

# CORTICAL BLINDNESS COMPLICATING ACUTE NEPHRITIS — A CASE REPORT AND ELECTROPHYSIOLOGICAL STUDY

G Devathanan  
W C Tan  
B K Boey  
P K Wong

## SYNOPSIS

A thirteen year old Chinese schoolgirl was admitted for generalised oedema, hypertension and haematuria. She was treated for acute nephritis. Three days later she developed sudden total blindness. Pupils were reactive and funduscopy was normal. CT scan showed translucent areas in the occipital poles and the EEG revealed abnormal occipital slow waves with loss of alpha rhythm. The pattern visual evoked response was done when she regained some vision to light. It was of normal latency and waveform but reduced amplitude. A week after onset of blindness she showed complete recovery with normal CT scan, improving EEG and normal visual evoked response. The underlying mechanism is probably focal spasm of branches of the posterior cerebral arteries with consequently a good prognosis.

## INTRODUCTION

The term cortical blindness is aptly confined to visual loss arising from bilateral lesions of the occipital cortex and the immediate underlying white matter, excluding more anterior lesions affecting the optic radiations (1). The wide variety of causes include leucodystrophies, carbon monoxide poisoning, encephalitis, meningitis, occlusive arterial disease, hypotensive crisis, tumours and vertebro-basilar artery angiogram (1, 2, 3, 4, 5, 6). Our patient, a schoolgirl, was unique in that she presented with sudden cortical blindness following hypertension secondary to acute post-streptococcal glomerulonephritis. The only other similar reported case was by Bramwell et al (7) in 1915. Her symptoms were fairly localised to the occipital cortex unlike a typical hypertensive encephalopathy and illustrates the susceptibility of the visual cortex to ischaemia. We also had the opportunity to demonstrate the value of the pattern visual evoked response in this condition, and the characteristic EEG changes.

## CASE REPORT

A thirteen year old Chinese schoolgirl presented with puffiness of the face, haematuria, fever and headache for three days. Previously she was in perfect health and she had no visual symptoms during admission.

She was slightly tachypneic, febrile, with a blood pressure of 160/110. Her face was puffy, with pitting ankle oedema, bilateral pleural effusion and ascites. She was not in cardiac failure. Neurological examination was normal. A diagnosis of acute glomerulonephritis was made.

**Department of Medicine  
National University of Singapore  
Singapore General Hospital  
Outram Road  
Singapore 0316**

G Devathanan, MBBS, M Med, MRCP  
Senior Lecturer

W C Tan, MBBS, MRCP, FRACP  
Senior Lecturer

P K Wong, MBBS, FRCP (E), FRCP  
Professor and Head

**Department of Radiology  
Tan Tock Seng Hospital**

B K Boey, MBBS, DMRD, FRCR  
Senior Radiologist & Head

Urine examination showed 35-45 red cells, although no white cells and casts were seen. 24 hours urinary protein measured 1.64 gm. Urea and electrolytes were normal. Antistreptolysin O titre was 630 Todd units suggesting recent streptococcal infection. Serum albumin was 3.1 gm/100 ml.

The patient was treated with methyldopa 250 mg bd, frusemide 40 mg bd and Penicillin. She made good recovery and the blood pressure dropped to 140/100. There was a good diuresis.

On the 3rd hospital day she woke up in the night complaining of total blindness. She groped with her hands and could not feed herself. There was a lack of visual fixation and pendular drifting of the eyes was noted. Fundoscopy was normal. The only other positive neurological sign was slight nuchal rigidity.

Six hours later when examined by an ophthalmologist she denied blindness and confabulated when presented with a few familiar objects to identify. She was unable to perceive even light.

CT scan on the same day showed bilateral translucent areas (Fig. 1) which did not take up contrast dye. EEG showed bilateral occipital slow waves with loss of alpha rhythm especially in the left (Fig. 2). CSF examination was normal.

The patient was not anti-coagulated as other parameters were improving. On the 7th hospital day she perceived movements, followed by flashes of light and images. Eventually perception of colour returned. By the 10th day she could see clearly. A second awake EEG recording showed some improvement and appearance of some alpha rhythm. The pattern visual evoked response was first performed on the 8th hospital day when she had some vision and was able to see and concentrate on the checker board pattern. The test was performed using a Medelec MS6 with a VS6 stimulator and a black and white TV monitor. Each square subtended an angle of 1° at the macula and the active recording needle electrode was placed 2 cm above the inion along the midline. Flicker rate was at 2/sec. The results are given in table 1. Latency was measured to 1st positive peak and amplitude was from 1st positive peak to the following negative peak. The latencies were normal but the amplitude responses were significantly reduced in both eyes. The test was repeated 4 days later when she regained normal vision and showed improvement in the amplitude of the visual evoked responses.

