Medical Journal of Zambia (1977), 11, 6, 172.

Good pasture's Syndrome in a Zambian woman.

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SUMMARY

The clinical and pathological characteristics of Goodpasture's syndrome are described and a case reported in which there were classical clinical and pathological manifestations of this condition. This is the first report of this entity in an indigenous black African.

INTRODUCTION

In 1919 a case of pulmonary haemorrhage associated with anaemia and albumenuria was described by Goodpasture whose name is generally given to this syndrome now. The autopsy findings in this original case were pulmonary alveolar haemorrhages and proliferative glomerulonephritis. There have been several reports of this condition subsequently but it is a rare entity there being only 52 reports in the literature up to 1964. The aetiology of the disease is unknown.

Goodpasture's syndrome is rare in women with a sex ratio of 9:1 (Benoit et al. 1964) and the only report of this condition in a non-caucasian is in an American negro (De Gowin et al, 1963). The disease has not been reported in children and most commonly occurs in young adults the peak incidence being between the ages of 16 and 25. Thereafter, there is a steady decline in incidence with age.

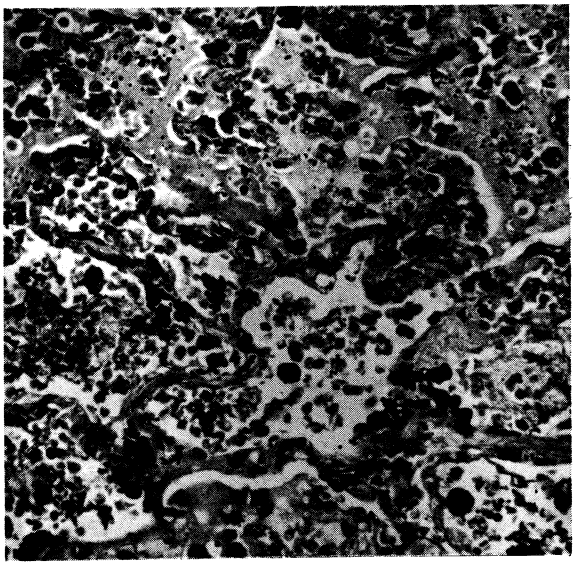
The presenting symptoms in the reported cases were remarkably constant. Haemoptysis was almost invariably present and was absent in only 1 of 53 cases. This was the usual presenting feature. Other symptoms in order of frequency was exertioal dysponea, fatigue, weight loss, chest pain and haematuria. Clinical examination generally revealed pallor and in rather more than half the patients widespread rales were heard in the lungs. Surprisingly hypertension was only present in two patients and in both of these a raised blood pressure had been recorded before the onset of their disease. (Heptinstall and Salmon, 1959) (Cruikshank and Parker 1961). Fundal changes were also surprisingly uncommon again occuring in only two patients (Cruikshank and Parker, 1961) (Saltzman et al. 1962).

Examination of the urine revealed invariable proteinuria which varied from small amounts to over 10 grammes in 24 hours (Benoit et al. 1964). Haematuria, usually microscopic, was present in most cases. and granular casts presents in about half the cases. Normocytic normochromic anaemia was generally present the degree of which depended on the extent of the pulmonary haemorrhage. Azotaemia occurred in the late stages of the disease in all patients but was not usually present initially. The chest X-ray was abnormal in the majority of patients characteristically showing bilateral confluent shadows densest in the perihilar regions.

Pathological findings in Goodpasture's syndrome are confined to the lungs and kidneys. Grossly the lungs are heavy and contain focal areas of red brown consolidation due to intra-alveolar haemorrhage with acute necrosis of alveolar walls and fusiform fibrous thickening of the alveolar septa.

The kidneys are as a rule enlarged and pale.

FIG. I



Lung with extensive intra-alveolar hamorrhage, hemosderin laden macrophages and firosis in some intra-alveolar septa.

