CLINICAL REPORTS

Idiopathic midline destructive disease—case report and review of the literature

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Summary

A case of idiopathic midline destructive disease (IMDD) in a 39-year-old male is described. Clinical features and diagnosis are discussed. The value of radiotherapy in management is highlighted.

KEY WORDS: midline granuloma syndrome, radiotherapy.

Introduction

Midline granuloma includes a wide spectrum of bacterial, fungal and neoplastic diseases in addition to lesions of unknown origin (Table 1). A subgroup with distinct clinicopathological features—idiopathic midline destructive disease (IMDD) has recently been identified (Fauci, Johnson and Wolff, 1976). The entity is characterised clinically by destructive lesions always localised to the upper respiratory tract and pathologically by non-specific acute and chronic inflammation with variable amounts of necrosis. Malignant or atypical cells are invariably absent and no infectious agent can be identified by culture or special staining. Appropriate diagnosis of this disorder is of paramount importance as, although uniformly fatal if left untreated, local radiotherapy with relatively high dosage has resulted in long-term clinical remission (Tsokos, Fauci and Costa, 1982).

Case report

A 39-year-old male presented with a 5-month history of nasal stuffiness, purulent nasal discharge and increasing swelling and ulceration of the anterior aspect of the nose. Pain developed over the maxillary sinuses in the 3 months before admission. Antibiotics failed to affect the clinical course. The patient had no systemic symptoms or relevant past history.

Physical examination showed an afebrile man with

TABLE 1. Diseases which may present clinically with the "Midline granuloma" syndrome.

Infectious diseases
Bacterial
Fungal
Parasitic
Inflammatory diseases of unknown aetiology
Wegener's granulomatosis
Idiopathic midline destructive disease
Neoplastic
Squamous cell carcinoma
Lymphoma
Polymorphic reticulosis
Rhabdomyosarcoma

an erythematous, tender ulceration on the tip of his nose, involving the left side and inferior aspect of the nasal septum (Fig. 1). The lesion was covered with yellow purulent material. No other abnormality was detected.

Laboratory investigations showed normal haemoglobin, white cell count and differential. The erythrocyte sedimentation rate (ESR) was 16 mm/hr (Westergren method). Serum electrolytes, liver enzymes, urinalysis, serologic test for syphilis, serum protein electrophoresis and chest X-ray were all normal, as was a bone marrow examination. Sinus X-ray revealed mucosal thickening of both maxillary antra. Soft tissue swelling of the left nostril without underlying erosion was also demonstrated.

Biopsy of the nasal mucosa showed non-specific acute and chronic inflammation with a variable amount of necrosis. There were no atypical cells and no evidence of vasculitis. Staining and culture of the material for fungi and tubercle bacillus were negative.

FIG. 1. Ulceration of nose in patient with IMDD.

On the basis of the clinicopathological findings, a diagnosis of IMDD was made. The patient was managed by radiation therapy and was given 28 treatments to a total dose of 50 Gy. (5,000 rads). This was well tolerated and resulted in regression of the nasal ulceration.

Discussion

IMDD is a relentless progressive localized, destructive inflammatory process. It predominantly involves the nose, paranasal sinuses and palate with erosion through contiguous structures, particularly the face. The majority of patients present with pansinusitis and destructive lesions of the nasal septum or hard palate (Fauci *et al.*, 1976). Untreated, the disease is uniformly fatal (Lober, Kaplan and West, 1982) with death occurring, usually after an extended illness, from meningitis secondary to erosion to the meninges, haemorrhage, sepsis or inanition. Both sexes may be affected, and the mean age of onset is 35 years. Although the cause is unknown, it has been proposed the disorder results from a fulminant inflammatory response to an unknown antigen (Walton, 1959). The development of hypersensitivity type skin reactions in two cases and disseminated inflammatory and vasculitic disease in a third is in keeping with this hypothesis (Fauci *et al.*, 1976; Tsokos *et al.*, 1982).

Diagnosis is essentially one of elimination (Fauci *et al.*, 1976). There must be no evidence of generalised disease, either inflammatory or neoplastic. A local infectious agent must be excluded by staining and culturing biopsy material for bacteria, mycobacteria and fungi. The presence of lymphoma and carcinoma must be sought by histological examination of multiple and deep tissue samples (Lober *et al.*, 1982). Polymorphic reticulosis, regarded by some as an atypical lymphoma (Batsakis, 1979) should also be included in the differential diagnosis. Histologically it shows pleomorphic lymphoreticular cell proliferation with a perivascular pattern of growth and the presence of atypical cells. In contradistinction, the infiltrate in IMDD is composed exclusively of inflammatory cells (Tsokos *et al.*, 1982) and infiltration of vessel walls, which is characteristic of polymorphic reticulosis, does not occur. In addition, polymorphic reticulosis is not always restricted to the upper respiratory tract.

Distinction between IMDD and Wegener's granulomatosis is of paramount importance as management differs. Clinically, Wegener's granulomatosis is not as destructive in its upper airway manifestations, virtually never eroding through the soft tissues of the face or palate. Unlike IMDD, it is a truly systemic disease (Wolff *et al.*, 1974). Pathologically, the characteristic feature of Wegener's granulomatosis is diffuse small vessel granulomatous vasculitis (McDonald, DeRemee and Weiland, 1981) this rarely if ever, occurring in IMDD. Interpretation of the histological findings in conjunction with the clinical presentation should therefore allow separation of these two entities.

The value of relatively high-dose local radiotherapy (4,000–5,000 rads in 5 weeks) to the upper airways in the management of IMDD has recently been realised (Fauci *et al.*, 1976; Tsokos *et al.*, 1982; Fitzgerald, 1982). Such treatment has resulted in some fatal central nervous system complications and, in a number of patients, second malignancies have developed. Some of these side effects should be reduced by careful radiotherapy planning. Although initially causing improvement, low dose radiation (1,000 rads in 10 days) is followed by re-activation and progression of tissue destruction (Fauci *et al.*, 1976). Response to chemotherapy is unsatisfactory, steroids are of no value (Harrison, 1974), and surgical removal of involved tissue may cause progression of the lesions (Casuccio and Yamagisawa, 1981). Reconstructive and plastic surgery, which may be of cosmetic and functional value, should be delayed for at least 1 year following completion of radiotherapy (Casuccio and Yamagisawa, 1981).

In summary, subgroups of patient presenting clinically with a midline granuloma syndrome can be recognised. Distinction and characterisation of these

diseases is essential for appropriate management. Cytotoxic therapy has been shown to be highly effective in the treatment of Wegener's granulomatosis (Fauci and Wolff, 1973).

When tumour is identified, treatment is dictated by histology and extent of involvement. An infectious aetiology requires anti-microbial agents. Finally, there is the entity called IMDD in which radiotherapy is of proven benefit.

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(Accepted 25 May 1983)