in December. In June, 1962, the erythrocyte sedimentation rate was 90 mm./hr. He had by then ulnar paresis with a certain degree of muscular atrophy of the arms, which was interpreted as cervical spondylosis. This improved after radiotherapy. Abdominal symptoms appeared at the end of 1962, and a large tumour was palpated in the abdomen. The patient died in April, 1963.

**Necropsy**

There was a mucus-forming cancer in the abdominal cavity, probably originating from the appendix. Moderate aortic atheromatosis, mild coronary sclerosis, and moderate sclerosis of the basal cerebral arteries with

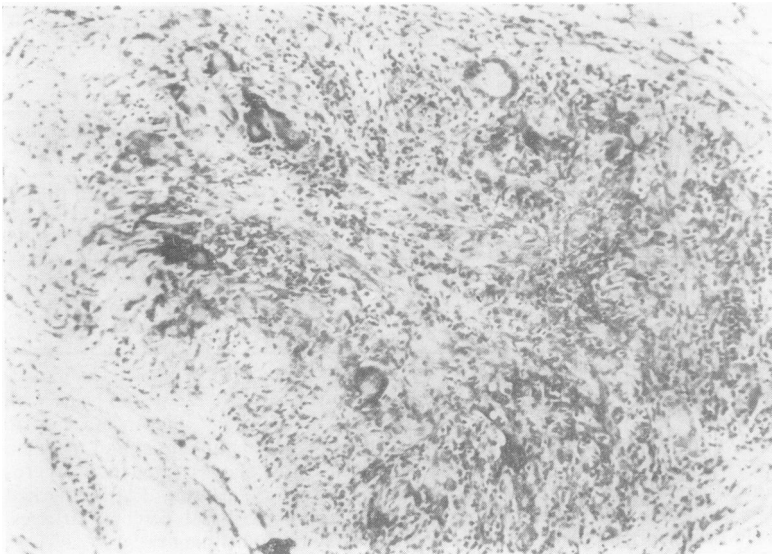


FIG. 4 *Case 4. Biopsy specimen, 1961. Obliterated artery with destroyed wall, many giant cells and lymphocytes. Haematoxylin and eosin  $\times 120$*

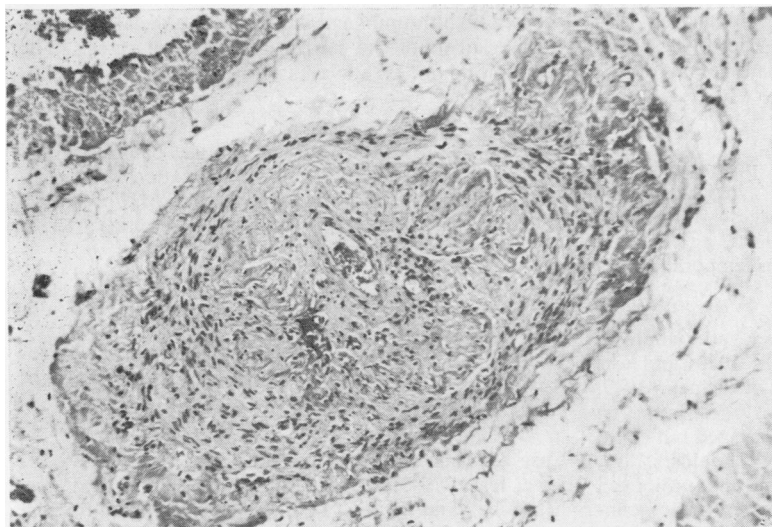


FIG. 5 Case 4. Necropsy specimen. pronounced fibrosis of all layers of the wall, narrow lumen, scanty round cell infiltration. Haematoxylin and eosin  $\times 120$

encephalomalacia of the cerebellum. The great vessels were not examined histologically.

**SUMMARY**

Histologically verified temporal arteritis with disturbances of vision 2 years before death. Responded favourably to corticosteroid treatment. Died from abdominal carcinoma. Temporal artery thin and shrunken with fibrotic wall (Fig. 5).