She was discharged on the 20th hospital day. There was no further evidence of oedema, haematuria or hypertension. Perimetry was normal. Angiogram was

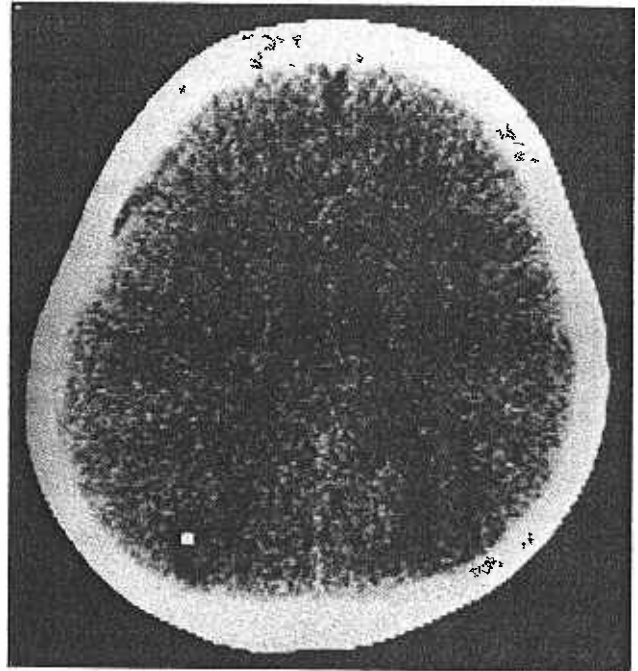


Fig. 1(a) 1st CT Scan (without contrast)  
Case of Cortical Blindness with bilateral occipital translucent areas.

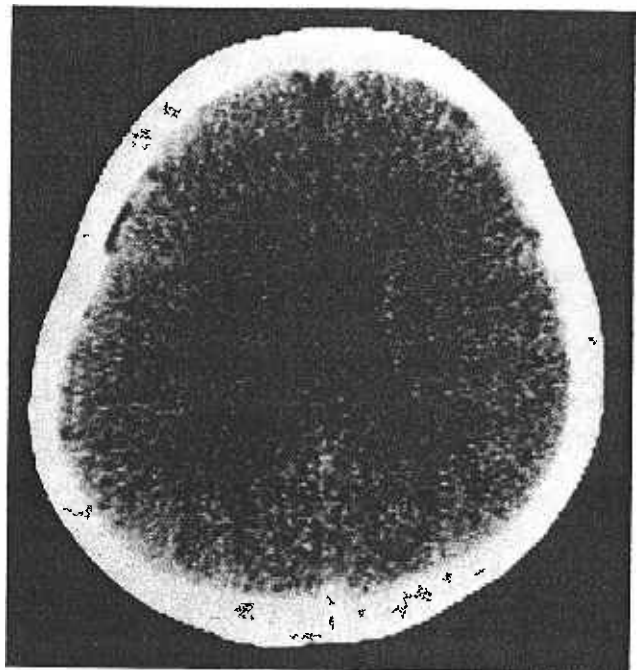


Fig. 1(b) 2nd CT Scan (without contrast)  
1 week later - normal.

**Table 1**  
**Visual Evoked Responses In A Patient With Cortical Blindness Complicating Acute Nephritis**

	Right Eye Amplitude & Latency to P <sub>1</sub>	Left Eye Amplitude & Latency to P <sub>1</sub>	Normal Amplitude & Latency to P <sub>1</sub>
1st Recording	4 uV 99.2 msec	4.2 uV 98.2 msec	9.0 uV (4 uV ± 2 SD)
2nd Recording 4 days later	12.0 uV 100.0 msec	9.1 uV 99.0 msec	latency below 107 msec (3 SD)

