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ORIGINAL COMMUNICATION

Karl Pfadenhauer
Hermann Weber

Giant cell arteritis of the occipital arteries

A prospective color coded duplex Sonography study in 78 patients

Abstract *Context* Occipital headache and nuchal pain may indicate the involvement of the occipital arteries (OCCA) in temporal arteritis (TA). Recently high resolution color coded sonography (CCDS) has greatly improved the imaging of small lumen arteries. *Objective* The aim of the present study was the demonstration of TA of the OCCA in comparison with the superficial temporal artery (STA) by means of CCDS in patients with nuchal and occipital pain suspected of suffering from TA. *Design* Prospective study of 78 patients comparing CCDS findings of the OCCA and of the STA with the clinical diagnosis and the biopsy results. *Results* 27 patients received the diagnosis TA; there were 51 other diagnoses. CCDS of the OCCA reached a sensitivity in diagnosis of 63 % and a specificity of 100 % and in histology of 65 % and 100 % respectively. CCDS of the STA reached a diagnostic sensitivity of 78 % and a specificity of 94 % and of a histological sensitivity of 77 % and specificity of 82 %. Reversibility of CCDS abnormalities was monitored in 5 patients over a period of 13 to 42 days. *Conclusions* Involvement of the OCCA in TA patients is a frequent finding and may be the only pathological phenomenon in some patients with nuchal pain, occipital headache and occipital scalp tenderness. CCDS of the STA and OCCA contributed to the diagnosis of TA with a high rate of perivascular hypoechogenic abnormalities (stenoses and occlusions) and a low rate of these abnormalities in the control patients. However, CCDS cannot differentiate between inflammatory and degenerative artery disease and has limitations concerning spatial resolution. Before CCDS may replace biopsy in clinical practice the accuracy of the criteria recommended above should be tested in larger groups of patients.

Key words temporal arteritis · occipital artery · duplex sonography

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K. Pfadenhauer, MD · H. Weber, MD
Dept. of Neurology
and Clinical Neurophysiology
Klinikum Augsburg
Augsburg, Germany

Dr. K. Pfadenhauer (✉)
Stenglinstraße 2
86156 Augsburg, Germany
Tel.: +49-821/400-2991
Fax: +49-821/400-2691

Introduction

Occipital headache, nuchal pain and occipital scalp tenderness [5, 9, 10, 21] may indicate the involvement of the occipital arteries (OCCA) in temporal arteritis (TA). Radiological [3] and ultrasound [20] studies have demonstrated the involvement of OCCA in 66–69 % of patients suffering from temporal arteritis. Temporal arteritis of OCCA may even be the only manifestation of arteritis in some patients [9]. Recent high resolution color coded duplex sonography (CCDS) has greatly improved the imaging of small lumen arteries and has been introduced in the diagnosis of TA by Schmidt et al. [16]. In the superficial temporal artery (STA) perivascular hypodense areas (so called halos) and stenoses and occlusions have been found.

The aim of the present study was the demonstration of giant cell arteritis (GCA) of the OCCA in comparison with the STAs by means of CCDS in patients suspected of suffering from arteritis of the occipital arteries.

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Patients

Over a period of 51 months we examined 115 patients who were suspected of suffering from TA. In 78 patients we examined the OCCA and the STAs because of occipital headache, nuchal pain and occipital scalp tenderness. A final diagnosis of TA was made in 27 patients (female/male: 22/5, age 52–90 years, median 76). Steroid treatment had been started in 19 patients before CCDS and biopsy. 26 patients underwent biopsy from the STA in 25, from the OCCA in 1 case. Histopathology demonstrated temporal arteritis in 23 patients. In all patients the diagnosis of TA was made according to the American College of Rheumatology 1990 (ACR) criteria for the classification of temporal arteritis [7].

As a control group we examined 51 patients (female/male: 36/15, age 39–88, median 72 years) who were given other diagnoses (arteriosclerotic vascular disease n = 18, autoimmune disease n = 16, polymyalgia rheumatica (PMR) n = 8 and other diagnoses n = 9). 17 patients underwent biopsy from the STA and no TA was found. In all patients only one biopsy was taken.

