Wegener's Granulomatosis

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Wegener's granulomastosis is a fatal syndrome of unknown cause, described first by Klinger[1] in 1931 and Wegener[2] in 1936. Wegener characterized it as a syndrome consisting first of necrotizing granulomata of the upper of lower respiratory tract; second, generalized necrotizing vasculitis involving arteries and veins, almost always involving the lungs; and third, renal decompensation, almost always leading to terminal uremia and death. We should like to present a case of this disease, because we have been unable to find out references to this disease in Japanese otolaryngological literature, though several reports are available in the Europian literature.

REPORT OF CASE

B. Y., aged 62 years, male. The patient was admitted to the Hospital of the Yamaguchi Medical Callege on December 14, 1961 for investigation and treatment of nasal discharge acompanied by bleeding on the right nostril and swelling on the right cheek for three months. Except for pleuritis on the left side 25 years ago and rheumatoid arthritis 20 years ago, since childhood, he was in his usual health until June, 1961, when he developed nasal obstruction on the right side slightly. In mid September, he noticed bloody rhinorrhea and swelling on the cheek on the right side without pain. A few days later he developed disturbance of vision, eye discharge and exophthalmus on the right side with fever. Local treatment to the nose and eye, and antibiotics afforded no improvement. Because of fever and nasal bleeding on the right side he was referred to our clinic for diagnosis and treatment. There was no history of allergy, no history of renal disease. His mother died of cancer of the stomach.

On physical examination, he was weak, appeared chronically ill. Blood pressure was 128/78. His temperature was 37.9°C. The hand joint and the elbow on the right side were unflexible.

Both ear drums were normal. The right nostril was filled with a redded soft mass, so that we could not examine the turbinates. The left nostril is normal. There was pus on the postnasal area without mass. There was no mass on the septum. The pharynx and larynx were normal. The cheek on the right side was swollen without pain. The ophthalmological consultant diagnosed conjunctivitis and ad-

vised local treatment. The rest of the examination was negative. The blood picture was one of mild normocytic anemia, with a white blood count of 24000 per cu. mm., of which 15 per cent were N. band, 81.5 per cent N. segmented, 0.5 per cent eosinphil, 0.5 per cent basophil, 11.5 per cent lymphocyte, and 4.5 per cent monocyte. Hemorrhagic study showed in Fig. 1. Systematic blood chemistric examination revealed in Fig. 2. Fluid examination showed in Fig. 3. Urinalysis was negative. The sputum and gastric content were negative for tubercle bacilli by culture and concentration. A chest x-ray showed a round mass lesion in the left upper lobe (Fig. 4). X-ray of the nose revealed that there is a cloudness on the right maxillar sinus and nostril with bony defect on the base of the orbit on the right side (Fig. 5). The tentative diagnosis of malignant tumour on the maxillar sinus extending the nostril on the right side was made, and on Dec. 22, 1961 a complete maxillectomy with neck dissection and ligation of the external carotid artery was done. There was no enlarged lymphgland on the right sided neck. The maxillar sinus and the nostril on the right side were filled with tumour, extending to the orbit on the right side, which was sent to the pathology. The pathologist made a diagnosis of Wegener's granulomatosis (Fig. 6). A few days later the biopsy taken from the kidney showed glomerulonephritis (Fig. 7). His temperature was elevated from 39° to 37.5°C nearly every day through Dec. 31.

Fig. 1. Hemorrhagic Study

Name Mr. H. Yamamoto Ward E.N.T. Referred by Dr. Mogi Date 1～4～62
Age 62 Sex Ⓜ. F.

Bleeding Time (Ivy's Method)		
Free flow	5 min.	1～6
Oozing of blood-tinged fluid	0 min.	1～4
Capillary Fragility		
(Tourniquet Test)	0	No petechiae
Clotting Time		Less than
Tube 2 16', 3 16', 4 17', Average 16' 20" min.	25 min.	
Clot Retraction		
Per cent serum expressed	53.0%	40～60
Hematocrit	34.0%	
Fluid volume per cent of the clot	13.0%	0～20
Clot characteristics		
Thrombocyte Count		
(No. per 100 oil immersion fields)	675	300～1000
Plasma Prothrombin Time		Control
(Quick's Method)	12.4 sec.	10.3 sec
Serum Prothrombin time		More than
(Prothrombin Consumption Test)	300> sec.	20 sec
(Other Test)		

Summary of Abnormalities
Interpretation
Hemorrhagic study is normal.

