

- (5) *Idem.*—*Practitioner*, December, 1916.
 - (6) *Idem.*—*Ibid.*, July, 1917.
 - (7) *Idem.*—*Brit. Journ. Derm. and Syph.*, July–September, 1917.
 - (8) *Idem.*—*Practitioner*, May, 1918.
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A NOTE ON MULTIPLE SMALL ANGIOMATA: ANGIO-KERATOMA AND MULTIPLE TELANGIECTASES.

By H. G. ADAMSON, M.D., F.R.C.P.

At a meeting of the Dermatological Section of the Royal Society of Medicine, on March 21st, Dr. Sibley* showed the case of a youth "for diagnosis," and a discussion arose as to whether this case was an example of multiple telangiectases, such as have been described by Osler and Parkes Weber, or of multiple purpuric lesions.

A comparison of Dr. Sibley's case with some other cases which have been recorded will, I think, make it clear that it belongs neither to the group of "multiple telangiectases of the skin and mucous membranes" associated with "a family form of recurring epistaxis" of Osler and Parkes Weber, nor is it an example of purpuric lesions; but that it is what has been described as the "aberrant form" of the angiokeratoma of Dubreuilh, Cottle, Mibelli, and Pringle.

In the *British Journal of Dermatology* for April, 1898, the late Dr. William Anderson published a case which is, in every respect, similar to Dr. Sibley's case, and in which the peculiarity of the eruption "rested in its widespread distribution, in the almost complete immunity of hands and feet, and in the absence of any tendency to chilblains or local embarrassment of circulation." The diagram of the distribution of the small angiomata in Anderson's case and that of the histological features would serve equally well for Dr. Sibley's case. As pointed out by Anderson (and, previously, by Dubreuilh), the warty condition found in cases where this complaint is limited to the hands and feet appears to depend upon purely local causes, is a secondary and accidental feature, and is slight or altogether absent when the affection occurs on other parts of the body.

Similar examples of this aberrant form of angiokeratoma have been recorded by Dubreuilh, by Fordyce, and by Sutton, in which

* See p. 109.

the lesions had unusual distributions on the trunk, the limbs, and the scrotum. The coloured plates showing the lesions on the scrotum in Fordyce's case, and in a similar case by Sutton, recall exactly the appearances of the scrotum in Sibley's case.

The peculiarity of the histological appearances, as shown in all the published drawings of sections of "angiokeratoma," and in Dr. Sibley's sections, is that the little blood-cysts are actually in the epidermis, and this is explained by all observers as resulting from a dilatation of the capillaries in the papillæ, which, pushing up into the epidermis, get cut off by lateral down-growth of the interpapillary processes