Anatomy

The occipital artery arises from the external carotid artery and passes through the sulcus of the OCCA along the medial border of the mastoid process. The diameter of the main stem of the OCCA is between 1 and 2 mm. On the way to the occipital scalp the OCCA penetrates the tendinous arch of the M. splenius capitis. From there it ascends over the occipital scalp and divides into a variable number of branches which supply the occipital skin and the occipital muscle. On the occipital scalp the artery lies in the dense subcutaneous tissue external to the galea aponeurotica. Lateral to the OCCA a small vessel, the posterior auricular artery, can be detected (Fig. 1).

The common trunk of the superficial temporal artery can easily be found when the artery passes through the parotid gland anterior to the tragus. From there the artery can easily be followed to the bifurcation and to the posterior and anterior branches. Both branches are embedded between the two layers of the fascia temporalis, which can be visualized clearly as two parallel hyperechogenic lines.

Methods

CCDS was performed in all cases before biopsy. The ultrasonographer was not blinded to the clinical situation. The decision to perform biopsy was made by the clinical staff in charge of the patient. All CCDS examinations were done by one experienced physician ultrasonographer (KP) using a Siemens Elegra high resolution ultrasound system and a multifrequency (6–9 MHz) linear transducer. Axial resolution was 0.45, lateral resolution 0.49 mm. Examination was started using standardized parameters including: B-Mode: Echo transmission frequency 9 MHz, dynamic range 50 Hz, gain 70 db. For color coded doppler sonography: Pulse repetition frequency (PRF) 3125 Hz, wall filter 50 Hz.

Examination of the OCCA started at the level of the tendinous arch of the M. splenius capitis, where the artery usually can be found 1–2 cm below the skin surface. From there the artery can easily be followed over the scalp. Routine examination included at least 3–4 cm long segments of the artery on both sides. Examination of the STAs started at the level of the tragus and included the common trunk and the anterior and posterior branches which were followed as far as possible (usually 4–5 cm)

A *halo* was diagnosed when periarterial hypoechogenic areas were detectable without local changes of the doppler frequency spectrum (Fig. 2). A *stenosis* was assumed in segments of the arteries where local increases of the doppler frequency spectrum and poststenotic flow disturbances were found. When there was no flow in the typical anatomical distribution of the OCCA and the STA was detected an *occlusion* of the artery was assumed. Widespread arteritis was assumed when perivascular hypoechogenic abnormalities extended to all parts of the artery.

Results

Occipital arteries

TA patients

CCDS of both OCCA was normal in 10/27 TA patients (37 %). Halos were found in 13, halo and stenosis in 3 and occlusion in 1 of the TA patients. About half of the abnormalities were bilateral. In one TA patient the only abnormal finding was a halo and a stenosis of one of the OCCA. Widespread halos were found in 7, segmental abnormalities in 10 TA patients. One of the OCCA of a TA patient was occluded.

Control patients

All OCCA of the 51 patients who were given other diagnoses were detectable and had normal arterial walls.