**Case 5, a housewife born in 1890,** had had headache with dizziness and impairment of vision on the left side in 1947; these were interpreted as signs of hypertension.

In 1955 she was admitted to hospital because of joint disease and a high erythrocyte sedimentation rate. She had then had 3 months' pain in the shoulder joint, and mild pain in the knees and hips. The erythrocyte sedimentation rate was 40–62 mm./hr. The temporal arteries were felt as hard cords and were tender to palpation.

Phenylbutazone gave relief for a few months. The erythrocyte sedimentation rate was then 74–102 mm./hr.

In 1960 she had herpes zoster in the left axillary region. She was re-admitted to hospital in May, 1963, because of weakness and dizziness which had been developing since half a year. After 17 days in hospital she had respiratory difficulty, terminally ventricular fibrillation, and pulmonary oedema.

*Necropsy*

There was mild, diffuse cerebral atrophy and small, old, and recent areas of encephalomalacia. The proximal part of the aorta was widened and had a thin wall with barely discernible transverse folds, and, distally, increasing calcifications of intimal plaques.

*Microscopic examination*

The thoracic aorta showed rarefaction and splitting of the elastica with capillaries, surrounded by lymphocytes and some polymorphs. No giant cells. Sections of the

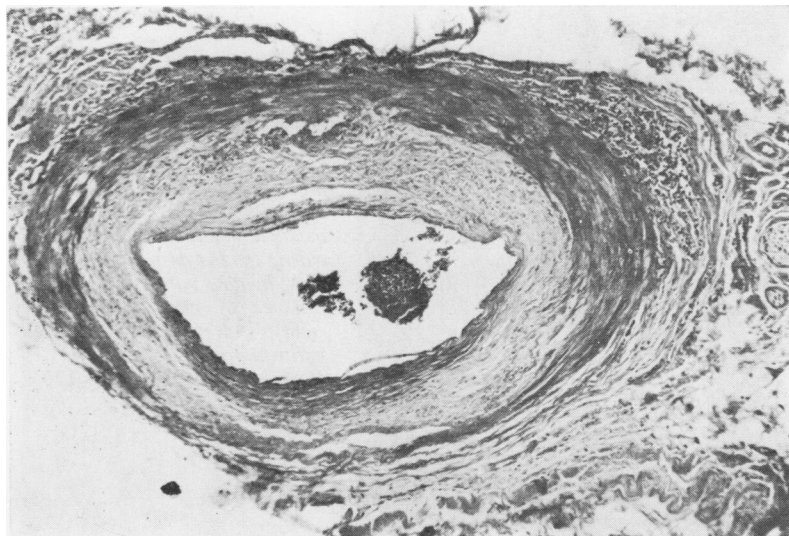


FIG. 6 Case 5. Necropsy specimen. Rather wide vessel with slightly thickened media. Adventitial fibrosis with marked round cell infiltrations. Haematoxylin and eosin  $\times 75$

left common carotid artery showed dilatation and similar rarefaction of the elastica. The right artery appeared normal. There was no inflammation of the femoral arteries.

#### SUMMARY

Illness 8 years before death possibly polymyalgia arteritica, but not diagnosed. Diffuse symptoms some months before death. Vascular changes with microscopically verified inflammation of aorta and carotid artery at necropsy. Mild changes in temporal artery (Fig. 6).

**Case 6, an engineer born in 1875,** had had symptoms of gastric ulcer since the middle of the 1940s and was admitted to the department of internal medicine in 1949. He was anaemic and had a high erythrocyte sedimentation rate of 112 mm./hr; malignancy was suspected but could not be proved. He had also had pain in the shoulders, hips, and knees, and complained of intense and prolonged headache. Temporal arteritis was apparently not considered as a possible diagnosis. A stone was found in the urinary bladder. In 1953 he again had symptoms of ulcer with gastric retention. Urinary retention in 1953-54 led to transvesical prostatectomy with extraction of the bladder stone in May, 1954. The same year he was submitted to gastric resection because of haematemesis. The operative specimen revealed no signs of a malignant disease. The erythrocyte sedimentation rate on that occasion was 40-80 mm./hr. During the following years he gradually deteriorated; in 1962 shortness of breath and swelling of the legs supervened and in the Spring of 1963 he had jaundice and ascites. He died in June, 1963.

