Onyalai — A Disappearing Disease Entity

A CASE REPORT AND REVIEW OF THE LITERATURE

BY

A. C. B. WICKS, M. B., M. R. C. P.
Lecturer in Medicine, University of Rhodesia

Onyalai is an interesting condition both clinically and histologically. It is characterised by haemorrhagic bullous eruptions of the mucous membranes, usually in the mouth, accompanied by thrombocytopaenia and occurs predominantly in young adult males.

A case of onyalai is presented along with a review of the literature.

A Case Report

VC, a 35-year-old African female was admitted to Harare Hospital with a history of having been well until three days prior to admission when she first developed severe frontal headache and general malaise. One day later she noticed sore-like lesions on her tongue and a few hours later similar lesions developed on the inside of her cheeks. These lesions rapidly enlarged and bled easily with the slightest trauma.

There was no history of drug taking, haematuria or a bleeding disorder in the family or early life. She had never been in hospital before.

The main features on examination were those of an ill apyrexial woman with haemorrhagic bullae in the mouth. One bullus was situated on the lateral aspect of the tongue and measured approximately 2 cm. x 1 cm. in diameter. Four other similar lesions, but rather smaller, were on the buccal mucosa of both cheeks. All were oozing blood. A few purpuric areas were also seen on the skin of the neck and in the right supraclavicular fossa. The tonsils were enlarged but not inflamed, and no lesions were seen in the vagina. There was no lymphadenopathy, splenomegaly or jaundice. All other systems were normal.

Progress

Initially, 20 mg. prednisolone was given followed by 10 mg. q.d.s. Because of the purpura she was transfused with one unit of fresh blood. During the first 24 hours her condition remained unchanged, but a fresh ulcer developed on the lower lip while the other mouth ulcers improved with remarkable rapidity. Three days after admission the mouth ulcers had healed and the skin purpura had faded; however, platelets were still only 27,000 per cu. mm. Two days later the platelets had returned to normal and the patient was clinically well with no evidence of either bullae or purpura. On outpatient follow-up with a reducing regime of prednisolone she made a completely uneventful recovery.

Discussion

Historically, this disease was recognised a long time ago by the indigenous population of Rhodesia. The earliest description was by an educated African layman called Jason Machiwanika, in the early part of this century. He described the disease called Mhuka in Shona, or N’ombe in Cimanyika, both much feared by the local populations. It was characterised by itching and small swellings all over the body associated with uncontrollable bleeding from the nose and mouth. He thought it was related to the heavy rains and stated that it was uniformly fatal.

Jackson,2 a Native Commissioner wrote in his report of 1902 that a disease was fairly common in his area called “inkomo” which was characterised by a discharge of blood in clots from the nose and roof of the mouth.

A more general awareness of this disease in Central Africa resulted from Wellman’s (1904) description of this same bleeding disorder which he called onyalai. The derivation of this name is not apparent from his writing. He reported further cases in 1905, 1907 and 1908.

Little mention was made of the disease again until 1925 when Ellis, a government medical officer described the clinical features of onyalai in a report submitted to the Medical Director of Southern Rhodesia.

Four years later, Scott (1929) added further notes on the disease. However, in the 1930’s and early 1940’s a large number of cases began to be reported. Gilkes (1934) writing from Broken Hill in Northern Rhodesia described 53 cases. Three years later, Blackie (1937), writing from Sali-
Onyala, was the first author to establish that thrombocytopaenia was present in this condition.

Further accounts were reported from South West Africa (Helman, 1938), Tanganyika (Keevil, 1938), Nyasaland (Shelley, 1938) and Bechuanaland (Morgan & Squires, 1940).

However, during the past few years little mention has been made of this condition in the literature. At a recent conference in Malawi, Edington, Hodgkinson and Seftel described 15 cases of onyala seen at the Jane Furse Memorial Hospital in the Transvaal over a 16-month period—September, 1968, to January, 1970.

In the Mashonaland area of Rhodesia very few cases have been seen over the last few years and this appears to be the experience of most practitioners working in the rural areas, where before, it was a disease fairly commonly encountered.