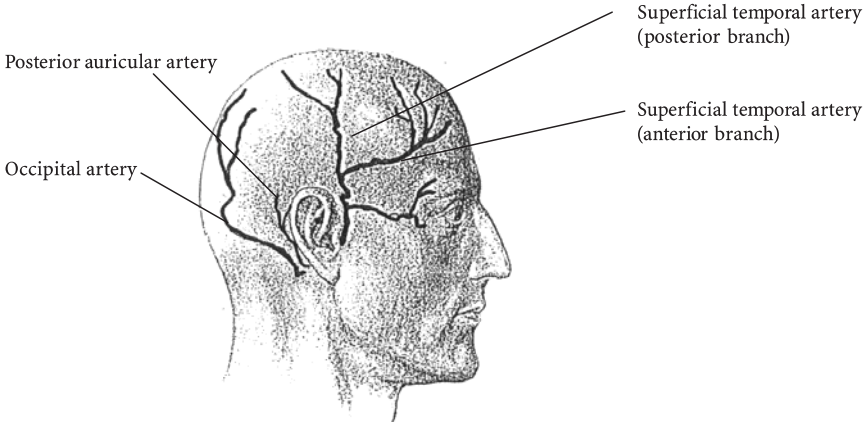

Fig. 1 Anatomy of the superficial temporal artery and the occipital artery

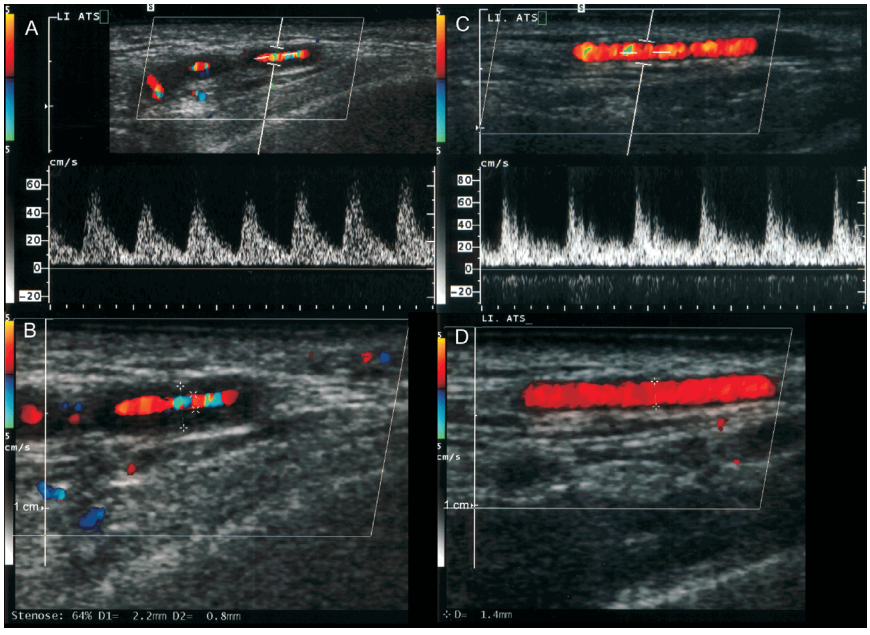

Fig. 2 Color-coded duplex sonography of the superficial temporal artery in a 73-year-old patient (patient 4 from table 2) suffering from temporal arteritis (**a, b**) and in a 75-year-old healthy woman (**c, d**). CCDS longitudinal sections from the anterior branch of the STA show normal perivascular tissue (**d**) and hypoechogenic perivascular tissue corresponding to active giant cell arteritis (**b**). Corresponding pulsed-wave-Dopplerspectra from the STA (**a, c**) showing normal results

Superficial temporal artery

TA patients

6/27 patients (22 %) had normal findings. In all other TA patients abnormalities were found including 14 patients with halos as the only abnormalities. Halos in combination with stenoses were found in 5, occlusions in 2 patients. In these 2 patients with bilateral occlusion, on initial examination a large hypoechogenic area was seen in the typical anatomical distribution of the common trunk of the STA and the anterior and posterior branches. Widespread halos were found in 12, segmental abnormalities in 7 TA patients.

Control patients

In 3 patients from the control group false positive halos were found, bilateral in one of them. In these cases the halos were restricted to one segment of the arteries. Final diagnoses were arteriosclerotic vascular disease in 2 and Waldenströms disease in 1 patient. In all 3 patients biopsy failed to demonstrate giant cell arteritis. In the case with bilateral halos biopsy showed arteriosclerosis, in the 2 other patients with unilateral halos biopsy showed fibrotic changes in the intima layer.

CCDS findings on follow up after steroid treatment was started

In 5 TA patients CCDS follow up was done showing recanalization of occlusions of both STAs in 2, resolution of one halo/stenosis in 1 and resolution of halos in 2 patients between day 13 and 42 after initial CCDS. In 1 case with occlusions of both STAs an initial examination conducted 5 months before the diagnosis of TA was made, showed no pathological findings.

Histopathological diagnosis

In 23 TA patients (85 %) the diagnosis was confirmed by positive histological findings. In 3 cases biopsy was negative and one TA patient refused biopsy. In all these patients the diagnosis was made by at least 3 other positive ACR-criteria, a prompt steroid response and exclusion of other diseases. Biopsies were taken according to the CCDS findings from the STA in 25 and the left OCCA in 1 patient.

Comparison between CCDS findings, diagnosis and biopsy results

The sensitivity and specificity of CCDS for the diagnosis of TA was evaluated for the OCCA and STAs, comparing the CCDS findings with the biopsy results and the final clinical diagnosis. The results are summarized in Table 3.