#### Necropsy

There was acute and chronic cholangiohepatitis with signs of cirrhosis and cardiac failure with pulmonary oedema and general stasis. Pronounced aortic atheromatosis with partly coalescent, largely-eroded plaques

covered with thrombi and increasing in a distal direction. Mild sclerosis of the cardiac and cerebral vessels. The vessels were not examined histologically.

#### SUMMARY

14 years before death headache, muscle pain, and high erythrocyte sedimentation rate. Investigation showed no malignant disease. Not treated with steroids. Progressive deterioration. Necropsy revealed no general arteritis but changes in temporal arteries.

**Case 7, a woman born in 1871,** became suddenly ill, and died soon after arrival in hospital. No known history of earlier disease. Macroscopical severe atherosclerosis but no signs of generalized arteritis. Slight fibrosis of temporal artery.

**Case 8, a housewife born in 1887,** had pain of the left lower leg in 1943, and in 1949 'neurosis' with dizziness, headache, and unrest. In 1958 she had pain of the muscles of the right upper arm, later pain in the arms and legs, and stiffness and difficulty in lifting the arms. Erythrocyte sedimentation rate 53-57 mm./hr. The symptoms were interpreted as cervical spondylosis and myalgia. The condition responded well to sodium salicylate and meprobamate.

She was admitted to hospital on October 1, 1963, with severe chest pain. Electrocardiogram showed signs of anterior infarction. She deteriorated and died 5 days later.

#### Necropsy

There was a thrombus in the left descending coronary branch with extensive infarction of the anterior wall and septum of the left ventricle and fibrinous pericarditis. In the aorta and great arteries there was pronounced atheromatosis with yellow-white, raised plaques, increasing distally and there with calcifications. The great vessels were not examined microscopically.

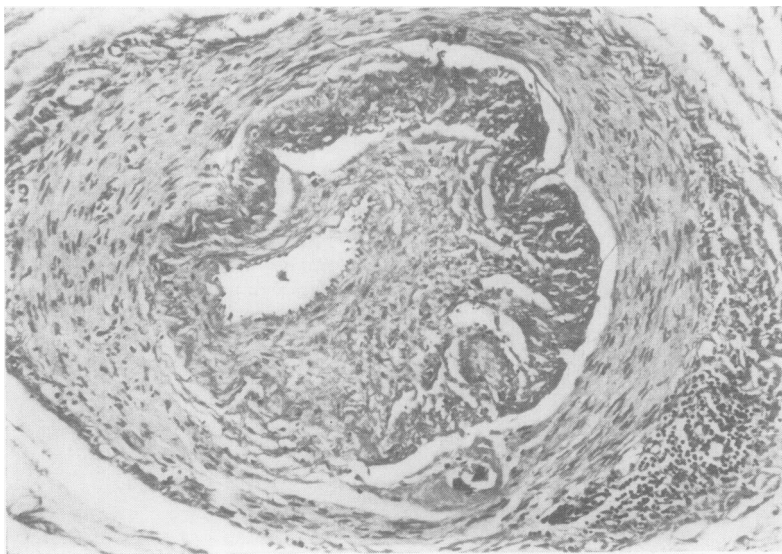


FIG. 7 *Case 11. Necropsy specimen, Narrow lumen with marked intimal fibrosis. Splitting of media probably artefact. Internal elastic lamina destroyed in lower part, hyaline patch with possibly residues of giant cells (lower right). Adventitial round cell infiltrations. Haematoxylin and eosin  $\times 120$*



**SUMMARY**

Suspected disease (polymyalgia rheumatica?) with muscle pain and high erythrocyte sedimentation rate 5 years before death. Necropsy revealed chronic inflammation of temporal arteries.

