

Wegener's Granulomatosis - Case Report

Pages with reference to book, From 285 To 287

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Abstract

Wegener's disease is one of the lethal forms of necrotizing arteritis. A case involving a young man is presented. The purpose of this report is to inform the Clinicians that a diagnosis of Wegener's granulomatosis or any other arteritis should always be considered in a vague presentation which fails to respond to adequate treatment. The patient's life can be improved and the survival rate in proven cases has been proposed from months, now to many years (JPMA 32:286, 1982).

Introduction

Wegener's granulomatosis is one of the clinical syndromes characterised by segmental inflammation and necrosis of blood vessels, chiefly involving the medium and small calibre arteries, especially of the respiratory tract and kidneys and also by clinical manifestations which vary considerably (Walton, 1958; Carrington and Liebow, 1966). At times it is difficult to differentiate between the various types of necrotising arteritis and may present with an overlapping syndrome complex. Hence a judgement can only be made on a clinical basis.

Case Report

A 19 year old male presented himself at Ayub Medical Hospital with a few months history of dysphagia and nasal obstruction. Initial physical examination revealed a young man of weak physique, blood pressure was within normal limits, pulse rate 130/min and temperature 99.0°F. His haemoglobin was 10.5 G/L, total and differential counts were within normal limits, and ESR was 140 mm 1st hr. x-ray of pharynx, larynx and trachea showed an extensive ulcerative lesion involving much of the area even the nasal septum was not spared. X-ray chest showed a large nodular mass on the lower right lobe with central cavitation which was unlikely to be tubercular. Other areas of the lung and heart were unremarkable. A tentative diagnosis of Wegener's granulomatosis (Carrington-Leibow Type-limited form) was made initially because the kidneys were unaffected. The patient was put on steroid therapy with some improvement, over a period of 6 months. The patient ran a downhill course with fever, hemoptysis and slight albumin in the urine. He ultimately expired 6 months after the initial diagnosis was made due to excessive bleeding from the nose and pharynx.

A biopsy was taken from the posterior nasal wall, measuring 0.8X0.3 cm. On microscopic examination, the tissue appeared to be polypoid, covered with stratified squamous epithelium showing focal ulceration, subepithelial necrotic debris, acute and chronic inflammatory cells, necrotizing vasculitis of small and medium sized vessels (Fig. 1)



Fig. 1. Photomicrograph showing acute and chronic inflammation and necrotic vasculitis (arrow) X100 H.E.

and few multinuclear (Fig. 2) giant cells.



Fig. 2. Photomicrograph showing a Giant cell (arrow) X100 H.E.

The entire picture was in consistence with Wegener's granulomatosis/Wegeners vasculitis/Lethal midline granulomatosis.

Discussion

Necrotizing arteritis comprises of a group of syndromes generally separable on the basis of clinical and histological appearance, and by the distribution of the lesion in vascular system. The findings have overlapping features and may vary considerably. An exact categorisation may prove difficult, if not impossible, because of the limited expression of the disease, in some cases. Nevertheless, an attempt at separation of various forms of vasculitis is always appreciable because of differences in prognosis and therapy.

The vascular changes that occur in rheumatoid arthritis (Dixon et al., 1958), systemic lupus erythmatosis (SLE), progressive systemic sclerosis (Norton and Nardo, 1970) and childhood dermatoarthritis (Banker and Victor, 1966), is also considered by many authorities as vasculitis.

Varieties of necrotizing vasculitis and other forms of arteritis are:-

1. Polyarteritis nodosa.
2. Hypersensitivity angiitis.
3. Arteritis associated with serum sickness.

4. Schonlein-Henoch purpura (Anaphylactoid purpura).
5. Wegener's granulomatosis.
6. Allergic granulomatous angiitis.
7. Giant cell arteritis.
8. Vasculitis associated with Systemic Lupus Erythematosis.
9. Vasculitis associated with progressive systemic sclerosis (Scleroderma).
10. Vasculitis associated with rheumatoid arthritis.
11. Vasculitis associated with childhood dermatoarthritis.
12. Takayasu's arteritis.

Wegener's type of arteritis/granulomatosis was first described by Wegener in 1936 and 1939. The disease is characterised by areas of granulomatous necrosis in the upper and lower respiratory tract, vasculitis involving both the arteries and veins, especially of lungs and focal glomerular nephritis (Walton, 1958; Carrington and Liebow, 1966). The disease has aroused great interest because of its relationship with other forms of polyarteritis and various granulomatous diseases of the upper respiratory tract, reported as Lethal Granuloma of midline Facial Structures and Malignant granulomas. The disease affects adults of either sex and is manifested by clinically purulent sinusitis, rhinitis, tracheo-bronchitis and pneumonia. Constitutional complaints, including fever are prominent and may dominate the clinical picture. In addition, there may be transient polyarthralgia or arthritis, as well as polyneuritis, parotitis and evidence of myocardial dysfunction. Anaemia and leukocytosis are generally the rule and eosinophilia is common. An elevation of serum IgA is also seen (Fauci et al., 1971; Israel et al., 1977). The development of renal involvement is usually marked by the appearance, microscopically, of hematuria and leukocyturia. The renal disease tends to progress rapidly and in the past many patients died of uraemia within weeks or months (Nielsen et al., 1967). Carrington and Liebow (1966) have described a limited form of Wegener's disease, in which all manifestations are present, but some of the organs are spared, especially the kidneys. They have labelled it as a 'limited, form of Wegener's Disease. X-rays may disclose one or more nodular densities 1-8 cm in diameter. There may be cavitations in these lesions, representing granulomas. In case the kidneys are involved, there is thrombosis and necrosis of loops or lobes of glomerular capillary tufts (Israel et al., 1977). The prognosis in Wegener's granulomatosis is generally accepted as worse, although limited and less severe forms of the disease may have better prognosis. Corticosteroid therapy is less effective. A number of cases have now been reported in which complete remission has occurred following therapy with alkylating agents and other immunosuppressives. The drug most effective so far has been cyclophosphamide (Raza et al., 1975.).

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