MIDLINE LETHAL GRANULOMA COMPLICATING PREGNANCY: CASE REPORT

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ABSTRACT

A case of midline lethal granuloma in a 28-year-old female Nigerian patient is reported. Oral, ocular and nasal lesions were present and these preceded a spontaneous abortion of a three month old pregnancy. The clinical course of the disease and its similarity to other granulomatous diseases, which are generally classified as midline granuloma syndrome, are highlighted. The prognosis is poor but early diagnosis and treatment appears to improve a patient’s condition.

INTRODUCTION

The term “midline granuloma syndrome” (MGS) is clinically used to describe a broad spectrum of diseases characterised by aggressive and progressive destruction of the mucosa and adjacent structures of the midline and upper respiratory tract (1). Since Wegener (2), first described three patients with a condition starting as rhinitis with disseminated disease and ending in renal failure, a number of cases have been reported in the American and European literature.

The term “Wegener granulomatosis” (WG) has been adopted when the systemic aspect of the disease are recognised (3). Although it was the first recognised cause of these syndromes, its clinical relationship to nasopharyngeal T-cell lymphoma in its early states has been emphasised (2). Some authorities (3,4), recognise that midline lethal granuloma (MLG) and WG are one and the same disease but differentiated by systemic involvement of the latter. The uncertainties surrounding the clinical and pathological presentation of these diseases, which are similar, are demonstrated by these confusing suggestions. These are further compounded by the view of some authorities that frown at the blanket use of the term “midline lethal granuloma” - arguing that not all are lethal and clearly not all are granulomatous (5), nor do they all begin in the midline (6).

The MLG and WG may be rare in Africa. Adekeye (7), in 1977 was only able to find three references to the diseases in Africa. Furthermore, he reported three cases of the MLG and WG in Nigerians. The purpose of this paper was to report a case of midline lethal granuloma complicating pregnancy in the hope that it will also add to the literature of the few cases reported from Africa.
Figure 1
Vasculitis of small arterioles with a chronic inflammatory cell infiltrate of the walls/stroma deep to the edge of the palatal ulcer. H&E X128

No giant cells were seen. These features are consistent with midline lethal granuloma. The patient was immediately commenced on azathioprine 150 mg and 10 mg of prednisolone daily for one week, which was to be reviewed with or without improvement of the patient. There was slight improvement in the patient’s condition as the disease did not progress further after one week of treatment. However, the patient voluntarily discharged herself to seek alternative treatment.

DISCUSSION

The existence of a destructive granuloma of the face whose natural history is different from that seen in the nasal lesion of WG has long been recognised (8). It is a rare disease which was originally thought to be due to an excessive inflammatory reaction but which many now believe to be an atypical form of lymphoma (9). These lesions belong to a group of disorders whose clinicopathological course is similar. They are generally classified as midline granuloma syndrome. Two distinct types are recognised, as WG and the Stewart type (3). The latter lesions have been known by various clinical descriptive terms such as non-healing midline granuloma (NHMG) (9), MLG (7), and malignant granuloma (MG) (4). A third clinicopathologic sub-classification termed idiopathic midline destructive disease (IMDD) has also been described but there are doubts now whether this form exists (1). It has been emphasised that after the exclusion of WG, nearly all the remaining cases presenting as MGS are peripheral sinonasal angiocentric T- and/or NK-cell lymphomas (1). The controversy that arose about these lesions belong to a group of disorders whose pathological entity of lethal midline granuloma: a case report. The poor prognosis of MLG is well known. The need to diagnose the disease early, correct biopsy technique, the use of T-cell markers where available to eliminate lymphomas, aggressive and appropriate use of steroids and cytotoxic chemotherapy treatments are recommended.

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REFERENCES