

## Neurologic Manifestations of Churg-Strauss Syndrome

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**Background** : Churg-Strauss syndrome (CSS) is characterized by disseminated vasculitis with multi-organ involvement. The purpose of this study was to determine the frequency and the types of neurologic involvement in a series of patients with CSS. **Methods** : We reviewed the medical records of 16 patients (seven men and nine women, age = 41.9 (18.6) with CSS who were examined at Seoul National University Hospital. The diagnosis of CSS was based on the presence of asthma, peripheral eosinophilia (more than 10% eosinophiles), and histopathological or clinical findings of vasculitis. **Results** : Of the 16 patients, 12 (75%) had neurologic involvement. Nine (56%) had neurologic symptoms as initial presentations of CSS. Peripheral neuropathy was detected in nine patients (56%) ; six had multiple mononeuropathy, and three had distal symmetric polyneuropathy. Three patients (19%) had cerebral infarctions; in two of them, a delayed diagnosis of CSS caused the recurrence of ischemic stroke. Corticosteroid therapy combined with immunosuppressive agents usually yielded improvement or stabilization of symptoms. **Conclusions** : Neurologic involvement is common in CSS, usually manifesting as peripheral neuropathy. Neurologic symptoms are important initial manifestations at the time of diagnosis of CSS. Furthermore, cerebral involvement is not uncommon; thus any neurologic symptoms in patients with asthma or eosinophilia prompts an aggressive diagnostic approach to CSS.

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**Key Words** : Churg-Strauss syndrome, Neurologic manifestation, Peripheral neuropathy, Cerebral infarction

(allergic granulomatosis and angiitis) (Churg-Strauss syndrome) 가 , 2 30 63% Lanham 3 16 75% , Sehgal 4 47 62% 가 1951 Churg Strauss'가 5-7 가 . 1977 Chumbley

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1) , 2) 가 X  
 , 3) ( , 36.3 (±19.0)  
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 2. 1987 1998 (Table 1).  
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 16 7 , 16 12 (75%)  
 9 , 17 74 9 (56%), 3  
 41.9 (±18.6) (19%)  
 14 , 3  
 (10 ), (9 ), (2 ), (2) 44.3 (±18.3)  
 ), (1 ), (1 ), (1 ) 4.5  
 2 (Table 1). 2

**Table 1.** Clinical data for 16 patients with Churg-Strauss syndrome (CSS)

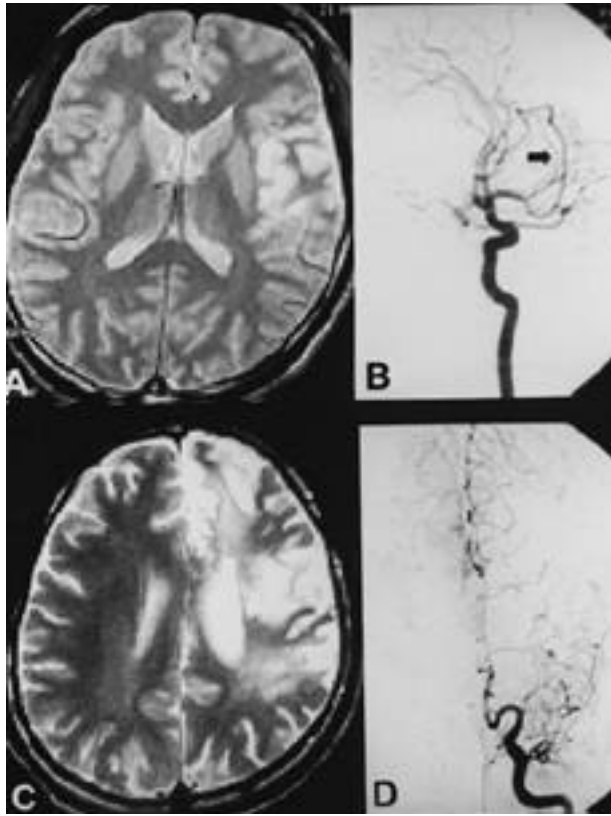
	Range (Mean)
Age at diagnosis of CSS	17-74yr (41.9yr)
Age at onset of asthma	5-73yr (36.3yr)
Duration of asthma before diagnosis of CSS	0*-20yr ( 5.7yr)
Age at onset of neurologic symptoms	17-74yr (44.3yr)
Duration of asthma before neurologic symptoms	0*-20yr ( 4.5yr)

\* Two patients were confirmed to have asthma by metacholine broncho-provocation test at the time of neurologic symptoms and diagnosis of CSS.

**Table 2.** Nerve lesions in 6 patients with multiple mononeuropathy on nerve conduction studies

Case	Sex/age	Tested limbs	Ulnar		Median		Radial		PT	Peroneal	Sural	Subclinical lesion
			M	S	M	S	M	S				
1	M/66	B u/e, L l/e	L		R>L	L				L		Peroneal
2	M/74	L u/e, B l/e		L						L>R	B	
3	F/35	B u/e, R l/e	B	R>L	L	R	L	L>R		L		
4	F/46	B u/e, B l/e	R>L	R>L	B	B	B	B	L>R	B	L	
5	M/18	B u/e, R l/e	R>L	R>L		B			R	R	R	L u/e, B l/e
6	F/62	L u/e, L l/e		L	L	L			L	L	L	

Abbreviations : PT=posterior tibial; M=motor; S=sensory; R=right; L=left; B=bilateral; R>L=right is worse than left; u/e=upper extremity; l/e=lower extremity



**Figure 1.** Brain MRI and cerebral angiogram performed in the patient 1 of table 4: brain MRI shows left MCA infarction (A), and cerebral angiogram shows left MCA stenosis (arrow, B) at the first stroke event. Follow-up brain MRI shows more extended left MCA infarct and newly developed left ACA infarct (C) and left internal carotid angiogram shows MCA occlusion and ACA stenosis (D) when stroke recurred.

**Table 4.** Clinical and laboratory features of 3 patients with cerebral infarction

Case	Sex/age	Risk factors	Symptom & signs	Lesion on MRI or CT	Angiographic findings	Echocardiogram	Comments
1	M/22	None	Aphasia, right hemiparesis	Left MCA, ACA large infarction	Left MCA occlusion & ACA stenosis	Normal (TTE & TEE)	Recurrent stroke
2	M/34	None	Aphasia, right hemiparesis	Multifocal infarcts in left frontal, parietal, and right frontal	Multifocal narrowing of intracerebral arteries	Normal (TTE)	Recurrent stroke
3	M/39	Smoking (40PY)	Visual field defect & dysarthria	Multifocal infarcts in both frontal, parietal, occipital cortex	Not done	Normal (TTE & TEE)	Remained well after steroid treatment

PY=pack year; MCA=middle cerebral artery; ACA=anterior cerebral artery; TTE=transthoracic echocardiogram; TEE=transesophageal echocardiogram

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 3) 가  
 가 . 1990 The American College of Rheumatology 가 807  
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 6가 .<sup>8</sup> 1) , 2)  
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 16 -  
 75% , Chumbley  
<sup>2</sup> 63%, Lanham<sup>3</sup> 75%, Sehgal<sup>4</sup> 62%  
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12 (75%) 16  
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9 가

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