Petechia haemorrhages are a constant finding. Histologically there is evidence of rapidly progressive glomerulonephritis typical of glomerular basement membrane disease (Proskey et al., 1970) with dominant

proliferative changes, crescent formation and increasing glomerular fibrosis as the diseases progresses.

The pulmonary and renal lesions are due to circulating antibodies directed against antigens common to both the alveolar and glomerular basement membranes (Koffer et al., 1969). Why these antibodies are present however is unknown.

Case Report

E.N. was a 35 year old African woman of the Ndebele tribe, born in Rhodesia but resident for many years in Lusaka. She had never previously been admitted to hospital and claimed to have enjoyed good health until about 1 week before her admission to hospital when she developed a profuse haemoptysis which was accompanied by central chest pain and continued unabated until her admission.

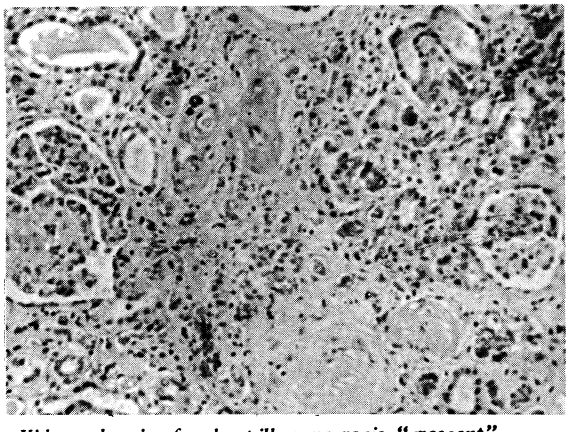
On admission she was fully conscious and well orientated but pale and breathless. The main findings were in the chest crepitations being heard in all parts of the lung but predominantly at the bases. The blood pressure was 140/100 m.m.mg. but there were no signs of hypertensive disease. The ocular fundi were normal. Blood and protein were present in her urine.

After her admission there was a rapid deterioration in her condition with continuing haemoptysis, progressive oliguria, terminal coma and death 36 hours after admission.

At autopsy apart from moderate concentric hypertrophy of the left cardiac ventricle there were changes only in the lungs and kidneys.

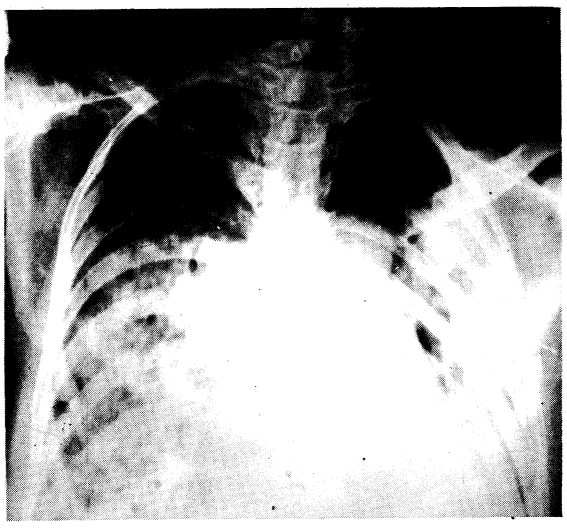
Grossly both lungs were heavy (left = 700 gms right = 1100gms) with smooth cut surfaces and an increased amount of foamy fluid. There was diffuse consolidation and dark red confluent patches. Microscopically the lungs showed areas of intra-alveolar haemorrhage and occasional proliferation of fibrous tissue in the alveolar septa. The kidneys weighed 150 gm each. The capsules were easily removed and showed a diffuse, fine granular surface, grayish in colour

FIG. II



Kidney showing focal capillary necrosis, "crescent" formation and fibrosis in glomeruli.