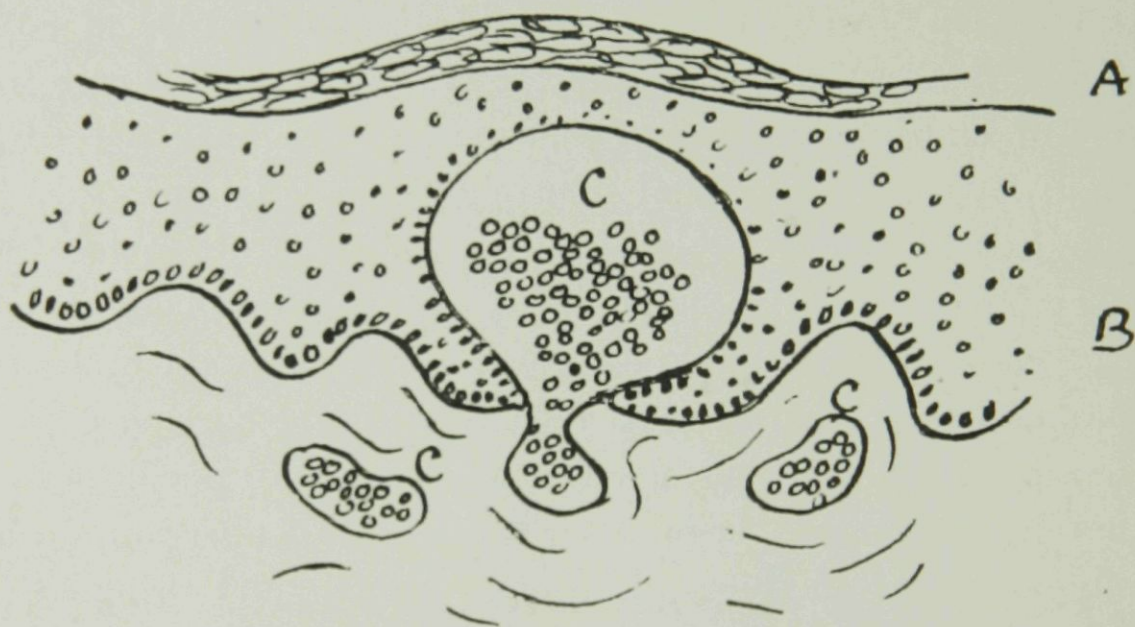


Diagram showing how the dilated papillary vessels become cut off by down-growth of the interpapillary processes to form blood-cysts in the epidermis. A. Horny layer of epidermis. B. Stratum Malpighii. C. Angiectases occupied by blood-vessels.

(see diagram). The early stages of this process are well shown in Wisniewski's drawings in the *Archiv für Dermatologie und Syphilis*, Bd. xlv, 1898, and there is a good coloured plate in Joseph and van Deventer's *Atlas of Cutaneous Morbid Histology* (Pl. II, fig. 4). Many of these blood-cysts eventually become cut off entirely from the derma, and thus is explained why it is sometimes impossible to express their contents, so that they simulate purpuric lesions. Sometimes the contents of these little blood-cysts become converted into a granular detritus, and sometimes they come to the surface and become exfoliated. All regard the verrucosities as secondary and accidental.

It should be noted that the angiomas in these cases of "aberrant angiokeratoma" are not of the tufted or spider-like type known as

"Nævus araneus," which may occur singly or multiple. Clinically, they bear a closer resemblance to the "seed-like" angiomata, known as "De Morgan's spots," which occur on the trunk in elderly persons, and, according to some observers, particularly in those who are the subject of malignant tumours; although, from the ætiological side, they ought probably to be distinguished from these.

They ought, perhaps, to be distinguished too from the small angiomata which may occur over a limited area of the trunk or limbs, as in the case of "bilateral telangiectases" described by Colcott Fox, and in a similar unpublished case of the writer's with unilateral distribution, which are possibly examples of segmental nævus.

They differ also in many respects from the cases of "multiple telangiectases" described by Osler and Parkes Weber, in which the angiomata, for the most part of the nodular type, but sometimes stellate, occurred particularly upon the cheeks, lips, and ears, and on the mucous membranes of the nose and buccal cavity, and were associated with a family form of recurring epistaxis.

A very full record of the published cases of multiple telangiectases or angiomata, a useful commentary on those cases, and a careful endeavour to sift them into their proper order was made by the late Dr. T. Colcott Fox in a paper in this Journal in 1908 while discussing the position of his case of "Bilateral Telangiectases of the Trunk, with a History of Marked Epistaxis in Childhood and Recent Rectal Hæmorrhage."

But the proper classification of these various forms of multiple angiomata or telangiectases is obviously a difficult matter until we know something more of their causation and ætiological relationships. It is not clear that the type of lesions form a sufficient basis for their classification, since the various types—the punctiform, the raised nodular, and the spider-like or stellate—may sometimes occur together in the same case. There does, however, seem to be a distinction between the lesions found in the cases of the angiokeratoma group, whether true or aberrant forms, and those of other groups, in that the blood-cysts in the angiokeratoma are often situated in the epidermis itself, while in the other forms the dilated blood-vessels or cyst-like formations are beneath the epidermis. In the most recently published case of the Osler type of telangiectasis, namely, that recorded by Dr. Norman Paul in the *British Journal of Dermatology*

for January–March, 1918, the photo-micrograph of a section shows the blood-cysts in the corium beneath the epidermis, but none actually in the epidermis. This is also the case in a drawing of a section made by myself of Dr. Colcott Fox's case of bilateral telangiectasis published in this Journal in 1908 (p. 148).