The most important findings are:

1. No patient from the control group had abnormalities in more than 2 arteries in contrast to the TA patients: 4 arteries were involved in 6 and 3 in another 6 TA patients all with positive biopsy results.
2. No patient from the control group had occlusions or

Table 1 CCDS findings in 27 patients with the final diagnosis AT suffering from occipital headache, nuchal pain and occipital scalp tenderness

Patient	Age (years)	A.occipitalis		A.temporalis supf.		Biopsy
		right	left	right	left	
1.	80	H	H	N	H	+
2.	80	H	H	N	H	+
3.	90*	H	H	H	H	+
4.	68*	N	H	H	H	+
5.	65	N	H	H	S/H	+
6.	52	N	N	N	N	+
7.	76	S/H	N	O	O	+
8.	68	N	N	H	N	+
9.	78	H	N	H	H	+
10.	74	H	H	H	H	+
11.	84*	H	H	H	S/H	+
12.	80*	O	N	H	N	+
13.	80	H	N	S/H	N	+
14.	63	N	N	O	O	+
15.	78	H	H	H	H	+
16.	78	H	H	H	H	+
17.	80	N	N	H	H	+
18.	76	N	H	H	N	+
19.	68	S/H	H	S/H	S/H	+
20.	77	S/H	N	N	N	+
21.	61	N	N	H	H	n. d.
22.	67	N	N	N	N	–
23.	72	N	N	N	H	+
24.	64	N	N	N	N	–
25.	77	H	N	S/H	S/H	+
26.	76	N	N	N	N	+
27.	58*	N	N	N	N	–

N normal; *H* halo; *S* Stenosis; *S/H* halo and stenosis; *O* Occlusion; *nd* not done; * male patients

Table 2 Summary of the final diagnoses and the results of CCDS of the occipital arteries (OCCA) and superficial temporal arteries (STA) in 78 patients

Diagnosis	CCDS			
	Occipital artery (OCCA)		*Temporal artery (STA)*	
Temporal Arteritis n = 27	Normal	10	Normal	6
	Abnormal	17	Abnormal	21
	Bilateral	8	Bilateral	14
	Unilateral	9	Unilateral	7
	Occipital and temporal arteries			
	Normal	5		
	Abnormal	22		
	4 Arteries	6		
	3 Arteries	6		
	2 Arteries	6		
	1 Arteries	4		
Other Diagnoses n = 51	Normal	51	Normal	48
			Abnormal	3
			Unilateral	2
			Bilateral	1

halo and stenosis: 8 TA patients had occlusions or halo and stenosis all with positive biopsy results.
3. In all patients with widespread abnormalities biopsies revealed GCA.
4. On follow-up in all TA patients steroid therapy led to a normalisation of the arterial wall abnormalities.

Discussion

Involvement of the occipital arteries in TA

In the TA patients the rate of abnormal CCDS findings in the OCCA was somewhat lower (63 %) than in the STAs (78 %). Bilateral abnormalities were also more frequent in the STAs (52 %) than in the OCCA (30 %). This is in accordance with previous ultrasound and radiological studies. Vinckier et al. [20] studied 59 biopsy-positive patients suffering from TA by examining the OCCA and the STAs by continuous wave doppler sonography. Abnormalities in the OCCA were found in 69 % and in the STAs in 81.5 % of his 59 patients. Arteriographic studies of the external carotid artery were performed by Godeau et al. [3] in TA patients. In 6 biopsy-positive patients GCA-typical abnormalities were found in the STAs. Involvement of the OCCA was detected in 4 patients. From multiple biopsy studies in some patients it is known that histopathological analysis for TA was negative in both STAs and positive only in the OCCA [4, 10]

Sensitivity of diagnostic methods in TA

The rate of false negative temporal artery biopsies is in the range of 10–61 % and pathological proof of TA may be difficult in some patients mainly because of the segmental nature of the disease [1, 2]. Therefore multiple biopsies can increase the rate of positive findings [4]. There are cases with positive biopsy findings from the OCCA after biopsies of the STA had been negative [4, 10]. In contrast to biopsy CCDS allows full length examination of the STA and the OCCA and monitoring of disease activity in response to therapy over time [16]. The main problem for CCDS concerning sensitivity is the limited spatial resolution, which may prevent the detection of small lesion [1]: The different extent of arterial wall inflammation has been recognized and described already by Horton [6] and Jennings [8] and ranges from severe occlusive necrotizing panarteritis with involvement of the adventitia to more restricted manifestations in the intima and media layer leaving the arterial lumen unchanged. In addition there may be particular histological aspects of the inflammation which also affect echogenity of the inflamed arterial wall.