FIG. III



Chest X-ray showing widespread confluent opacities.

with many petechial haemorrhages. The cortex was pale and the medulla was congested. The calyces and pelvis were normal. Microscopically the kidneys showed changes in most of the glomeruli. In some glomeruli there was capillary necrosis with fibrinoid like material, in others there was crescent formation and some showed complete fibrosis with minimal secondary change in the tubules. There was no evidence of vasculitis. Unfortunately, facilities for immunofluorescent studies were not available but even so the combination of pulmonary haemorrhage with glomerulonephritis is highly suggestive of Goodpasture's syndrome.

DISCUSSION

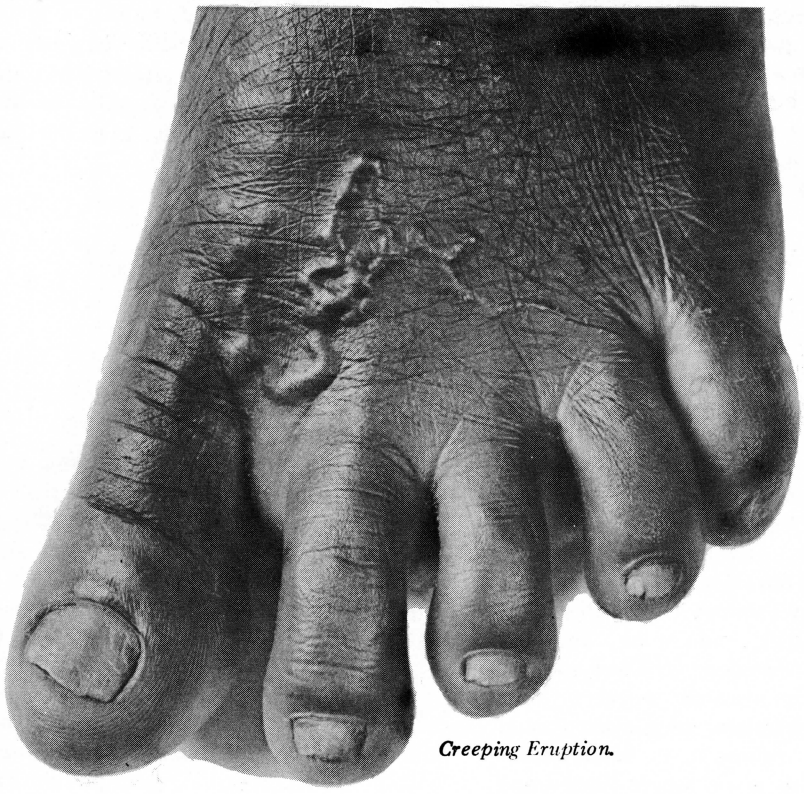
The clinical picture in this case is highly suggestive of Goodpasture's syndrome and this impression is upheld by the pathological findings. The interest of this case lies in the fact that this rare disease appears to be even more rare in non-Caucasions and this is the first report of this entity in an indigenous black African. The occurence in a woman makes this case even more unusual.

ACKNOWLEDGEMENT

We would like to thank the Medical Illustration Unit, Mr. M.A. Ansary for the photographs.

Spot the Diagnosis

by Dr. A.V. Ratnam – Lecturer in Dermatology.



Creeping Eruption.

Can you spot the diagnosis? For the answer see over page.

This patient presented with an itchy eruption on the foot of one week duration.

ANSWER

Creeping eruptions are caused by the presence in skin of the larvae of certain nematodes parasitic in animals, commonly the hook worms of cats and dogs. The areas most affected are those which come in contact with the ground-feet, buttocks and trunk. The characteristic feature of the condition is a progressive, pruritic cord which forms a bizarre pattern. The rate of advance is inconstant but usually 2 to 5cm daily. A reddish papule is often present at the active end.

Treatment of the creeping eruption may be local or systemic. Local treatment mainly consists of destructive methods like freezing and application of solid carbon-dioxide or liquid nitrogen. Among topical chemical agents reported successful are 25% Piperazine ointment, 2% Gammexane cream and Thiabendazole suspension. A variety of systemic medications including Antimonials. Hetrazone and Butazolidin have been reported effective but not uniformly. Thiabendozole in doses similar to those used for intestinal infections appears to be the most effective.

