

## Case report

# Dorsal thalamic haemorrhage complicating polyarteritis nodosa : a clinico-pathologic case report

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### Abstract

*Intracerebral haemorrhage is a rare complication of polyarteritis nodosa (PAN). We present the clinico-pathologic findings of a 62 year-old patient with systemic involvement by PAN, who developed temporary decrease of consciousness and a mild left hemisyndrome due to a small right dorsal thalamic haemorrhage. No clear cause for the haemorrhage was found on postmortem examination.*

**Key words :** Polyarteritis nodosa ; dorsal thalamic haemorrhage ; cerebral vessel pathology.

### Introduction

Central nervous system manifestations of polyarteritis nodosa (PAN) are rare and mostly appear in the late course of the disease (Ford and Siekert, 1965 ; Cohen *et al.*, 1980 ; Moore and Fauci, 1981 ; Reichhart *et al.*, 2000). Stroke occurs in approximately half of these cases of which one third are haemorrhagic (Ford and Siekert, 1965 ; Reichhart *et al.*, 2000).

In large thalamic haemorrhage, with ventricular extension, of any cause impaired consciousness, hemiparesis, pupillary abnormalities and vertical gaze impairment are the main clinical features, frequently leading to death (Weisberg, 1986). Small haemorrhages can, similarly to infarcts (Bogousslavsky *et al.*, 1988), be subdivided in anterolateral, posterolateral, medial and dorsal types (Kawahara *et al.*, 1986 ; Steinke *et al.*, 1992 ; Kumral *et al.*, 1995), of which the posterolateral one is the most frequent and the dorsal one rare (Kawahara *et al.*, 1986 ; Kumral *et al.*, 1995).

We describe the first clinico-pathologic case of dorsal thalamic haemorrhage complicating PAN.

### Case Report

This 62 year-old man was admitted to the department of internal medicine on the 6th of november 1971 because of severe anemia, heart failure and left pleurorrhea.

The prior history was unremarkable until january 1971, when he developed a left purulent otitis treated by paracentesis. Afterwards he complained of left hearing loss and hemicranial headache.

On the 25th of april 1971 he suddenly developed hoarseness and swallowing difficulties after a flue-like episode. There was a loss of 7 kg bodyweight. A left vagus nerve lesion was diagnosed. Blood analyses at that time revealed a sedimentation rate of 64 mm for the first hour, normal red bloodcell count and 7900 leucocytes/ml. SGOT was 480 IU/l and SGPT 740 IU/ml with a total bilirubin of 4.8 mg/dl. Protein electrophoresis, showed an increased gamma fraction. The probable diagnosis of viral hepatitis was made. No cause for the nervus vagus lesion could be retained at that time.

During the last admission the patient also developed abdominal pain with melena and had signs of a progressive renal insufficiency with haematuria and proteinuria. Bloodpressure was in normal range. Laboratory data revealed a sedimentation rate of 122 mm for the first hour, 2.000.000 Rbc/ml, platelets of 183.212/ml, total protein of 4,9 g/dl with 33% albumine and 31% gammaglobulin, SGOT of 840 IU/l, SGPT of 1015 IU/l, total bilirubin of 5.1 mg/dl, creatinine of 2.8 mg% and urea nitric acid of 150 mg%.

The diagnosis of PAN was made on muscle biopsy, showing necrotizing angiitis with thrombosis of medium-sized arteries (Fig. 1). The patient was afterwards treated with corticosteroids.

Three weeks after admission he became suddenly lethargic. On neurologic examination a moderate left inferior facial paresis was found without loss of strength in arm and leg. Speech was not disturbed. Tendon reflexes were brisk and symmetrical with a left Babinski sign.

Sensation was preserved and ocular movements were normal. He regained normal consciousness after one day and the focal neurological signs disappeared after two days.

His general condition gradually deteriorated and he died on the 11th of december 1971, due to his cardiac failure.