J. Mizushima

Fig. 2 Systematic Blood Chemistry

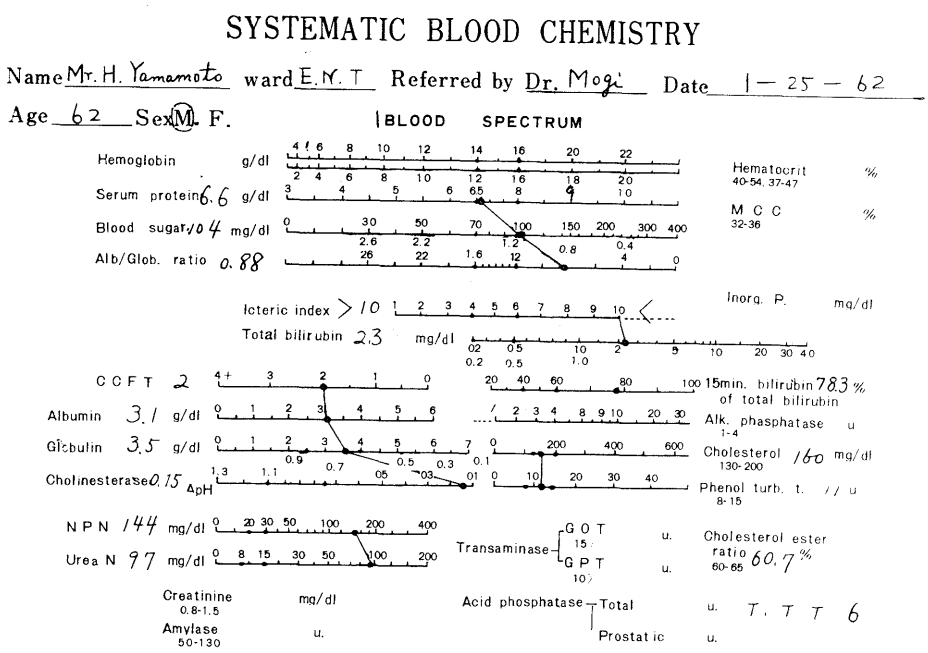

Fig. 3. Flud Examination

Name Mr. H. Yamamoto Ward E.N.T. Referred by Dr. Mogi Date 1~30~62
Age 62 Sex (M). F.

	Serum Electrolyte		Electrolyte Excreted in Urine 960 (volume ml/day)	
Sodium	129 mEq/l	136~148	42 mEq/day	130~260
Potassium	3.7 mEq/l	3.6~5.0	15 mEq/day	50~100 (27~ 40)
Calcium (total)	4.3 mEq/l	4.5~5.5 (4.8~5.7)	2 mEq/day	2.5~ 15
lonized Ca.	1.9 mEq/l	2.1~2.5		
Magnesium	2.2 mEq/l	0.8~2.0	7 mEq/day	4~ 16
Chloride	81 mEq/l	100~106	31 mEq/day	140~260
Bicarbonate	20 mEq/l	25~ 32		
Inorganic Phosphours	4.9 mEq/l	1.4~2.7	19 mEq/day	50~ 90 (2.5~ 40)
Protein g/dl,	16.5 mEq/l	16~ 20		

( ): normal value of child

Interpretation

Volume	deficit		excess
Electrolyte concentration	deficit	+	excess
Potassium	deficit		excess
Calcium	deficit		excess
Metabolic	acidosis		alkalosis +
Respiratory	acidosis		alkalosis
Plasma-to interstitial fluid shift			
Interstitial fluid-to-plasma shift			

The patient has metabolic acidosis associated with phosphatemia and with low excretion of salt in urine.

This is consistent with advanced renal dysfunction.