Reinecke and Kuwabara [14] found different histological types of inflammation ranging from an acute

Table 3 Sensitivity and specificity of color coded duplexsonography (CCDS) of the occipital artery and superficial temporal artery for the diagnosis of temporal arteritis and to confirm histopathological findings

	Occipital artery		Superficial temporal artery	
	Sensitivity (%) (positive tests/total)	Specificity (%) (negative tests/total)	Sensitivity (%) (positive tests/total)	Specificity (%) (negative tests/total)
CCDS and Diagnosis	63% (17/27)	100% (51/51)	78% (21/27)	94% (48/51)
CCDS and Histology	65% (17/26)	100% (17/17)	77% (20/26)	82% (14/17)

27 patients received the diagnosis TA (including 26 biopsy-controlled patients), 51 patients other diagnoses (including 17 biopsy-controlled patients)

stage characterized by massive leukocyte infiltration, wall necrosis and edema to more granulomatous and fibrotic changes in more chronic stages of the disease.

Diagnostic specificity

Schmidt et al. [16] consider the demonstration of a perivascular hypoechogenic area in the STA as a GCA-specific phenomenon indicating the presence of inflammatory periarterial edema. This has been questioned by Ventz et al. [19], who found a perivascular halo in 2 patients with arteriosclerotic disease proven by biopsy.

Our study shows that false positive results are possible, because CCDS cannot differentiate between arteriosclerotic and arteritic wall changes. However, they occur at a low rate of 6% in the STAs and they were completely absent in the OCCA. In addition occlusions and stenoses were not observed in patients with other diagnoses than GCA.

Comparison with other Studies

Our findings concerning sensitivity of CCDS in TA patients are in accordance with other studies [13, 15–18]. The low rate of halos in the study by Lauwerys et al. [11] may result from patient selection. There is a difference in the rates of stenosis and occlusion as evident by the 80% in the study by Schmidt et al. [16] and the 19% in our patients, a difference which might be only in part explained by the selection of more severely affected patients. For the diagnosis of temporal arteritis CCDS of the STAs and OCCA is employed as a new diagnostic approach using a technology at the upper limit of its technical capacity. Up to now experience with this rare disease is limited and the results are more investigator-dependent than in other vascular diseases ultrasound is applied to. Important other factors affecting diagnostic sensitivity and specificity as well include patient selection, diagnostic criteria, examination technique and ultrasound equipment:

For these reasons the range of diagnostic sensitivity (9–100%) and specificity (61–86%) in the available studies reporting on biopsy controlled patients [11, 13, 15, 16, 17, 18, 19]) is not surprising.

The occipital arteries were studied in 2 studies including 20 [15] and 33 [18] patients with the diagnosis TA and 66 patients with other diagnosis [15]. In both studies no biopsies from the OCCA were performed. No abnormalities were seen in one study [15], while halos were found in 9% of the patients studied by Schmidt et al. [18]).

Conclusion

Involvement of the OCCA in TA patients is a frequent finding and may be the only manifestation in some patients with nuchal pain, occipital headache and occipital scalp tenderness. CCDS of the STAs and OCCA contributes to the diagnoses of arteritis with a high rate of perivascular hypoechogenic abnormalities (so called halos), stenoses and occlusions. The main advantage of CCDS is a full length visualisation of the entire STA and OCCA. However, there is a low rate of these abnormalities in the control patients because CCDS cannot differentiate between inflammatory and degenerative artery disease. On follow up CCDS abnormalities resolve in GCA patients under steroid treatment. In patients with unavailable or negative biopsy results CCDS of the OCCA and STAs may add a substantial contribution to the diagnosis of TA and can help in the selection of affected arterial segments for biopsy. The most specific CCDS findings are: widespread involvement of more than 2 of the 4 arteries examined, halo and stenosis and the reversibility of these abnormalities under steroid treatment. Before CCDS may replace biopsy in clinical practice the accuracy of the above recommended criteria should be tested in larger groups of patients. Progress in ultrasound technology and the use of high resolution high frequency ultrasound systems may be helpful on the way to accurate atraumatic diagnosis of TA.

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