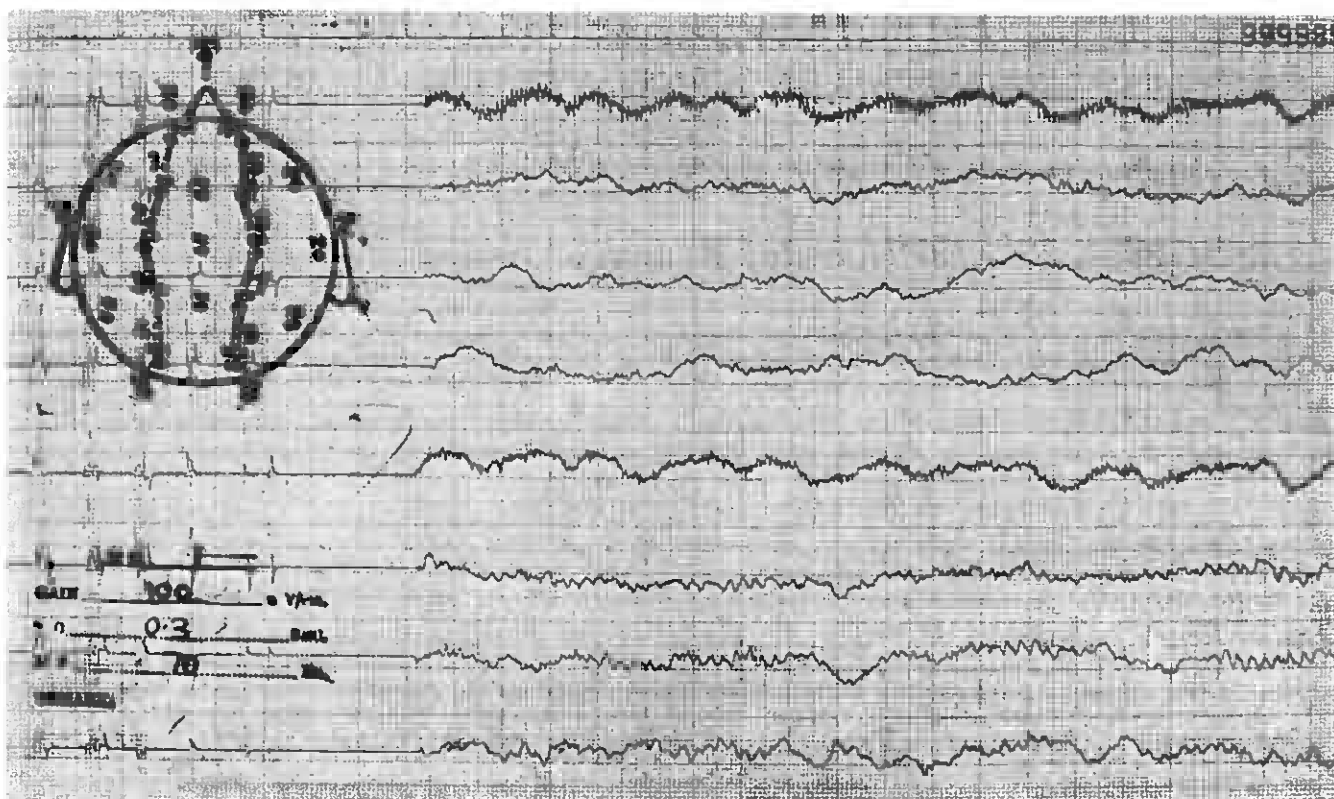


Fig. 2(a) EEG in a patient with cortical blindness complicating acute nephritis. EEG was done in the acute phase and shows slow waves more prominent posteriorly. Significant loss of alpha rhythm over left occipital region.

