

Angioid streaks in beta thalassaemia minor

FRANK P KINSELLA AND DAVID J MOONEY

From the Research Unit of the Royal Victoria Eye and Ear Hospital, Dublin

SUMMARY We report what we believe to be the first recorded case of angioid streaks in a patient with beta thalassaemia minor. The occurrence of angioid streaks in a patient with a relatively normal iron balance and only very mild haemolysis may be explained by the combination of pregnancy with associated multiple transfusions in a myopic patient where an inherent defect in Bruch's membrane may exist.

Angioid streaks, representing breaks in Bruch's membrane, with the typical ophthalmoscopic appearance of reddish brown bands radiating outwards from the optic disc, were first described in 1889 by Doyne.¹ Subsequently they have been described in a number of conditions, the most notable being pseudoxanthoma elasticum,² Paget's disease of bone,³ and certain haemoglobinopathies, particularly sickle cell disease.^{4,5}

We report a patient with beta thalassaemia minor who was found to have angioid streaks in both fundi.

Case report

In July 1982 a 42-year-old woman of Italian extraction had beta thalassaemia minor diagnosed by a haematological unit, having presented with a long history of mild anaemia with more severe exacerbations needing multiple blood transfusions during each of three preceding pregnancies (the last one being in 1973). Apart from symptoms of mild anaemia she was otherwise asymptomatic, and general physical examination was normal. There was no family history of thalassaemia.

In February 1986 she presented with a three-month history of blurred vision in her left eye. Examination revealed a visual acuity of 6/60 in the right eye and 2/60 in the left eye, improving to 6/18 and 6/36 respectively with -3.50 dioptré sphere correction in each eye. Angioid streaks were noted in both fundi, radiating outwards from the optic discs and passing through both maculae (Figs. 1 and 2).

Laboratory investigations were as follows: Hb 11.5 g/dl, mean cell volume 61 μm^3 reticulocyte count 3%

(normal less than 2%). The following were normal: serum Fe, total iron binding capacity, serum folic acid, serum B₁₂, erythrocyte folic acid, haptoglobins, and thyroxine. Hb electrophoresis showed Hb A=94.7%, Hb A2=5.3% (normal less than or equal to 2%). A blood film showed occasional microcytic hypochromic red blood cells in addition to rod, pear, and target cells; leucocytes and platelets were normal.

A skin biopsy showed no evidence of pseudoxanthoma elasticum, nor was there any evidence, clinical, biochemical, or radiological, of Paget's disease of bone.

Over a four-year follow-up the haemoglobin remained fairly constant (between 10 to 11 g/dl) with only a very mild haemolysis (as shown by haematological indices) while the patient received no treatment.

Discussion

Angioid streaks have been described in several haemoglobinopathies, including homozygous sickle cell anaemia,⁴ sickle cell trait,⁶ sickle cell haemoglobin C disease,⁷ and sickle cell thalassaemia,^{8,9} but until recently their occurrence in the thalassaemias has not been described (Table 1). In 1979 angioid streaks were noted in a patient with haemoglobin H

Table 1 *Angioid streaks in the thalassaemias*

Type	Reported cases	Authors
Haemoglobin H disease	1	Daneshmend ¹⁰
Beta thalassaemia major	1	Gibson <i>et al.</i> ¹¹
	1	Theodossiadi <i>et al.</i> ¹²
Beta thalassaemia intermedia	1	Singerman ¹³
Beta thalassaemia minor (our report)		

Correspondence to Mr David Mooney, Royal Victoria Eye and Ear Hospital, Adelaide Road, Dublin 2, Ireland.

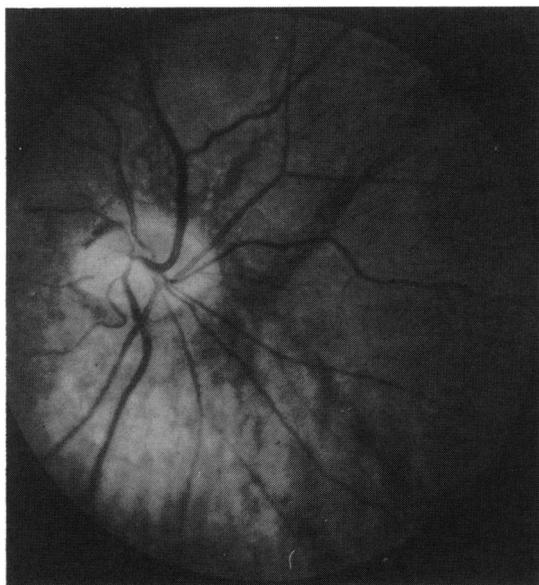


Fig. 1 Fundus photograph of right optic disc showing angiod streaks.