Takahashi

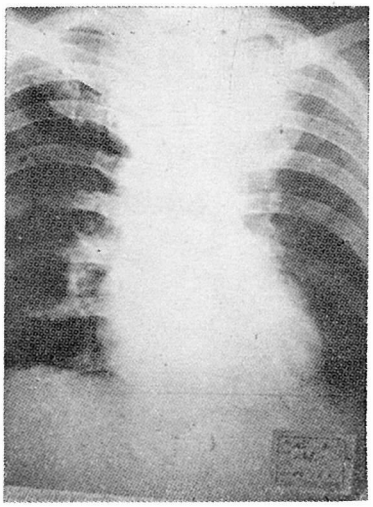

Fig. 4

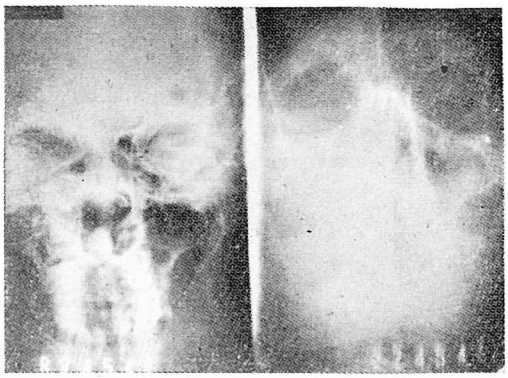

Fig. 5

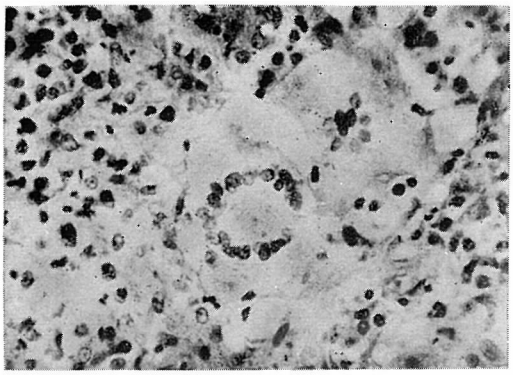

Fig. 6

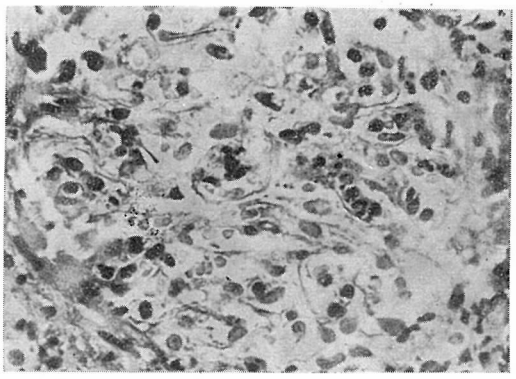

Fig. 7

During admission the patient was given Achromycin, Streptomycin and predonine, without good results. On Feb. 6, 1962 the patient lapsed into a deep coma and died several houral later.

COMMENT

Fahey and associates[3] in 1954 reviewed twenty-two cases of Wegener's granulomatosis collected from the literature, and added seven patients of their own cases. Since then Milner[4], McCallum[5] and Leggat and Walton[6] have reported this disease bringing the tolal to thirty-five cases.

In diagnosis the following diseases should be differentiated; 1) specific infectious granulomatous diseases, 2) sarcoidosis, 3) the rare progressive granuloma gangrenes-

cens involving the nose and face, which is probably closely related to Wegener's granulomatosis although not ordinarily associated with either vascular or renal lesions, 4) polyarteritis nodosa, and 5) allergic granulomatosis and angitis.

Wegener's granulomatosis should be considered a subgroup of polyarthritis nodosa with certain anatomical and clinical peculiarities. Godman and Churg[7] noted several features of Wegener's granulomatosis which differentiate it from the microscopic form of polyarthritis nodosa. In former, there is the peculiar predominant and aggressive charactec of the necrotizing lesions in the respiratory to tract, and renal involnement occurs with impressive regularity and severity. Tissue eosinophilia is not commonly found in Wegener's syndrome, and the clinical stigmas of allergy are usually absent. They comment on the continuous spectrum of tissue changes from pure necrosis and granuloma formation to pure angitis in the various manifestations of polyarteritis and allergic angitis and granulomatosis.

SUMMARY

Wegener's granulomatosis is characterized by necrotizing granulomatous lesions of the upper respiratory tract or lungs necrotizing vasculitis and focal glomerulonephritis terminally usually in uremia. One additional case is reported.

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