Book (Journal) Review

Volume 33. Number 1, January 1977 Publisher: The British Council.
65 Davies Street London W1Y 2AA Price: £5.00

This issue of the British Medical Bulletin reviews the subject of Disseminated Sclerosis (D.S.). Emphasis is given to the fact that little is definitively known about the aetiology, pathology and treatment. Authors of various papers have dealt with the subjects of epidemiology, pathophysiology, early diagnosis, the role of virus antibodies, immunology and biochemistry. This issue has been done very well and it is recommended that both research workers, postgraduate students and practising physicians should take the opportunity of reading it. Its value is even greater in the developing world where library and reference facilities are inadequate.

D.S. has been known to medical science for more than a century. It is probably the commonest neurological disorder, affecting adults of either sex, in Northern Europe and White settlers in America, Australia and South Africa. Its incidence is low in the original inhabitants of tropical and subtropical countries; in fact it may be completely non-existent. Appreciation of these facts has drawn attention to the role of latitude, genetic, dietary and environmental factors in the production of this disease. A large number of aetiological theories have been proposed from time to time. Allergy, infection by spirochaetes and viruses, auto-immune disturbances and thrombosis of cerebral venules due to disordered coagulation of blood, have all had their adherants. But there is no denying the fact that we still do not know the cause of this disease and this review reveals this in a most lucid way.

It has been very widely accepted that diagnosis of D.S. should be considered if objective signs are demonstrated at two or more sites in the Central Nervous System. This still remains true today. In the paper dealing with Diagnosis and Classification the authors have tried to evaluate the significance of evoked potential methods for objective detection of clinically silent lesions and the importance of examination of C.S.F. for oligoclonal IgG in the diagnosis of early cases.

The role of viruses and their antibodies in sera and C.S.F. has been discussed in some detail. This section has succeeded in presenting the masses of data in a clear and understandable form. It has been pointed out that measles virus may be involved in its pathogenesis. The virus probably initiates the disease in childhood and auto-aggressive inflammatory immune reaction accounts for much of the destruction in C.N.S. in later life. The patient ultimately dies because of the undesirable side effects of this process. The reaction takes place between the antigen (virus) and the immune system of the body.

Since a definitive cure still eludes us, treatment evolves around pilliative and ameliorative measures. The doubtful value of various dietary regimens, steroids, drugs against spirochaetal, rickettsial and virus infections and the role of immuno-suppressive therapy has been nicely presented in the last chapter.

A lot more work needs to be done to understand the basic cause of this steadily progressive malady. There is every hope that current multicentric research will solve this riddle in time.

P. SIKAND.

OMISSION

This table was to be published with Dr. Losada's paper but inadvertently ommitted.
Reference: J.S. Losada, Medical Journal of Zambia (1977) 11, 3, 73.

TABLE 1

Results of treatment with intra-arterial continuous. infusion of methotrexate in head and neck cancer – Dr. J.S. Losada: Medical Journal of Zambia (1977) 11, 3, 73.

Tumour Site	Number of Cases	Age	Sex		Reduction in Tumour Size		Objective Improvement & Symptomatic Relief	Total Failure
			M	F	Complete	Incomplete		
Maxillary sinuses	22	50–76	20	2	2	11	10	9
Nasopharyrix	9	34–48	8	1	–	8	8	1
Hypopharynx	8	65–75	8	–	–	8	8	–
Palate	5	42–80	4	1	–	5	4	–
Salivary glands	4	50–55	4	–	–	4	4	–
Tonsils	2	48–58	2	–	1	1	2	–
Larynx	2	62–70	2	–	–	2	2	–
Tongue	2	51–68	2	–	–	1	1	1
Middle ear	2	55–75	2	–	–	1		1
Skin	2	65–75	2	–	–	2	2	–
Lip	1	70	1	–	–	1	1	–
TOTAL	59	–	55	4	3	44	42	12