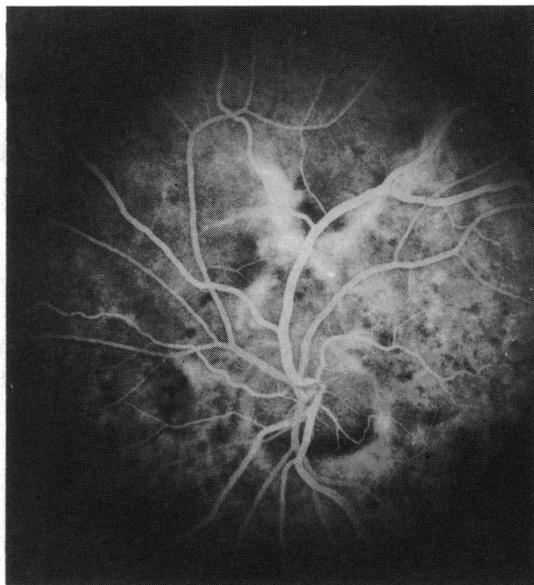


Fig. 2 Fluorescein angiogram of left fundus demonstrating angiod streaks above optic disc.

disease¹⁰ (an alpha thalassaemia), and their occurrence in two cases of beta thalassaemia major^{11,12} and in one of beta thalassaemia intermedia¹³ has since been reported. We now report a case of beta thalassaemia minor in which angiod streaks were observed.

Unlike the earlier reported cases of beta thalassaemia major and intermedia, which had iron overload syndromes (from serial blood transfusions for anaemia), our patient was in a normal or slightly negative state of iron balance, with only very low grade haemolysis.

The pathogenesis of angiod streaks in the haemoglobinopathies is uncertain and may be due to either inherent defects or chronic iron deposition in Bruch's membrane—though this latter explanation has recently been disputed.¹⁴ A possible explanation for the occurrence of angiod streaks in our patient with a relatively normal iron status and very mild haemolysis is that iron deposition in Bruch's membrane occurred during the repeated pregnancy-associated transfusions in a myopic patient in whom a defect in Bruch's membrane may already have existed.

References

- 1 Doyne RW. Choroidal and retinal changes—the results of blows to the eyes. *Trans Ophthalmol Soc UK* 1889; **9**: 128–31.

- 2 Goodman RM, Smith EW, Paton D, *et al.* Pseudoxanthoma elasticum: a clinical and histopathological study. *Medicine* 1963; **42**: 297–332.
- 3 Terry TL. Angiod streaks and osteitis deformans. *Trans Am Ophthalmol Soc* 1934; **32**: 555–73.
- 4 Goodman G, von Sallmann L, Holland MG. Ocular manifestations of sickle cell disease. *Arch Ophthalmol* 1957; **58**: 655–82.
- 5 Condon PI, Serjeant GR. Ocular findings in elderly cases of homozygous sickle cell disease in Jamaica. *Br J Ophthalmol* 1976; **60**: 361–4.
- 6 Gerde LS. Angiod streaks in sickle cell trait haemoglobinopathy. *Am J Ophthalmol* 1974; **77**: 462–4.
- 7 Condon PI, Serjeant GR. Ocular findings in haemoglobin SC disease in Jamaica. *Am J Ophthalmol* 1972; **74**: 921–31.
- 8 Goldberg MF, Charache S, Acacio I. Ophthalmologic manifestations of sickle cell thalassaemia. *Arch Intern Med* 1971; **128**: 33–9.
- 9 Condon PI, Serjeant GR. Ocular findings in sickle cell thalassaemia in Jamaica. *Am J Ophthalmol* 1972; **74**: 1105–9.
- 10 Daneshmend TK. Ocular findings in a case of haemoglobin H disease. *Br J Ophthalmol* 1979; **63**: 842–4.
- 11 Gibson JM, Raychaudhuri P, Rosenthal AR. Angiod streaks in a case of beta thalassaemia major. *Br J Ophthalmol* 1983; **67**: 29–31.
- 12 Theodossiadis G, Ladas I, Koutsandrea C, Damanakis A, Petroustos G. Thalassaemia et neovaisseaux sous-rétiniens maculaires. *J Fr Ophthalmol* 1984; **7**: 115–8.
- 13 Singerman LJ. Angiod streaks in thalassaemia major. *Br J Ophthalmol* 1983; **67**: 558.
- 14 Hamilton AM, Pope FM, Condon PI, *et al.* Angiod streaks in Jamaican patients with homozygous sickle cell disease. *Br J Ophthalmol* 1981; **65**: 341–7.

Accepted for publication 20 February 1987.