Wolff and Limarzi (1946) reported the case of a 30-year-old woman presenting with 50 cm. of gangrenous small bowel prolapsing through the internal os. The bowel was devoid of its mesentery, and at operation 150 cm. of gangrenous small bowel was excised. The perforation in the uterus measured 3 cm. She made a good recovery. Simon, Kaufman, and Mastellone (1951) described the case of a woman aged 25 who presented with small bowel prolapsing through the internal os. It was at least three days before laparotomy and resection of the damaged bowel was carried out. In spite of the delay her general condition remained satisfactory. After the operation she made a good recovery. Potts and Petzing (1953) and Howkins (1952) recorded one case each of bowel injury associated with perforation of the gravid uterus.

An astonishing feature of these three cases reported from the United States and of our case was the absence of shock in spite of severe damage to the small bowel culminating in gangrene. This is probably due to the minimal bleeding as a result of retraction and clotting of the torn mesenteric vessels, and also partly to the "tourniquet" effect of uterine retraction. In the case reported by Howkins, though the general condition was satisfactory initially, the patient progressively deteriorated as the bleeding continued unarrested into the peritoneal cavity.

Furthermore, the absence of purulent peritonitis even after 24 hours of the assault was striking in our patient. This, we believe, was due to the isolation of the cut ends of the bowel within the uterine cavity. With the lapse of sufficient time perforation of the gangrenous bowel and fulminating peritonitis would no doubt have proved fatal.

This young woman remained well one year after loss of all except approximately 20% of her small bowel. Her weight remained steady, though below normal. She seemed likely to develop vitamin-B₁₂ deficiency in the near future if not treated; otherwise there had been no major metabolic disturbance, though she continued to have very considerable steatorrhea.

Summary

A West Indian woman aged 23 was admitted with a history of lower abdominal pain and vaginal bleeding of 24 hours' duration. A total of 360 cm. of small bowel was removed during criminal abortion. At laparotomy a further segment of gangrenous jejunum and ileum was excised, together with a lacerated uterus. On completion, approximately 105 cm. of healthy jejunum and 5 cm. of terminal ileum remained. Post-operatively, profuse diarrhoea and steatorrhea occurred, in consequence of which she was maintained for an experimental period of 10 days on a synthetic diet consisting of medium-chain triglyceride, with the disappearance of steatorrhea and return of normal faecal-fat levels. At the end of one year her weight remained steady.

Our thanks are due to Mr. T. F. Dee for photographic services. We also wish to thank the Criminal Investigation Department of the Birmingham City Police for providing valuable information and allowing us to use photographs of specimens.

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Medical Memoranda

Blue Rubber Bleb Naevus

Brit. med. J., 1966, 2, 931-932

Bean (1958) coined the term "blue rubber bleb naevus" for a syndrome of a particular type of vascular naevus having the appearance of vascular bladders on the skin and mucous membranes and at times associated with gastro-intestinal bleeding. Only about 12 cases of the condition have been reported, and the latest reviews are by Rice and Fischer (1962) and Butterworth and Streat (1962). The purpose of this paper is to describe two more cases, one with very widespread naevi and the other showing few scattered lesions. Both cases demonstrate some differences from those previously recorded, notably the absence of pain and sweating of the lesions and the history of bleeding from sites other than the gastro-intestinal tract. The two patients also show inheritance through three and five generations respectively. Histological features and the differential diagnosis are discussed. Both cases were shown at the clinical meeting of the British Association of Dermatology Annual Meeting in July 1965.

CASE 1

A woman aged 56 had had a blue nodule at the right outer canthus since infancy, and this, together with a small lesion near the mouth, was removed by Mr. Denis Bodenham in 1948. When aged 39 further lesions in groups on the left upper arm and right shoulder and a few scattered in other areas of the body started to develop. Some lesions apparently followed pregnancies or occurred at injection sites. They have not given rise to pain but were slightly tender on pressure. She has had attacks of frequency and urgency of micturition and one episode of haematuria three years ago. Menorrhagia developed just before the menopause two years ago. There was no history of bleeding from other sites. The affected members of the family are shown in Fig. 1.

General and gynaecological examination revealed no significant abnormalities. Blue haemangiomas ranging in size from 2 to 20 mm. in diameter were present on the right shoulder, left arm, hands, legs, and left heel. Some larger lesions were surrounded by smaller satellites. The lesions were slightly tender on handling. Pressure emptied the haemangiomas of blood and gave a sensation of a gap in the underlying tissue. The mucous membranes were not affected. No excessive sweating was apparent over the lesions even after heating the limb encased in polyethylene.