**Case 9, a housewife born in 1889**, had diabetes diagnosed in 1946. Her mother, two sisters, and two brothers also had diabetes. The disease could not be controlled by dietary restriction, and insulin treatment was started in 1947.

In 1959 she was admitted to hospital. She had by then had a few episodes of loss of consciousness and some weakness of the left side, which was interpreted as the effect of cerebral ischaemia. She complained of pain in the muscles of the limbs and joints for 1 year and loss of weight during the last few months. Blood sugar values were normal, and the erythrocyte sedimentation rate 57–84 mm./hr. Judging from the patient's records arteritis was not suspected and she received no special treatment. In October, 1963, she was admitted to hospital with signs of infarction and died after a few days in a state of shock. Erythrocyte sedimentation rate 12–16 mm./hr.

**Necropsy**

There was a large, fresh infarction of the left ventricle, signs of cardiac failure with stasis, and pulmonary oedema. Pronounced aortic atheromatosis, increasing distally. The great vessels were not examined histologically.

**SUMMARY**

Diabetes mellitus since age 60. Muscle pain, weight loss, and raised erythrocyte sedimentation rate 4 years before death. Died with myocardial infarction. Necropsy changes in temporal arteries.

**Case 10, an unmarried woman born in 1876**, had previously felt well until 1961, when cardiac decompensation was diagnosed. The condition responded well to digitalis and diuretics.

She suffered a sudden left-sided central facial paresis and weakness of left arm in 1963, and died 3 days later from pulmonary oedema. The erythrocyte sedimentation rate on admission was 10 mm./hr.

**Necropsy**

There was a thrombus in a branch of the right medial cerebral artery with recent encephalomalacia in the posterior part of the right frontal lobe. Generalized arteriosclerosis with very advanced atheromatosis of the aorta with calcified plaques, distally coalescent. The great vessels were not examined microscopically.

**SUMMARY**

No known previous disease. No macroscopic signs of arteritis in great vessels but microscopical changes in temporal arteries.

**Case 11, an unmarried woman born in 1878**, had been admitted to the department of infectious diseases in 1955 because of fever and several weeks' weakness and

malaise. Erythrocyte sedimentation rate 129 mm./hr., blood pressure 200/100, and osteoporosis was diagnosed.

In 1962 she was admitted to the department of internal diseases because of cardiac decompensation, which improved on treatment with digitalis and diuretics. The erythrocyte sedimentation rate was 82–59 mm./hr.

She was re-admitted in November, 1963, in a poor general condition with cardiac decompensation, and died a few days later.

**Necropsy**

There was stenosis and insufficiency of the aortic valves with calcification. Coronary sclerosis with healed infarction of the posterior wall. Signs of cardiac insufficiency with pulmonary oedema. The aorta showed advanced atheromatosis with erosions and calcifications, increasing distally. The great vessels were not examined histologically.

**SUMMARY**

8 years before death treated for fever and raised erythrocyte sedimentation rate. Judging from hospital records, arteritis not considered. Death from acute cardiac failure. Temporal artery narrowed by fibrosis (Fig. 7).