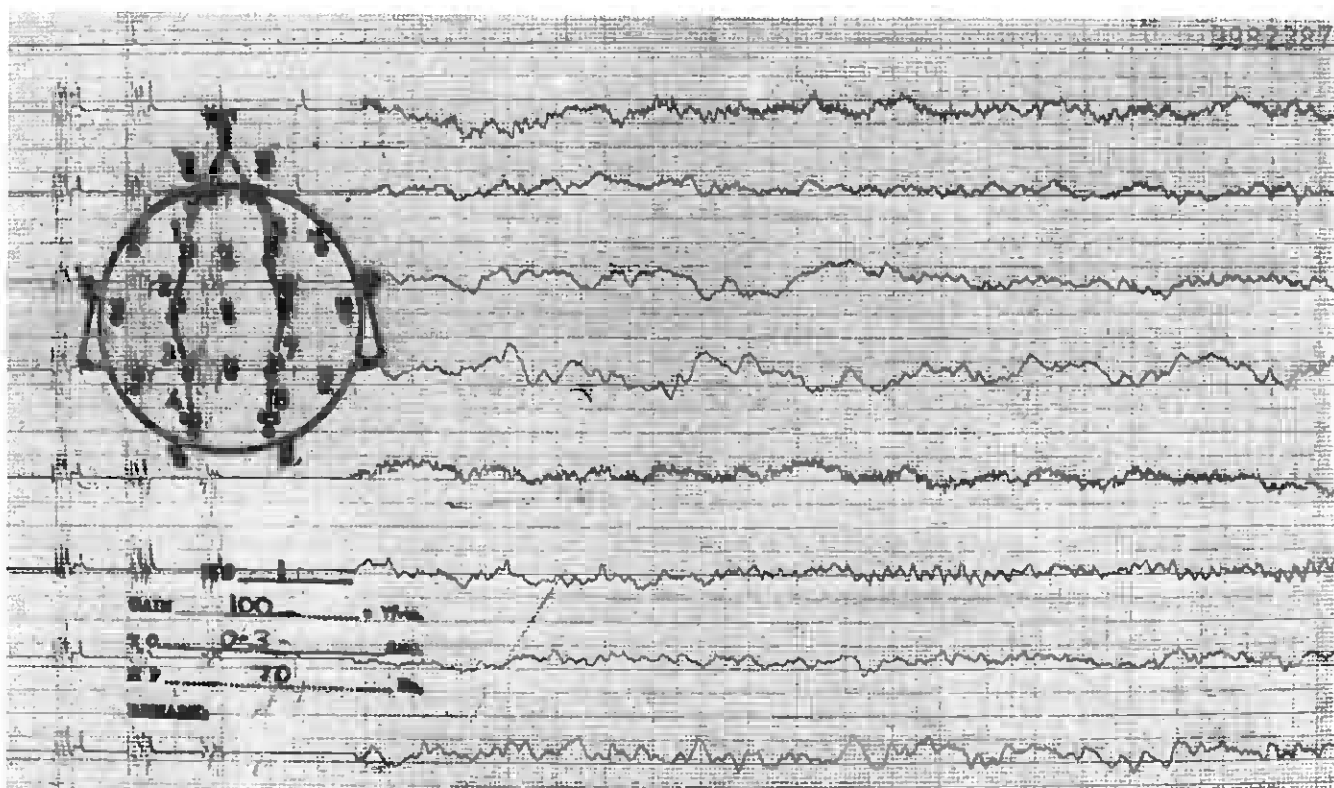


Fig. 2(b) Repeat EEG one week later when vision returned to normal clinically. Abnormalities still persisted but with some improvement. Repeat EEG six weeks later was within normal limits.

not performed at any stage as it was felt that this invasive procedure would not contribute to the management.

## DISCUSSION

### Clinical

Hysterical blindness was confidently excluded here because of the absent blink reflex to abnoxious stimuli, the presence of pendular nystagmus and her inability to feed herself. Denial of blindness is not a feature of hysterical blindness but is a common complication of cortical blindness when visual association areas and projection subcortical fibres of the left dominant occipital lobe are involved. When her vision was improving movements were first appreciated. Riddoch (8) had made the observation that movement perception may be present in the absence of perception of stationary objects. The transitory tendency to confabulate appeared similar to Korsakoff's psychosis. However we feel that this was not due to a defect in memory and involvement of limbic or diencephalic structures but rather arising out of her actual inability to see and interpret. The patient filled the gap by 'guessing' because of repetition of the question to identify an object visually and her desire to reply to the auditory request. In other words, confabulation occurs with Anton's syndrome.

### Mechanism

The CT scan did not distinguish between ischaemia with oedema and acute infarct. The rapid and complete resolution suggests strongly that ischaemia without infarct is the underlying pathology. The underlying mechanism is probably autoregulatory focal spasm of the posterior cerebral arteries in response to the elevated blood pressure. Such focal spasms of the posterior cerebral arteries have been reported in subarachnoid haemorrhages (2). Ensuring local oedema may compress the posterior cerebral arteries against the tentorium causing further ischaemia (9). Thrombus at the bifurcation or multiple microemboli would not be a likely mechanism as she is young, without evidence of underlying diffuse arterial disease, there is no source of emboli and the nephritis was subsiding rapidly. Special angiographic studies of the posterior circulation with tomographic cuts (3) might resolve this issue. However this investigation in itself is a well known cause for temporary cortical blindness, the suggested mechanism being transitory alterations of the blood brain barrier in the striate cortex (4, 5), Linderberg (10) has shown that the visual cortex, in particular Lamina 3 & 4 of Broadman, is prone to hypoxic injuries.

