SICKLE-CELL TRAIT

A Report of a Postoperative Neurological Complication

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SUMMARY

It is widely assumed that individuals with sickle-cell trait present no anaesthetic problems with the result that sickle-cell screening is not performed in Negro patients unless they are anaemic. This complacent attitude is potentially dangerous and may be the cause of occasional postoperative morbidity or mortality. This report is of a 12-year-old Negro patient with sickle-cell trait who was anaesthetized for an elective eye operation. After the operation, he developed signs of a haemorrhagic cerebral infarct with aphasia and hemiplegia.

The problems and risks in anaesthetizing a patient with sickle-cell disease are well documented (Gilbertson, 1967). Some authors have assumed that the sickle-cell trait (AS haemoglobin) presents no special problems (Searle, 1973; Gilbertson, 1967; Oduntan and Isaacs, 1971), while others suggest that the precautions taken with the SS disease patients should be taken with the “carrier” also (Howells et al., 1972; Schenk, 1964). The carrier may be at risk because it is likely that many patients who come for operation with a sickle-cell “trait” may remain undiagnosed because many physicians believe that it is unnecessary to test for HbS if the haemoglobin concentration exceeds 12 g/100 ml in Negro patients despite evidence that this is not so (Diggs and Diggs, 1972; Konotey-Ahulu, 1967; Bauer and Fisher, 1943).

CASE REPORT

A 12-year-old male Negro was admitted for cryotherapy to both eyes for lattice degeneration. The patient had developed secondary glaucoma followed by optic atrophy in the left eye after a hyphema resulting from an injury. Examination before operation revealed no abnormality except for a systolic murmur. His haemoglobin concentration was 12.3 g/100 ml and the haematocrit was 36.2%.

On the day before surgery, 10% phenylephrine and 1% cyclopentolate HCl (cyclogyl HC1) one drop in each eye, four times daily, were prescribed.

Premedication consisting of diazepam 5 mg and atropine 0.4 mg, was given 1 hour before surgery. After preoxygenation, anaesthesia was induced with thiopentone 175 mg, followed by suxamethonium 60 mg mixed with atropine 0.8 mg and an orotracheal tube was inserted without difficulty. Anaesthesia was maintained with 70% nitrous oxide in oxygen; tubocurarine 25 mg was given i.v. At the end of the procedure the neuromuscular blockade was antagonized with a mixture of neostigmine 3.5 mg and atropine 0.8 mg given i.v. When adequate spontaneous ventilation had returned, the endotracheal tube was removed and the patient was taken to the recovery room. No oxygen was administered during the transfer.

On arrival in the recovery room, oxygen was given from a mask. The patient remained unconscious (drowsy) and was given doxapram 50 mg to aid awakening. He responded well and appeared more awake. At this time he was seen to be able to move all his limbs although he did not speak. Eight hours after the end of surgery, the patient was transferred to the intensive care unit and on examination he exhibited a right hemiparesis and aphasia. On the 1st postoperative day the e.e.g. showed a slow wave focus in the left frontotemporal region. The brain scan was negative. A cardiologist examined the patient and was able to exclude the possibility of a paradoxical embolus. The patient’s aunt was questioned about sickle-cell disease and stated that indeed the child had a sickle-cell “trait”. The electroencephalogram pattern showed 37.2% HbS (in contrast to the normal 100% HbA).

On the 2nd postoperative day he was transferred to the ward in essentially the same condition. He developed nuchal rigidity and a lumbar puncture was performed. The cerebrospinal fluid was grossly bloodstained and xanthochromic. The opening and closing pressures were 590 and 270 mm H2O respectively. A left carotid arteriogram performed on the 7th postoperative day revealed: (1) straightening of the intracerebral vessels with a shift of the left anterior cerebral artery to the right and indicating cerebral oedema; (2) vasculitis of intracerebral vessels and; (3) a questionable decreased filling of the anterior portion of the sagittal sinus. A diagnosis of superior sagittal sinus thrombosis was made. After 2 weeks of physical and speech therapy, the patient made a good recovery and was discharged with only minimal weakness of the right upper arm.

DISCUSSION

The accepted management of the patient with sickle-cell disease includes the avoidance of hypoxia, cold and acidosis. However, a major problem is circulatory stasis which may cause gross desaturation in...
the venous blood and precipitation of the insoluble HbS with consequent sickling of the red cells. This causes microthrombi, sludging, and emboli which in turn cause the clinical features of sickle-cell diseases (haemolysis, aplasia, infarctions). The anaesthetic management of a patient with sickle-cell disease (homozygous SS, or the abnormal haemoglobins, SC disease, Sickle-Thalassemia) has been discussed fully in the literature. However, it has been stated that heterozygous patients do not present a problem (Watson-Williams, 1967; Holzman et al., 1969) and that heterozygous (a mixture of normal HbA with abnormal HbS) or “AS states give almost complete protection from lytic crises” (Ball, 1967). This assumption on the part of many physicians is perhaps responsible for the common practice of not testing for sickle haemoglobin unless the Hb concentration is reduced. Thus, a large proportion of these patients come to operation without the realization that they are heterozygous. McCormick (1961) was able to document post-mortem evidence of ante-mortem visceral infarcts in 22 out of 135 patients with proven sickle-cell “trait”. Konotey-Ahulu (1967) has stressed that sickling is possible in heterozygous patients and that its severity is dependent upon the actual amount of HbS rather than the ratio of the haemoglobin A and S. On the other hand Searle (1973) in a recent review of anaesthesia and sickle-cell states, concludes that patients with the sickle-cell trait do not incur an increased risk from anaesthesia. However, he notes that in one out of three deaths in patients with sickle-cell trait described in the literature, anaesthesia could have been the cause and that death could possibly have been prevented had the presence of the sickle-cell trait been known.

The percentage of sickle haemoglobin in the sickle-cell trait can vary from 25 to 45% (Neel, Wells and Itano, 1951). In-vitro studies suggest that in carriers who have a high concentration of HbS, the risk of sickling is not much less than in patients with sickle-cell disease (Howells and Huntsman, 1973).

It is at tissue level that desaturation occurs but no monitors are presently available to measure tissue oxygen tension. Oxygen saturation is the important index but, in current practice, the partial pressure of oxygen rather than oxygen saturation is measured. We believe that while it is desirable to monitor arterial blood gases, the measurement of central venous Po$_2$ is a more rational variable as it is in the venous blood that sickling will occur. It should be noted that in homozygous disease, even when haemoglobin is 100% saturated with oxygen, there is a trace of sickling; at 65% saturation and at 50% oxygen saturation, there is 75 and 100% sickling respectively, while in heterozygous disease with 40% HbS, sickling starts at 40% saturation (Dugdale, 1972). Thus the critical Po$_2$ is 30 mm Hg for SS and 20–30 mm Hg for AS (Howells et al., 1972). In the presence of any circulatory stasis or reduced cardiac output, oxygen extraction may be high and venous Po$_2$ may be reduced to dangerously low values in spite of a normal arterial oxygen tension.

REFERENCES


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