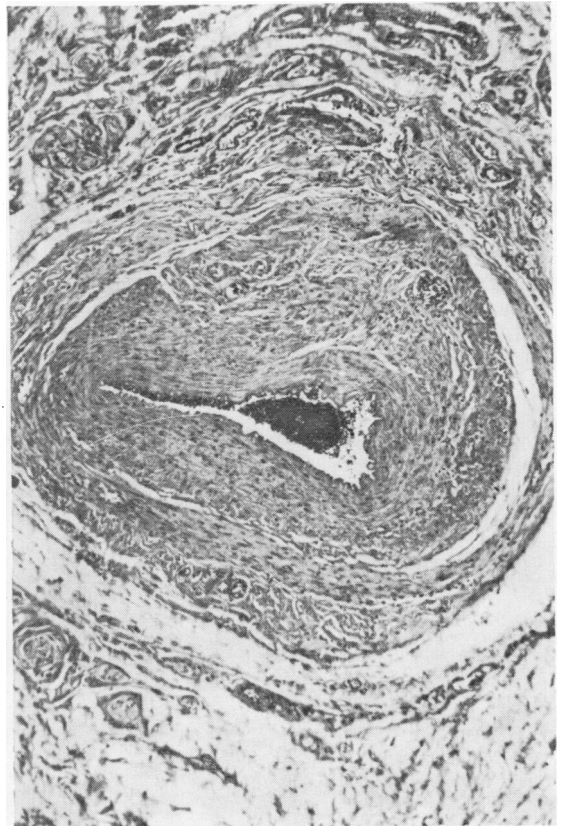


FIG. 8 Case 12. Necropsy specimen. Narrow vessel with fibrosis of media and adventitia with penetrating capillaries and diffuse, slight round cell infiltration. Haematoxylin and eosin  $\times 75$



**Case 12, a housewife born in 1880**, had had chronic leg ulcers since she was 40 years of age. In 1960 she had anaemia, probably after melaena, but the cause was not investigated. After 1962 she had repeated spells of jaundice without other symptoms, except loss of body weight.

In June, 1963, she had a severe attack of fever and jaundice, for which she was admitted to hospital. The liver was enlarged. The erythrocyte sedimentation rate was 118–122 mm./hr. Radiological examination revealed no stones but a widening of the common bile duct. The patient was sent home in a good condition 9 days later, but she returned in November, 1963, and died on the day of admission.

#### *Necropsy*

There were signs of hypertension with hypertrophy of the left heart. Severe constrictive coronary sclerosis with small, recent infarction of the left ventricle, signs of cardiac insufficiency with pulmonary oedema and congestion. Cholecystolithiasis, widening of common bile duct, mild liver cirrhosis. Very severe atheromatosis of the aorta with coalescent calcifications in the abdominal portion. The great vessels were not examined histologically.

#### SUMMARY

No evidence of previous arteritis. Temporal artery (Fig. 8) narrowed by adventitial fibrosis. Round cell infiltration.

**Kalmar Case 1, a tax collector born in 1889**, was operated upon in 1938 and treated with radiation for cancer of penis. He had back pain, especially in the lumbar region, with stiffness of shoulders since February, 1962. Radiological examination revealed osteoporosis with compression of L2, L4, and L5. Erythrocyte sedimentation rate 100 mm./hr. No signs of myeloma. Proteinuria and anaemia. Nonprotein nitrogen 40 mg./100 ml. Prostatic hyperplasia.

In August, 1962, he had haematuria and stones were

demonstrated in the urinary bladder. In January, 1963, he had acute chest pain and an electrocardiogram suggested infarction of the anterior cardiac wall. He died some days later.

#### *Necropsy*

There was generalized arteriosclerosis with thrombi in the anterior descending coronary branch and recent infarction of the septum and anterior wall of the left ventricle with cardiac rupture and haemopericardium. Cancer of the urinary bladder with perforation of the bladder wall, and engagement of adherent loop of ileum, left-sided ureteritis and pyonephrosis. Osteoporosis. No great vessels removed for microscopical examination.

#### SUMMARY

Muscle pain and high erythrocyte sedimentation rate one year before death. Temporal artery showed mild shrinkage and fibrosis.

**Kalmar Case 2, a housewife born in 1895**, had been healthy until in 1961 she had noticed a growing tumour in the left breast which was diagnosed as a cancer. She had increasing shortness of breath and pleural exudate. The tumour was inoperable, and she was given cyclophosphamide. The erythrocyte sedimentation rate rose to 162 mm./hr. She died in March, 1963.

#### *Necropsy*

There was widespread, left-sided mammary cancer with a large ulcer of the skin and an extensive growth in the thoracic wall and in several organs. The abdominal aorta showed moderate atheromatosis. No vessels were removed for microscopical examination.

