Cases in a Family

A girl aged 2½ was first brought to hospital with scabies, the child being carried despite her age. Inquiry showed that bullae had been present on her feet each summer since birth. The severity was such that she could not get out of the upstairs flat for four to five months each summer unless she was carried. Her younger brother, aged 1½, was already affected. Her mother was affected, as were the maternal grandmother, great-grandmother, and great-great-grandmother. The condition was so obviously inherited from the mother's family that the father (who claimed not to have known about it before marriage) was annoyed to the extent of threatening to leave the house and family. There was no other abnormality present, the hair, nails, and teeth all being normal.

The full family tree is shown.

The patient's mother had been badly affected as a child, but now (aged 27) was only occasionally troubled by bullae on the soles of her feet and where the dorsa of her feet were rubbed by shoe straps. The patient's grandmother suffered from hyperhidrosis and blistering of hands and feet all her life (she died of pneumonia aged 59). Even writing a short letter would produce blisters. The two previous generations were also known to have suffered severely until their death (in old age and at 45 respectively). The inheritance is that of a non-sex-linked Mendelian dominant.

The feet of both the patient and her brother showed areas of erythema and tense bullae up to 2 cm. in diameter. Each lesion would last up to a week and would then gradually heal without scar formation. On no occasion did examination show hyperhidrosis, but the mother insisted that they did sweat excessively. Bullae were not seen on any other area of the body.

Sodium hexametaphosphate powder, propantheline, and dapsone were used without effect. Treatment with corticosteroids (triamcinolone 2 mg. b.d.), however, rapidly controlled the blister formation in both the children. After the second day of treatment no new blisters developed, and after seven days the feet had virtually returned to normal. The children were once again able to return to their normal activities. After 15 days' treatment the mother stopped the tablets as the boy had developed diarrhoea (despite the fact that two other members of the household had the same complaint). When seen again, seven days later, several blisters had appeared. Steroids were restarted and the feet improved once more. Ten days later the mother again stopped the tablets because she thought the boy was jaundiced. When seen four days later there were two lesions on each of the boy's feet and one on the girl's right foot. The tablets were restarted, again with successful results, and treatment was continued for a further five weeks, by which time it was the end of September. This was the time of year when in previous years the feet had spontaneously ceased to give trouble. One blister developed at this time on the girl's right foot, but soon cleared.

Summary

A family with severe localized epidermolysis bullosa is described, 14 members of the family being affected. Corticosteroids depressed the formation of bullae. Relapse immediately followed cessation of treatment.

I should like to thank Dr. Netta Hay for permission to study and report this case.

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ADENOCARCINOMA OF CERVIX IN A PRIMIGRAVIDA AGED 18

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Invasive carcinoma of the cervix is seldom found during pregnancy. Chassar Moir (1956) quotes an incidence of "about 1 in 2,000" at the Glasgow Royal Maternity Hospital. However, this incidence may be unusually high, for Browne and Browne (1955) have given a figure of 1 in 4,000 from University College Hospital, London, and Hayden (1956) found 12 cases at the Chicago Lying-in Hospital between 1931 and 1954, an incidence of 1 in 6,817.

Rarer still is the disease in the young primigravida. Kistner *et al.* (1957) had only nine primigravidae among a series of 136 cases, and only two of these were under 20 years of age. In only 22% of 124 cases reported by Sadugor *et al.* (1949) were the patients under 30, and one of these was only 18; she was the youngest case found by us in the literature.

The following report of an 18-years-old primigravida with a cervical adenocarcinoma and an abnormal foetus is of interest because of its extreme rarity and because it emphasizes the importance of making a speculum examination in every case of vaginal bleeding during pregnancy lest the occasional cancer of the cervix is missed and treatment delayed.