Clinically onyala is usually characterised by a mild prodromal illness consisting of headache, malaise and general body aches, followed shortly afterwards by bleeding from the mucous membranes of the nose, mouth and occasionally vagina (Gilbert, 1943). Haematuria is uncommonly encountered in Rhodesia while it was a prominent feature in Wellman's (1908) cases in Angola. Occasionally bullous eruptions may be seen in the nasopharynx, larynx, trachea and oesophagus. Gastro-intestinal bleeding with lesions in the gut are less commonly seen. Purpura may or may not be seen in the skin.

The hallmark of this disease is a blood blister usually found in the mouth. These blisters characteristically ooze blood or bleed even with the slightest trauma. They often clear remarkably quickly as was noted in the case report above. These bullous lesions are rarely encountered in idiopathic thrombocytopaenic purpura. Occasionally relapses of these lesions occur and Gelfand (1948) estimated that this occurred in 25 per cent. of his patients.

Splenomegaly is not a common finding. On investigation the main abnormalities are found in the blood. Thrombocytopaenia is usually present and was first described by Blackie (1937), and confirmed by Gear (1938), and Stein and Miller (1943). However, Laufer (1953) noted that 50 per cent. of his cases did not exhibit this abnormality. Clot retraction is usually impaired and the bleeding time is prolonged with a normal coagulation time. Bone marrow examination is normal. (Stein and Miller, 1943).

Most reported cases are those of young adult males. The sex incidence is set out in Table I. Although most authors show this male predominance, Strangeway and Strangeway (1949), describing a large series in Angola, found the sex incidence to be equal.

The disease has only been described from the African continent and almost exclusively in the African races although Gear (1944) described three cases in Europeans and one case in a female of Chinese origin in South Africa. Gelfand (1954) and Shee (1961) both described single cases in Europeans in Rhodesia.

Wintrobe (1937) writing from the U.S.A. noted that idiopathic thrombocytopaenia was uncom-

<table>
<thead>
<tr>
<th>Author</th>
<th>Males</th>
<th>Females</th>
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<tbody>
<tr>
<td>Gilkes (1934)</td>
<td>48</td>
<td>4</td>
</tr>
<tr>
<td>Gear (1938)</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Morgan &amp; Squires (1940)</td>
<td>19</td>
<td>5</td>
</tr>
<tr>
<td>Stein &amp; Miller (1943)</td>
<td>15</td>
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<tr>
<td>Squires (1950)</td>
<td>22</td>
<td>13</td>
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<tr>
<td>Gelfand (1954)</td>
<td>31</td>
<td>14</td>
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<td>179</td>
<td>57</td>
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Geographically, the disease has a fairly well-defined area of incidence (Wilkinson, 1953). As can be seen from Fig. 1 the area involved is bordered to the north by the equator, to the south by 28° latitude, to the west by the Atlantic and to the east by a line drawn from the Kenyan Highlands down to the eastern border of the
Rhodesias and Transvaal. The majority of cases reported have occurred in Angola, the Rhodesias, Bechuanaland and Northern Transvaal. However, sporadic cases have been reported from the Congo (Massey, 1907), Kenya (Welch, 1920) (Wilkinson, 1952), South West Africa (Helman, 1938), Tanganyika (Keevil, 1938), Nyasaland (Shelley, 1938), Nigeria (Jelliffe, 1950) and Uganda (Trowell, 1951).

Altitude appears to make little difference to the disease and it occurs in both rural and urban areas. In most series it occurs all the year round and is not related to any particular weather condition.

AETIOLOGY

The aetiology of the disease has remained a mystery; although many hypotheses have been put forward, none have found general acceptance. Stein and Miller (1943) considered that onyalai was a variant of idiopathic thrombocytopaenia which was first described by Werlhoff in 1735. Metz and Kramer (1943) considered that the term onyalai was misleading. They pointed out that in their experience blood bullous eruptions also occurred in the Bantu in such conditions as aplastic anaemia, megaloblastic anaemia, erythraemic myelosis and chronic I.T.P. They also felt that acute I.T.P. was commoner in the Bantu than Europeans. Wilkinson (1956) also noticed bullous eruptions in an East African patient who had aplastic anaemia. A summary of the differences appear in Table II.