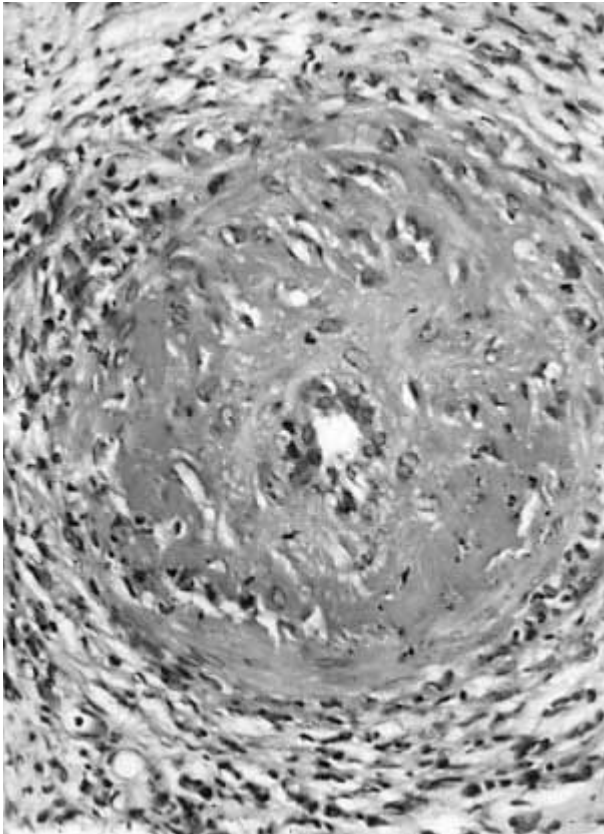


FIG. 1. — Necrotizing angitis of a medium-sized artery in the left quadriceps muscle biopsy (HE, 125 $\times$ ).

General necropsy revealed fibrinous pericarditis, bronchopneumonia with pleuritis, iron deposits in liver and spleen and segmental hyalinosis of both kidneys. Widespread arteritis lesions were found in the kidneys, liver, muscles and the left vagus nerve in the neck. Arterioles, capillaries and veins were not affected. No signs of systemic bleedings were observed.

The postmortem brain showed no obvious external lesions and the arteries displayed no atherosclerosis. On coronal sections of the cerebral hemispheres a lytic haematoma of approximately 1 cm<sup>3</sup> was found involving the dorsal part of the lateral posterior, lateral dorsal and dorsal medial nuclei of the right thalamus (Fig. 2). No other lesions were detected in the brain.

Histological examination on serial sections of the lesion were performed with Haematoxylin-eosin, Masson's trichrome and Phosphotungstic acid haematoxylin. The haematoma had disrupted the ependyma and bulged into the lateral ventricle. It was mainly composed of lytic red blood cells. At the border a large number of macrophages and siderophages were seen. Dilated capillaries and large reactive astrocytes were present. Arteries and veins had a normal appearance. No inflammatory, aneurysmal or fibrotic changes of the vesselwalls were observed (Fig. 3).

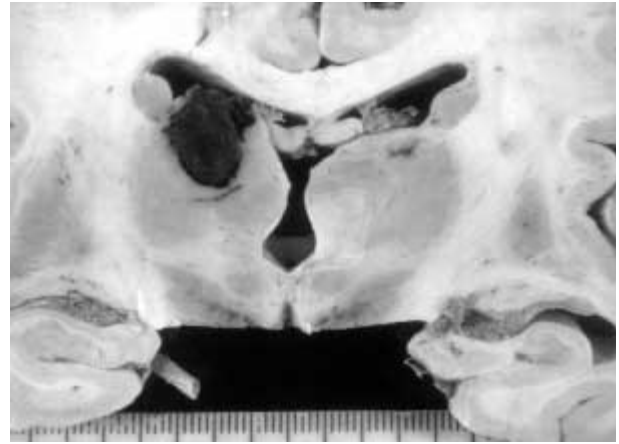


FIG. 2. — Coronal section of the cerebral hemispheres, showing a right thalamic haemorrhage involving the dorsal part of the lateral posterior, lateral dorsal and dorsal medial nuclei.