#### SUMMARY

Mammary cancer for 2 years. Clinical records contained no notes on arteritis but temporal artery showed mild changes.

**Kalmar Case 3, a woman born in 1881**, lived in the country and records were scanty. In the last 2 years she had been

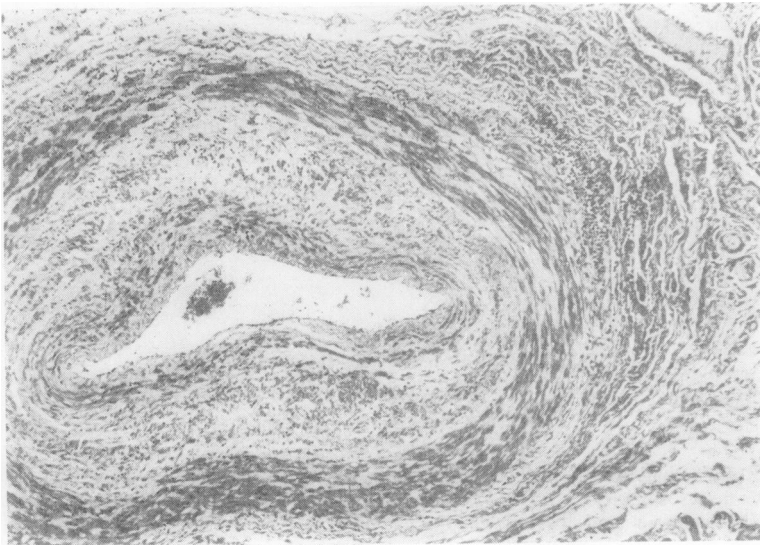


FIG. 9 *Kalmar 3. Necropsy specimen. Marked adventitial fibrosis with capillary proliferation and round cell infiltration. Slightly thickened media and intima. Haematoxylin and eosin  $\times 75$*

treated for heart decompensation and senile confusion.

In April, 1963, she was admitted to the department of infectious diseases with fever and cough, and was found to be in a poor general condition with mental confusion. Bronchopneumonia, heart decompensation, and cerebral arteriosclerosis were diagnosed. Erythrocyte sedimentation rate 65 mm./hr. She died after 4 days.

#### Necropsy

There was purulent bronchitis and widespread focal pneumonia. Above the ostium and along its entire length the aorta was markedly widened but with no distinct aneurysm. In the distal part of the abdominal aorta and above the left renal artery were parietal thrombi. The left renal artery was filled with thrombi with infarction of the kidney. A renal carcinoma twice the size of a walnut was found in the left kidney. No vessels were removed for histological examination.

#### SUMMARY

Scanty data. Poor general condition for last 2 years. Marked widening of aorta suggested arteritis. Temporal arteries showed chronic inflammatory changes (Fig. 9).

**Kalmar Case 4, a retired farmer born in 1881**, was healthy until he was admitted to hospital in July, 1963, after 3 weeks' fatigue and a few days with abdominal pain and vomiting. A blood transfusion was given but he died after 3 days.

#### Necropsy

There was duodenal ulceration and erosions in the cardia with intestinal bleeding. General arteriosclerosis with healed myocardial infarction. The ostium of the aorta was narrowed and calcified. The aortic arch and thoracic portion were markedly dilated with multiple calcified plaques, and similar severe changes were found in the abdominal aorta. No mural thrombi. Atheromatosis was also seen in the pulmonary artery. No great vessels were removed for histological examination.

#### SUMMARY

No known previous disease. Widening of thoracic aorta suggested aortitis. Fibrosis of temporal arteries.

#### Results

Active temporal arteritis was not found in any case, but the sixteen patients (5 males and 11 females) listed above had alterations, which were interpreted as postarteritic (Table). The mean age of the patients was 78 years (range 68 to 92).

