Supratentorial haemangioblastoma with polycythaemia

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SYNOPSIS A case is reported of a 21 year old man with polycythaemia and a vascular frontal lobe tumour that was histologically compatible with the diagnosis of haemangioblastoma. On review of the English literature no case could be found of polycythaemia associated with supratentorial haemangioblastoma.

It is well known that polycythaemia may complicate infratentorial haemangioblastomas. On review of the English literature no case could be found of polycythaemia associated with supratentorial haemangioblastoma. We report such a case.

CASE REPORT

W.S., a 21 year old West Indian man, was admitted in February 1974. One month previously he had developed diplopia on left lateral gaze and frontal headaches associated with vomiting. Abnormal physical signs were limited to the central nervous system. He was well orientated. His blood pressure was 120/80 mmHg. Fundoscopy revealed papilloedema. There was a left 6th cranial nerve palsy. There were no abnormalities apparent in the other cranial nerves. Power and sensation were normal in the limbs. The tendon reflexes were normal and plantar responses flexor.

Investigations revealed a haemoglobin of 19.3 g/dl haematocrit of 59%, white cell count 3 300 cells/mm³ with a normal differential and a platelet count of 180 000 cells/mm³. Blood urea, creatinine, electrolytes, liver function tests, serum transaminase, and a mid-stream specimen of urine were all normal. A ⁹⁹Tc brain scan showed an area of increased uptake of isotope in the right mid-frontal region. Right carotid angiography showed a vascular tumour with a pronounced right to left shift of the midline (Fig. 1). A chest radiograph and an intravenous urogram were within normal limits.

Further investigations revealed a blood P_{O₂} of 110 mmHg, a P_{CO₂} of 42 mmHg, a pH of 7.37 and

HCO₃ concentration of 23 mmol/l. An oxygen dissociation curve was normal. A red cell mass estimation using ⁵¹Cr labelled red cells was 65.5 ml/kg (normal range 28 ± 8 ml/kg).

FIG. 1 Right carotid angiogram showing a vascular frontal lobe tumour.

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At operation, a vascular tumour measuring $4.3 \times 3.7 \times 2.6$ cm was removed from the right frontal lobe. There was no apparent communication with the meninges. The histological appearances were of a highly vascular angioblastic tumour without a capsule. The tumour was composed of a fine vascular network of capillary channels lined by endothelial cells with no demonstrable basement membrane on PAS staining. These channels dilated in places to form cavernous lacunar spaces. The vessel network was outlined by fine reticulin fibres which passed between the stromal cells. The stroma consisted of numerous clear vacuolated foamy xanthomatous cells. No mitotic figures were seen. The cells were rich in lipid droplets. The absence of interstitial nervous tissue and negative Van Gieson

stain served to exclude a cerebrovascular malformation (Fig. 2).

The postoperative course was uneventful. After operation the patient's haemoglobin fell gradually over the ensuing 16 days to 12.1 g/dl, remaining at a similar level on subsequent follow-up.

DISCUSSION

The present report describes a hitherto undescribed association between supratentorial haemangioblastoma and polycythaemia confirmed by red cell mass estimation. It is well documented that infratentorial haemangioblastomas are associated with polycythaemia and in one series the incidence was as high as 49% (Palmer, 1972).

The concept of a humoral stimulating factor as the fundamental stimulus for red cell production was first proposed by Carnot and Deflandre (1906). Waldman *et al.* (1961) presented experimental evidence for the production of an erythropoiesis stimulating factor by a cerebellar haemangioblastoma. The erythropoiesis stimulating factor has been found to be similar when derived from renal carcinoma, cerebellar haemangioblastoma cyst fluid and urine from anaemic patients (Rosse *et al.*, 1963).

Up to 1962 Morello and Bianchi (1963), in their review of the literature, could accept only seven histologically proven cases of supratentorial haemangioblastomas and added two cases of their own. At least 12 further instances of supratentorial haemangioblastomas are mentioned in the literature (Stein *et al.*, 1960; Hoff and Ray, 1968; Palmer, 1972) but, unfortunately, no histological or angiographic findings are reported in the majority and in no case is there mention of associated polycythaemia.

The morphological and angiographic appearances of the tumour in our report fulfil the criteria for the diagnosis of haemangioblastoma set out by Morello and Bianchi (1963). The histological features in our patient are also consistent with the diagnosis of angioblastic meningioma, although the absence of mitoses and apparent connection with the meninges favours the diagnosis of haemangioblastoma. There is, however, so close a morphological resemblance between these two types of tumour that the grounds for separating them are unsatisfactory (Russell and Rubinstein, 1971). We

FIG. 2 Photomicrograph of section of the tumour. H and E, $\times 250$.

believe that the occurrence of polycythaemia in our patient is further evidence for the existence of supratentorial haemangioblastomas.

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