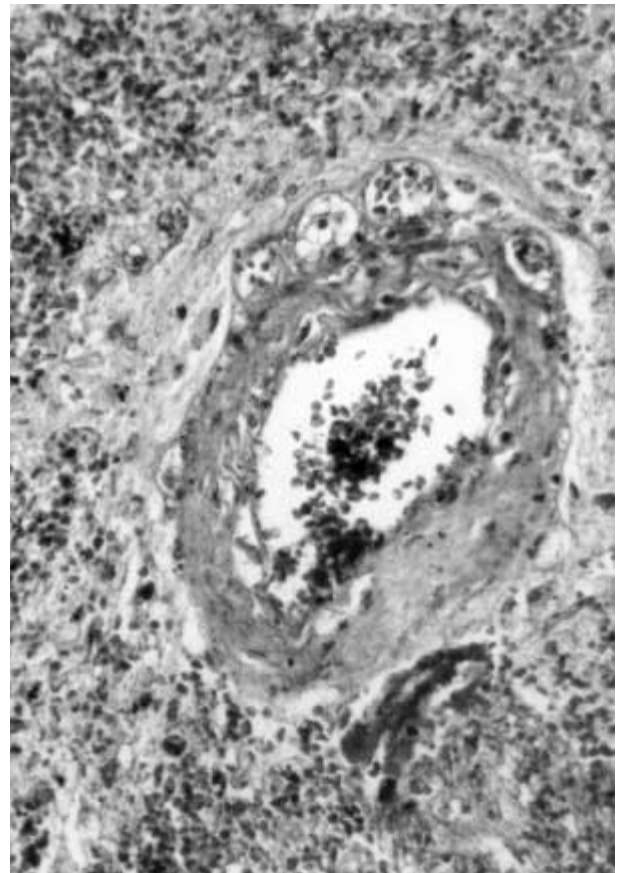


FIG. 3. — Normal appearance of a small artery at the border of the thalamic haemorrhage (Masson's trichrome, 125 $\times$ ).

The neuropathologic findings were consistent with a two week-old small dorsal haemorrhage of the right thalamus.

### Discussion

This clinico-pathologic case report is remarkable by the association of a small dorsal thalamic haemorrhage and PAN.

The patient fulfilled the criteria of PAN of the American College of Rheumatology (Lightfoot *et al.*, 1990) but not from the Chapel Hill Consensus Conference (Jenette *et al.*, 1994), who would have classified this case as microscopic polyangiitis (MPA), due to the presence of glomerulonephritis. However, the differentiation between MPA and PAN remains difficult and there is overlap between both conditions (Watts *et al.*, 1996). Arguments for a classical PAN are that biopsy and autopsy revealed only necrotizing inflammation of medium-sized or small arteries, without involvement of arterioles, capillaries and veins. Also the fact that the patient had a prior history of hepatitis points to an association with PAN (Guillevin *et al.*, 1995). In the early seventies the association of hepatitis B virus and PAN was just described (Gocke *et al.*, 1970).

Temporary decreased consciousness and mild transient motor deficit were the main clinical features of the small dorsal thalamic haemorrhage in our case (Kawahara *et al.*, 1986 ; Kumral *et al.*, 1995) As the lesion was restricted to the dorsal part of the lateral posterior, lateral dorsal and dorsal medial nuclei in the territory of the lateral posterior choroïdal artery (Plets *et al.*, 1970) no pupillary abnormalities or gaze impairment were observed. Those ocular disturbances are found in more extended lesions in the vascular territory of the posterior choroïdal artery (Devic *et al.*, 1964).

No vasculitis lesions or microaneurysms were found in the brain of our patient, despite still active lesions in other organs. Also no fibrotic thickening as remnant of healed vasculitis lesions of the deep perforating arteries was observed. Some atherosclerotic mechanism, induced by corticosteroids alone or combined with cyclophosphamide or azathioprine has been suggested rather than the vasculitic arterial changes to explain the late occurrence of strokes in treated patients with PAN (Cohen *et al.*, 1980 ; Reichhart *et al.*, 2000 ; Reichhart and Bogousslavsky, 2001). However, this case report illustrates that the occurrence of a cerebral haemorrhage in PAN may not be related to the underlying disease, nor to its treatment.

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