In only two cases were there positive biopsies of the temporal artery 2 years before death. Nine patients had had diffuse symptoms with muscle pain, headache, and raised erythrocyte sedimentation rate 1 to 14 years before death; these symptoms could be interpreted as due to temporal arteritis. In five cases the records contained no data about previous diseases and the patients (*e.g.* Cases 7 and

K4) were not seen in hospital before their final disease.

One of the cases with verified arteritis and two with a suspicious previous medical history also showed histological changes in other vessels consistent with arteritis. In two cases the aorta was markedly dilated and thin-walled, but was not examined microscopically.

During the last 10 years there have been on average eleven temporal artery biopsies per year in Malmö, of which five showed histological arteritis and six did not.

As biopsies were made in practically all suspected cases, the incidence of clinically diagnosed temporal arteritis was about 2 in 100,000 annually.

The principal diagnoses at necropsy of the sixteen cases showed the same range of diseases as the necropsy series in Malmö as a whole. Temporal arteritis was not a main or contributory cause of death.

#### Discussion

Temporal arteritis is a disease of old age and it has even been suggested that the condition should be designated 'arteritis of the aged' (Paulley and Hughes, 1960). The present series confirms this general impression. The numbers are too small for any definite conclusion to be drawn from the marked sex difference (11 women; 5 men); the disease seems to be equally frequent in men and women, but in some other series females have been more frequent (Dixon, Beardwell, Kay, Wanka, and Wong, 1966).

The histological picture of active temporal arteritis is characteristic. The lumen is usually reduced and the intima thickened by fibrosis. An inflammatory cell infiltrate is present, mainly localized in the media and adventitia (Fig. 2). Round cells are always present, and characteristic giant cells may or may not be seen (Fig. 4). The internal elastic lamina is often destroyed and the giant cells seen in connection with elastic fragments. (Harrison, 1948; Kimmelstiel, Gilmour, and Hodges, 1952; Bevan, Dunnill, and Harrison, 1968). In the adventitia there may be fibrosis and a proliferation of capillaries; there is often cellular infiltration as in the media but usually no giant cells. The end stages of arteritis are not so well known. They were discussed by Kimmelstiel and others (1952), who stressed the role of the elastic changes in the active phase. They studied temporal arteries from a series of elderly patients and found elastic destruction. The difference between post-arteritic and senile lesions consisted in the absence of a giant cell and granulomatous reaction in the latter. Ainsworth, Gresham, and Balmforth (1961) also studied the relation of post-arteritic changes to degenerative changes

due to age and sometimes found it difficult to make a distinction.

In the present series atherosclerotic changes were not specially studied. The inner elastic lamina was usually well distinguished in sections stained with haematoxylin and eosin and was preserved. Calcification in the elastic lamina occurred without noticeable reaction. In connection with atheromatous plaques in the intima, the elastic lamina was sometimes thin or destroyed, but there was no pronounced medial or adventitial alteration. The characteristic lesions were intimal fibrosis with narrowing of the lumen and vascularization of the media (Figs 3, 5, and 7). These were seen more often in cases with diagnostic biopsies and/or highly suspicious clinical histories. Adventitial fibrosis and vascularization were not more pronounced in these cases than in those in which the diagnosis was 'uncertain'. Round cell infiltration may be more significant but was not a certain sign of the disease (Figs 6, 8, and 9).

The distribution of lesions in the temporal arteries was discussed histologically by Harrison (1948) and clinically (radiologically) by Gillanders, Strachan, and Blair, (1969). The distribution is segmental and the changes can vary in different sections from the same artery. In the present study no specific variation in the histological picture was characteristic, although narrowing of the lumen could be more pronounced in one of the three sections of the same artery. There was no consistent difference between the right artery and the left artery.

Positive histories were obtained in cases with positive biopsies in only two instances. The more questionable cases had vague histories, which is typical of the disease (Harrison, 1948; Paulley and Hughes, 1960; Harrison and Bevan, 1967; Nosenzo, Dughera, Macchioni, Judica-Cordiglia, and Gelato, 1967).

In one patient the duration of disease was 14 years, indicating the chronicity of the condition and the persistence of histological changes in the vessels.