Without attempting a complete or final classification of these angiomas upon ætiological grounds, about which we know at present so little, it may be useful, perhaps, to divide them into clinical groups as follows :

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| Nævi. | { | <ol style="list-style-type: none"> 1. Nævus araneus, either single or multiple.
(A case of multiple telangiectases of the Nævus araneus type described by myself in the <i>St. Bartholomew's Hospital Reports</i> in 1909, and again in this Journal by Dr. Sibley in 1914, possibly belongs to this group.) 2. Multiple telangiectases with segmental distribution—a form of segmental nævus.
(The case of bilateral telangiectases of Dr. Colcott Fox and a similar unpublished case of the writer's with unilateral distribution.) |
| Acquired. | { | <ol style="list-style-type: none"> 1. De Morgan's spots—the bright red, slightly raised, seed-like growths common on chest and trunk in later middle life (associated, according to some writers, with malignant tumours or diseases of the liver, but frequently seen without this association.) 2. Telangiectatic lesions of the skin associated with Graves' disease (Hyde). 3. "A family form of recurring epistaxis with multiple telangiectases of the skin and mucous membranes." The angiomas in these cases are mainly of the raised nodular type, and situated especially on the cheeks, lips, ears, and nasal and buccal mucous membranes.
(Cases of Osler and Parkes Weber and many others, most of which are noted by Weber and by Colcott Fox.) 4. An aberrant form of the angiokeratoma of Mibelli and Pringle, in which the lesions occur particularly upon the scrotum, trunk, and extremities. Sometimes a family complaint, but unassociated with recurring epistaxis.
(Cases of Fordyce, Anderson, Sutton, Sibley, etc.) |

REFERENCES.

(1) PARKES WEBER.—"Multiple Hereditary Developmental Angiomas (Telangiectases) of the Skin and Mucous Membranes with Recurring Hæmorrhages," *Lancet*, 1907, vol. ii, p. 160 (with an epitome of all the recorded cases of the Osler type of multiple telangiectases).

(2) FOX, T. COLCOTT.—"A Case of Bilateral Telangiectases of the Trunk, with a History of Marked Epistaxis in Childhood and Recent Rectal Hæmorrhage," *Brit. Journ. Derm.*, May, 1908, vol. xx, p. 145 (drawing of histology, p. 148).

(3) PAUL, S. NORMAN.—“Hereditary Angiomata (Telangiectases) with Epistaxis,” *Brit. Journ. Derm.*, January–March, 1918, p. 27 (photo-micrograph of histology).

(4) DUBREUILH.—“Angiokératome plan,” *Ann. de Derm. et de Syph.*, 1893, p. 379.

(5) ANDERSON.—“A Case of Angiokeratoma,” *Brit. Journ. Derm.*, April, 1898, p. 113 (diagram of distribution and drawing of histology).

(6) FORDYCE.—“Angiokeratoma of the Scrotum,” *Journ. of Cut. Dis.*, 1896, p. 81 (coloured drawing).

(7) SUTTON.—“A Clinical and Histological Study of Angiokeratoma,” *Journ. Amer. Med. Assoc.*, July 18th, 1911, p. 189 (drawing of scrotum).

CONGENITAL UNILATERAL NÆVUS (“NÆVUS UNIUS
LATERIS”) OF ANGIOKERATOMATOUS STRUCTURE,
WITH LOCALISED HYPERKERATOTIC OUTGROWTHS
ON THE CONGENITALLY TELANGIECTATIC BASIS.

By F. PARKES WEBER, M.A., M.D., F.R.C.P.

THE patient, A. S—, aged 15 years, was a healthy and well-developed lad excepting for a congenital vascular nævus, which involved a great part of the left lower extremity, including the gluteal region. The vascular nævus was a diffuse superficial capillary and venous angioma, some of the superficial veins being greatly dilated as well as the capillaries. On this congenitally telangiectatic basis there were some hyperkeratotic outgrowths about the knee,

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