Investigations.—Blood count normal. Occult blood tests on stools negative. Repeated specimens of urine showed no erythrocytes,

leucocytes, or other abnormality. Skin biopsy (Dr. O. C. Lloyd): the section showed the nodule to be a cavernous haemangioma with smooth muscle in the walls; a few sweat glands were incorporated.

FIG. 1.—Case 1. Family tree.

CASE 2

A man aged 48 had had dark-blue vascular lesions of various sizes since childhood, but new ones were continually developing. They were painless and did not alter with change of temperature. He had an unexplained attack of haematuria in 1961. In 1963 he began to suffer from attacks of dizziness and sweating, tingling of the tongue, and vomiting. In January 1964 he had a number of minor attacks of nose-bleeding, and in December 1964 a slight haemoptysis occurred. The affected members of the family are shown in Fig. 2.

FIG. 2.—Case 2. Family tree.

General examination revealed a well-nourished healthy man. B.P. was 120/70. Fundi were normal. Neurological examination showed no abnormality. There were numerous dark haemangiomas, ranging in size from 1 to 20 mm., on the hands, arms, feet, and legs, and a few on the trunk. These could be compressed, giving a sensation of pushing the blood through a gap in the underlying tissue. They were not tender. A few lesions under the tongue were associated with tortuous dilated veins. No sweating over the lesions was apparent.

Investigations.—Radiographs showed no abnormal opacity in the lung fields and no bony erosion in the skull. E.E.G. showed no focal lesion or evidence of epileptic diathesis. The pattern of response to photic stimulation was the one typically seen in migraine. Scanty erythrocytes were present in urine on one occasion. Blood cytology was normal. E.N.T. examination showed no naevi in the nose, post-nasal space, mouth, larynx, pharynx, or external and middle ear. The audiogram was normal. Cystoscopy disclosed some urethral stricture and trabeculation in the bladder. Bronchoscopy was normal. Skin biopsy: sections show the nodule to be a

cavernous haemangioma with smooth muscle in its walls; the appearances are the same as those seen in Case 1.

COMMENT

The two cases reported show some of the typical features of blue rubber bleb naevus. This consists of widely scattered groups of haemangiomas varying in number from a few to hundreds and in size from 1 to 20 mm. The lesions themselves are well described by the title; they are blue and can be emptied by continued pressure. When they are empty the finger encounters a gap in the supporting tissues, suggesting that the haemangioma has herniated through the dermis. Our cases have not shown the increased sweating over the lesions previously described, nor have the lesions been painful except on heavy pressure.

The importance of the condition lies in the association with gastro-intestinal bleeding as stressed by Berlyne and Berlyne (1960), and if the skin lesions are few in number this underlying cause of the bleeding could be missed. Neither of the two cases described above had gastro-intestinal bleeding, but haematuria had occurred in both, epistaxis and haemoptysis in Case 2, and menorrhagia in Case 1, though the latter symptom had occurred just before the menopause and may be unconnected. That bleeding is likely to occur from other sites is suggested by the necropsy findings of Rice and Fischer (1962), who demonstrated lesions in the lungs, pleura, peritoneum, lower intestine, and skeletal muscle. The cause of the attacks of dizziness in Case 2 was not discovered. No evidence of a focal disturbance was obtained, but a small cerebral haemangioma would be difficult to demonstrate.

Fine *et al.* (1961) describe what they thought to be the typical histology of the condition. There are large numbers of dilated vascular spaces whose walls may be thrown into folds. The lining spaces of the cells are often cuboidal. The vascular spaces are separated by smooth muscle in some areas and fibrous tissue in others. Sweat glands are closely related to many of the vascular structures. In both our cases smooth muscle was apparent in the walls of the haemangiomas.

Some cases previously recorded have not had a family history of haemangiomas, but Butterworth and Stearn (1962) comment that it appears to be transmitted as an autosomal dominant abnormality with no evidence of sex linkage, and this is confirmed in our patients.

The differential diagnosis is not difficult. In hereditary haemorrhagic telangiectasia (Osler's disease) the haemangiomas are flat, red, and often show telangiectatic vessels. Bean (1958) mentions that larger cyanotic lesions may occur and a patient we have seen recently with this condition showed a large number of blue blebs over the face with some resemblance to blue rubber blebs, though pressure demonstrated that they were interconnected and really a mat of grossly dilated vessels.

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