That temporal arteritis is part of a generalized arterial disease has been stressed by many authors (Cooke, Cloake, Govan, and Colbeck, 1946; Harrison, 1948; Russell, 1959; Nosenzo and others, 1967). The present survey included only the temporal arteries, but in three cases, on clinical grounds or because of changes seen at necropsy, histological examination of the large vessels was made and arteritic changes were found. In two further cases the aorta was markedly dilated, but no microscopical study was made.

Temporal arteritis is not a rare disease. Its frequency, however, seems not to have been studied before. Ainsworth and others (1961) found two cases in 39 necropsies in patients over 60 years of age. Clinical material was mostly collected at

random. Hamrin and others (1965) related a series of 52 cases of polymyalgia arteritica seen during a period of 5 years, which they regarded as a manifestation of a generalized giant cell arteritis, in a population of about 100,000 inhabitants. This gave an incidence of about 10 per 100,000 persons per year.

Hamrin and his co-workers have taken a very active interest in the disease and cases from the whole district are referred to them. In Malmö, we found an incidence of about 2 per 100,000 persons per year, but all cases may not have been referred to hospital. Earlier Horton, Magath, and Brown (1934) and Cohen and Harrison (1948) noticed more cases in farmers and country dwellers, but Harrison (1948) mentioned that this did not seem to be the rule. Cohen and Harrison also noticed that information about occupational and home background was often not given. The difference between our figures from Växjö and Malmö might suggest a preponderance of the disease in rural populations, but figures from the present series are too small for valid conclusions to be drawn.

A clinical study made by Nosenzo and others (1967) in one hundred old persons revealed nine cases of temporal arteritis, a remarkably high incidence. The patients had had varying clinical symptoms, such as headache, psychic disorders, diffuse pain, and fever, all had anaemia and most of them a high erythrocyte sedimentation rate. Temporal artery biopsy was done in only some of the patients.

Dixon and others (1966) correlated temporal arteritis and polymyalgia rheumatica with other forms of rheumatic disease. They found the ratio between patients of all ages with rheumatoid arthritis and those with temporal arteritis or polymyalgia rheumatica to be 12 : 1; in patients aged 70 years or more rheumatoid arthritis was found twice as often as polymyalgia rheumatica and temporal arteritis. The frequency of rheumatoid arthritis has not been studied in the present series.

There was no correlation with any other special diseases and changes in temporal arteries had no apparent significance in the patients' symptomatology or in the cause of death. The prevalence in the necropsy series was higher than the clinical incidence and this suggests that many cases remain clinically undiagnosed.

### Summary

The town of Malmö has about 250,000 inhabitants. About 65 per cent. of all persons dying in the town are examined *post mortem* at Malmö general hospital. The temporal arteries from all adults (1,097) who died during a one year period were examined histologically.

Microscopical examination revealed narrowing,

intimal fibrosis, and vascularization of the media and adventitia, with adventitial fibrosis and a varying degree of round cell infiltration in twelve cases. These changes were interpreted as sequelae to arteritis. The patients' clinical histories were studied. Two had had histologically verified arteritis 2 years before death, and seven had had periods of headache, muscle pain, and general malaise, which could be interpreted as rheumatic polymyalgia or temporal arteritis, 1 to 14 years before death. Some also had a markedly raised erythrocyte sedimentation rate. Arterial changes were seen in 1 per cent. The prevalence of temporal arteritis in this necropsy series was thus much higher than the

incidence of about two cases per 100,000 population per year, estimated from positive biopsies during the last 10 years.

Sequelae to arteritis seem to have a characteristic persistent histological pattern. The findings suggest that temporal arteritis is often undiagnosed and that patients dying from other causes are also suffering from this condition.

Temporal arteries from 230 necropsies in a mainly rural district showed suspicious post-arteritic changes in four cases, in two of which there was a possible history of temporal arteritis, but the numbers are too few for conclusions to be drawn about differences between urban and rural areas.

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