In the classical hypertensive encephalopathy there is general but patchy oedema of the brain with exudates and petechial haemorrhages (11). Patchy necrosis of walls of small arteries allows escape of fluid. In this patient the changes are more prominent in the occipital region and in that sense is atypical. Moreover, the blood pressure was not very high at the onset of blindness. Byrom (12) demonstrated focal vasoconstriction in some vessels using hypertensive

rats. Lassen and Agnoli (13) by studying regional cerebral blood flow have demonstrated in man, increase in cerebral blood flow when there is an acute blood pressure rise. This could be due to a breakdown of autoregulatory mechanisms. Lassen's observations do not necessarily contradict Byrom's experimental observations of vasospasm since one phase could well lead to another i.e. vasoconstriction followed by vasodilation when autoregulation is lost. Such micropatho-physiological changes are well documented in the systemic peripheral circulation.

### Electrophysiological changes

EEG changes in cortical blindness have been reviewed by Bergmann (1). Figure 2 shows slow waves prominent in the occipital region with loss of alpha rhythm although patient was awake and quiet. There was complete resolution of EEG abnormalities only a month later. EEG remains a useful and quick way of differentiating hysterical blindness from organic cortical blindness. Minimal or early lesions may not show radiologically but may manifest as EEG abnormalities, by the presence of posterior slow waves. EEG response to flickering light was also studied by Bergmann.

The pattern visual evoked response obviously could not be done when she was totally blind and unable to fixate. Interpretation of results would be doubtful. With returning vision we demonstrated that while latency and waveform were normal on both sides, the amplitudes were reduced inspite of variability of this parameter. Delay in latency is due usually to myelin disease and in ischaemia myelin is relatively resistant. Occipital neurons, however, in layer 4 especially, suffer easily from hypoxia and this may account for reduced amplitude. Normal latency and waveform might imply good prognosis.

### Prognosis:

The chances of recovery in cortical blindness depends entirely on the cause and is variable. Those due to acute reversible hypotension, cardiac arrests and angiograms, have a good prognosis. Hoyt (2, 14) states that recovery may continue for months later. Optimism is therefore worthwhile as illustrated by our patient.

## REFERENCES

1. Berman PS: Cerebral Blindness, an analysis of 12 cases with especial reference to the Electroencephalogram and patterns of recovery. *A M A Arch Psychiat* and *Neurol* 1957; 78: 568-84.
2. Hout WF, Newtown TH: Angiographic changes with occlusion of arteries that supply the visual cortex; *N Z Med J* 1970; 72: 310-7.
3. Grimsdale, H: A case of gas-poisoning with unusual ophthalmological complications; *Brit J Ophthal* 1934; 18: 443-6.
4. Horwitz: MH, Lewis W: Temporary cortical blindness following angiography; *J Neurosurg* 1974; 40: 583-5.
5. Studdard WE, Davis DO, Young SW: Cortical Blindness after cerebral angiography; *J Neurosurg* 1981; 54: 240-4.
6. Mod DJ, Kwun YA: Cortical blindness as a complication of haemodialysis; *J Pediatr* 1978; 93: 890-1.

7. Bramwell B, Balton JS, Robinson W: Bilateral lesion of the occipital lobes with retention of macular as distinct from panoramic vision; *Brain* 1915; 38: 447-72.
8. Zappia RJ, Enoch JM, Stamper R, Winkelman JZ, Gay AJ: The Riddoch phenomenon revealed in non-occipital lobe lesions; *Brit J Ophthal* 1971; 55: 416-20.
9. Jefferson A, Sheldon P: Transtentorial herniation of the brain as revealed by the displacement of arteries; *Acta radiol* 1956; 46: 480-98.
10. Lindenburg R: Compression of brain arteries as pathogenic factor for tissue necrosis and their areas of predilection; *J Neuropath and Exper Neurol* 1955; 14: 223-43.
11. Adam R and Vander E: Vascular diseases of the brain: *Annual review of medicine* 1953; 4: 213-52.
12. Byrom FB: Pathogenesis of hypertensive encephalopathy and its relation to malignant phase of hypertension; experimental evidence from hypertensive rat; *Lancet* 1954; 2: 201-11.
13. Lassen MA, Agnoli A: The upper limit of autoregulation of cerebral blood flow – on the pathogenesis of hypertensive encephalopathy; *Scand J Cl and Lab Invest* 1972; 30: 113-5.
14. Hoyt WF, Walsh FB: Cortical Blindness with partial recovery following acute cerebral anoxia from cardiac arrest. *AMA Archives of Ophthalmology* 1958; 60: 1061-9.