HEREDITARY TELANGIECTASIA IN A ZULU FAMILY

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In 1953 Theron[1] reported the first case of hereditary telangiectasia in an African, a Venda man whose father was also found to have the condition. Three Negro cases have been reported in America[2] but no further cases in the African seem to have been described.

The following is a further case which presented in a Bantu, in which a fairly extensive family history could be elicited.

Case History

C.N., Zulu female, 49 years. Frequent nose bleeds for some years. Admitted to King Edward VIII Hospital 1958; diagnosed hereditary telangiectasia, nasal mucosa cauterized, failed to return for follow-up. Bleeding resumed soon afterwards. Admitted to Clairwood Hospital 1962 because of more severe bleed.

Findings. CCF, BP 120/70 mm.Hg, Hb 6·7 G/100 ml. Numerous small telangiectases in nose, palate, tongue, lips and finger. Of these only the first and last appear to have bled.

CCF responded to treatment with bed rest and blood transfusion, etc., but nose bleeds recurred repeatedly.

With the help of the Health Educator, the family history was obtained covering 5 generations, and 5 members of the family who lived in and around Durban were examined. A family tree was constructed from details (Fig. 1).

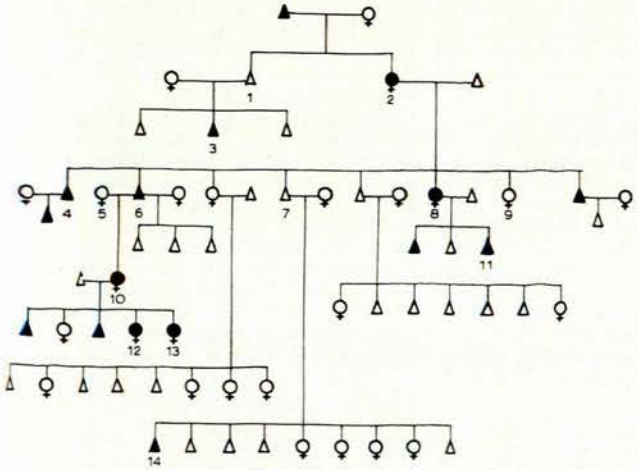

Fig. 1. Family tree of the Zulu family with hereditary telangiectasia.

Family History

Particularly strong evidence was obtained in the following:

No. 2. The patient's own father died from repeated haemorrhages.

No. 4. The eldest brother was known to have had melaena stools and has a son who has nose bleeds.

No. 5. A sister-in-law was examined. Neither telangiectasia, nor a history of bleeding were found.

No. 6. A brother, the husband of No. 5, is a bleeder and was treated at a hospital on the Rand. He married a second time; no children of the second marriage appear to be bleeders.

No. 8. The patient. Two of her 3 sons are nose bleeders but live out of Durban and could not be examined.

No. 9. A sister from Pietermaritzburg was examined. No telangiectasia and no history of bleeding were obtained.

No. 10. In a niece, the daughter of No. 5 and No. 6, typical nasal and oral telangiectases were found; she bleeds frequently.

Nos. 12 and 13. Daughters of No. 10, aged 12 and 2 years, were examined. No. 12 has typical telangiectases but No. 13 showed only one on lip. No history of bleeding.

No. 14. This case is interesting because of being the only bleeder among 9 siblings, whereas a ratio of 1 : 1 is expected.

DISCUSSION

The condition is said to be carried by an autosomal dominant gene, with which this family tree is compatible if cases No. 1 and No. 7 are assumed to be affected too. This is possible because affected persons do not necessarily always bleed and the history is not reliable if there is only mild bleeding.

Treatment was ineffective in our case. This corresponds to most people's experience. The following methods have been advocated but give only temporary relief:

Bleeding can be controlled by the local application of fibrin foam, 'oxycel' or similar preparation, but recurrence is inevitable. Iron often does not control the anaemia and repeated blood transfusions may become necessary, as in this patient.

The most hopeful approach so far has been that of Saunders.[3] He reports the results of skin grafting the nasal septum in 15 cases. It is not possible to cover all the affected mucous membrane, but since the nasal septum is the most frequent source of haemorrhage and most subject to trauma, if this can be covered by skin, bleeding becomes less frequent and less severe. If necessary, further grafts may be attempted at a later stage.

SUMMARY

A Zulu patient suffering from hereditary telangiectasia is described. Elaboration of the family tree confirmed the expected distribution of affected individuals.

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