Case Report

A primigravida aged 18 years and 3 months first attended the Belfast City Hospital Antenatal Clinic at the twenty-second week of pregnancy. There had been no vaginal bleeding since the last menstrual period, which had begun on October 23, 1958. The only abnormality noted was that the uterus was slightly larger than the period of

amenorrhoea. Vaginal examination was not performed. Two weeks later she complained that two episodes of bright-red vaginal staining had occurred since her last examination. Admission to hospital was arranged though the staining had ceased.

On admission hydramnios was diagnosed and an x-ray film showed a single anencephalic foetus. Speculum examination revealed a cauliflower-like, friable, clinically malignant growth about 1 in. (2.5 cm.) in diameter arising from the anterior lip of the cervix. A piece of the tumour was detached by the examining fingers and sent for urgent histological report.

Microscopy demonstrated a highly anaplastic adenocarcinoma of somewhat papillary structure, many of the cells of which showed a vacuolated cytoplasm. There was no tendency to undergo squamous change. No transition to mesenchymal-like elements was found and there was nothing to suggest the possibility of a mixed mesodermal tumour of the sarcoma botryoides type.

On April 15, 1959, at the twenty-fifth week of pregnancy, examination under anaesthesia confirmed that the growth was clinically confined to the cervix (stage I). The foetus,

which was small and anencephalic, was delivered by hysterotomy through a mid-line upper uterine segment incision.

Post-operative convalescence was smooth and the lochia was normal. The patient was discharged from hospital 13 days after delivery.

On May 18 she was readmitted; and on May 20, five weeks after delivery, radical hysterectomy, with removal of a large cuff of vagina, was carried out. Though pelvic glands and fascia were cleared in the usual manner, the right ovary was conserved in deference to the patient's youth and doubtful prognosis. Three pints (1.7 litres) of blood were transfused during operation. Post-operative convalescence was again uneventful and the patient was discharged on the fifteenth post-operative day.

Examination of the operation specimen (Fig. 2) confirmed the previous finding of an anaplastic adenocarcinoma originating from the cervix. There was no direct infiltration of the parametrium, but, unfortunately, microscopical metastases were found in the parametrial lymph nodes and in a gland from the right obturator fossa. The external iliac glands were not involved.

The patient was last reviewed on November 16, 1960, 18 months after hysterectomy. She was apparently in excellent health and was free from symptoms. Questioning revealed that coitus was normal and there had been no menopausal symptoms. The vagina was almost normal in length and its vault was perfectly healed. Nothing abnormal was detected during vaginal, rectal, and abdominal examination.

Discussion

The prognosis for this patient must be regarded as poor. Bonney (1941), Taussig (1943), and Meigs (1951) have given figures of 23%, 21%, and 26% respectively for the five-year relative apparent recovery rate after radical hysterectomy in the non-pregnant patient with involved pelvic nodes. The results of treatment of cancer of the cervix during pregnancy have been discussed by Kistner *et al.* (1957) in a careful review of 136 cases, and by Holzaepfel and Ezell (1958) in a separate review of 28 cases. These authors came to the conclusion that the results of treatment during the first two trimesters of pregnancy were no worse than for comparable stages of the disease in the non-pregnant patient, but when the disease was discovered and treated during the third trimester or within six months after delivery the results were much worse, only about one-third of the expected five-year survival rate being achieved. Danforth (1937) had previously put forward a similar view regarding the prognosis.

We do not propose to discuss the many aspects of the treatment of carcinoma of the cervix associated with pregnancy, but it is worth noting that most authors have preferred radiotherapy (after abdominal removal of the foetus if pregnancy has progressed beyond the first trimester). Surgical treatment was chosen in our case because conservation of some functioning ovarian tissue was desirable in this very young and recently married woman, and McCall *et al.* (1958) have recommended this policy. In retrospect, however, it is felt that it would have been better to proceed at once to radical hysterectomy at the time of caesarean section (as recommended by Donald, 1959) instead of waiting for five weeks, during which time some spread of the tumour may well have occurred.

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FIG. 1.—Photomicrograph of adenocarcinoma from the cervix ($\times 100$.)