### Table II

| Essential Differences Between Onyalai and Idiopathic Thrombocytopaenia Purpura |
|---------------------------------|---------------------------------|
| Onyalai | I.T.P. |
| Age | Young adults | Young adults and children |
| Sex | Mainly males | Mainly females |
| Race | African | European |
| Geography | Africa | Ubiquitous |
| Bullae | Common | Rare |
| Mortality | High | Low |

Mense (1906) thought that it may be due to poisoning from the Euphobiciae tree. Strangeway and Strangeway (1949) and Gilkes (1934) commented upon the poor nutrition of their patients and postulated that Vitamin C deficiency played a major role. Gelfand (1954) noted that most of his patients were well nourished and thought this explanation unlikely. Gear (1938) and Blackie (1937) noted that some of their patients had a positive Wassermann reaction.

Gilkes (1934) was the first to report that there may be a familial aspect in this disease. He described a family of four children, two of whom developed the disease within a year of birth. Squires (1943) described two brothers with the disease.

The most widely held view is that onyalai is the result of some sort of poisoning especially African medicine or drugs. Stein and Miller (1943) reported two African patients, both of whom admitted taking African medicine. In one of these patients they obtained some of the medicine used, made a suspension and injected this substance into a guinea pig. This produced haemorrhagic areas in most of the guinea pig's organs and a thrombocytopaenia. The classical bullae, however, were not seen. However, many authors note that there is often no history of African medicine although the average patient may not be very willing to tell the doctor about previous medications prescribed by the witch-doctor. Often patients only take African medicine after the symptoms first begin.

Apart from African medicine, drugs have been incriminated by many authors. Gear et al. (1944) made a strong case for drugs causing onyalai. They described three cases following neoaarsphenamine treatment while Rigby (1949) reported an onyalai-like picture in a patient who had been treated with N.A.B. and Bismuth. However, he noted that this was the only case he had seen associated with these drugs which had been widely used in his area. Phenobarbitone has also been incriminated. Gelfand (1954) reported seeing it in a European woman following phenobarbitone ingestion and noted that Stein and Miller (1943) had also seen a similar picture.

Wellman (1908) regarded onyalai as a specific disease entity and thought that it was an infectious process. Laufer (1953) postulated that it was an allergic manifestation with disease of the capillaries.

In this particular case report, as in many others reported, there was no history of ingestion of African medicine, drugs or previous medications. As far as the author is aware viral studies or platelet antibodies studies have not been carried out in this disease.

MORTALITY

The overall mortality in the above quoted series was 14 per cent. It is interesting that the severity of the disease, and likewise the mortality, varied from area to area. Morgan and Squires (1940) noted that although onyalai was widespread in Northern Bechuanaland the mortality was low and that the local population did not
have the same dread of the disease as reported
from other parts of Central Africa (Blackie, 1937).

Gilkes (1934) noticed that a subnormal tem­
perature was a bad prognostic sign, whereas a
good temperature response produced a better
prognosis. Stein and Miller (1943) felt that the
higher the temperature generally the more sick
the patient and the prognosis appeared to be
worse. Haematuria (Wellman, 1904; Squires,
1950) was found to have a bad prognosis.

The main cause of death was usually bleeding
internally and into vital structures elsewhere.

TREATMENT

There are as many conflicting reports on the
treatment of this disease as there are theories
of its causation. Wellman (1908) tried quinine,
alkaline salts, oil of turpentine, acetate of lead,
tannic acid, ergot and supra renal extract; all to
no avail.

Morris (1934) advocated intramuscular blood
and others confirmed the efficacy of this treat­
ment (Gilkes, 1934; Blackie, 1937; Gear, 1938;
Morgan and Squires, 1940). Gelfand (1954)
thought the result was non-specific in effect.
Arsenic in full doses was advocated by Gilkes
(1934) and Gear (1938). Strangeway and Strange­
way (1949), writing from Angola used large
doses of intravenous Vitamin C with excellent
results; in fact, they stated that without this treat­
ment 90 per cent. of their patients died. Laufer
(1953) used adrenaline and histamine with good
effect.

The consensus of opinion today appears to be
that steroids in high dosage are useful, as is
platelet enriched blood if it appears that a
catastrophic bleed is imminent. However, it would

seem that a large number of these patients will
spontaneously improve on no therapy at all.

In summary, the present situation would appear
to be that onyalai is a variant of I.T.P. and dif­
fers from I.T.P. in the bullous formation, the
geographical and racial distribution of the disease,
and male predominance. The mortality is variable
and the incidence of this condition appears to be
on the wane. No definite aetiological factors have
been universally incriminated and the treatments
of choice, although legion are probably: steroids
and platelet enriched blood.

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