FIG. 2.—Excised uterus (C) with vaginal cuff (B), showing the carcinoma (A) arising from the cervix.

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Medical Memoranda

Myiasis from the Tumbu Fly: Two Cases Recorded in Britain

Cordylobia anthropophaga (Blanchard), the tumbu or skin maggot fly, is widespread in Africa south of the Sahara, but is not found elsewhere. In West and Central Africa it is a common pest of rodents (especially rats), humans, dogs, and occasionally other animals. Man might not be an ideal host, because the maggot does not always fully mature in him.

The adult is a yellowish-brown fly, 6–12 mm. in length, which feeds entirely on plant juices. Eggs are normally laid on floor dust, dry sand, or clothing which has been soiled by urine or faeces, but if the elective site is too moist they are deposited near by. The larvae are hatched out on the third day and remain alive for about two weeks. During this period they will attach themselves to any host animal which touches them. The larvae of the tumbu fly penetrate the skin and grow rapidly, undergoing successive moults, and reaching maturation by the eighth to the tenth day, when they leave their "boils" and drop to the ground to pupate. At this time they usually measure 13–15 mm. in length. This is the time sequence in a perfect host. In humans, however, development may be much slower, and in an infected volunteer a third-stage larva measuring only 9 mm. in length has been removed on the fifteenth day. From the time factor alone, therefore, the chance of encountering tumbu infection in Britain is remote.

REPORT ON TWO CASES

The patients, a boy aged 3 years and his sister aged 6 months, had been living in Ghana. On the day after their return to England the girl developed small pimples which rapidly grew into "boils" (Fig. 1), and her brother was also found to have a spot on his scrotum and one on the middle of his back. She was given penicillin, but the "boils" continued to enlarge. The mother had taken every precaution against exposing the children to tumbu-fly infection, but, as they had been playing in a dry sand-heap three weeks previously, it was suspected that both of them might have been attacked by larvae of the tumbu fly, and the

girl was admitted to hospital. The boy was not in the least distressed. A maggot was expressed from the lesion on his back and no treatment was required. On the other hand, his sister had over 30 widely dispersed septic lesions. They varied from the size of a pea to that of a butter-bean, and the larger ones were oval-shaped; each was inflamed, with a hole in the centre which exuded a clear serous fluid. If this had been pus the resemblance to an ordinary boil would have been complete. Pressure or manipulation did not seem to be painful, but led to a slight movement within.

The girl was given chloral so that the lesions could be cleaned with a mild antiseptic. Afterwards ethyl chloride was sprayed on to the skin surface and a maggot was expressed through each hole (Fig. 2). Oral tetracycline was also given to forestall infection of the empty cavities. Next day there was a remarkable improvement, as the child was no longer miserable and the lesions already showed signs of healing. Three weeks later only faint marks indicated the sites of former lesions.

Investigations.—Blood: Hb, 77%; W.B.C., 14,200/c.mm. (70% neutrophils, 26% lymphocytes, 4% monocytes); marked neutrophil leucocytosis with a slight left shift. Repeated swabs from the "boils" showed no growth.

Examination of Larvae.—The larvae varied from 6–7 mm. to 11–12 mm. in length. Despite the variation in their size they were morphologically uniform, and were identified by Mr. Glyn Williams as third-stage larvae of the tumbu fly.

COMMENT

Myiasis, an infestation of the body by larvae of dipterous insects, may be (1) specific, from larvae of obligate parasites which feed on living tissue; (2) semi-specific, from larvae which normally feed on decaying flesh but which are facultative parasites of wounds and diseased tissues; and (3) accidental, from larvae that normally live on excrement and decaying organic matter, but which may accidentally reach the gut in contaminated food.

Specific myiasis in Britain occurs only when a maggot is deviated from its normal animal host. The animal

FIG. 1

FIG. 2

Fig. 1.—Tumbu "boils." Fig. 2.—Some of the tumbu larvae after expression from the skin. (The scale is in centimetres.)