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Acute Sjögren-like syndrome as the first manifestation of a generalized CMV infection in a patient with AIDS

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Abstract
We report the first case of generalized cytomegalovirus (CMV) disease in an AIDS patient who presented with an acute Sjögren-like syndrome and was diagnosed by parotid gland biopsy. All symptoms disappeared after a few days of intravenous ganciclovir therapy.

Key words: Acquired immunodeficiency syndrome; Cytomegalovirus; Parotid gland

Introduction

Cytomegalovirus (CMV) infection is almost universal among HIV-infected subjects. Asymptomatic viraemia and viruria are frequently observed in AIDS patients. However, only a few of them have specific organ involvement and need to be treated. Retinitis is the most commonly recognized disorder but oesophageal, intestinal, colic and lung involvements are also described (Schooley, 1990). We report on an acute parotitis which was the first clinical manifestation of a generalized CMV infection in a patient with AIDS.

Case report

A 30-year-old patient known to be HIV seropositive since 1987 presented with a swelling in the parotid area which was observed three days previously. Salivation became painful and a dry mouth was noted.

Significant medical history included an oesophageal candidiasis successfully treated with fluconazole. He had been treated with zidovudine for 18 months; didanosine had been started four months previously. Dapsone was given as a prophylaxis against *Pneumocystis carinii* pneumonia. The physical examination was normal except for obvious signs of bilateral parotid gland enlargement. Stensen duct orifices were erythematous but a purulent discharge was not observed. No cervical adenitis was present. His body temperature was 38°C. Laboratory findings disclosed a 45/mm^3 CD4$^+$ cells count and an elevated serum amylase level at 1280 I.U. (normal <150 I.U.), 90 per cent being specifically of salivary origin; antinuclear antibodies were absent. Echography demonstrated no sialolithiasis and salivary ducts were not dilated.

A bacterial origin was suspected and clindamycin 600 mg Qid was given. After a 14-day interval, the symptoms were still present and the patient had progressively lost 8 kg. A biopsy of the gland was performed. Histological examination of the fragment disclosed typical CMV intracellular inclusions. The presence of CMV was confirmed by specific immunoperoxidase staining. Viral culture of the biopsy yielded CMV. Bacterial, mycobacterial and fungal cultures remained negative. The patient was admitted to hospital. He had had CMV IgG antibodies detected a few months before this episode. CMV IgM antibodies were repeatedly negative. CMV viraemia and viruria were demonstrated. Ophthalmological examination identified asymptomatic peripheral retinal lesions and an obvious keratoconjunctivitis sicca syndrome.

Intravenous ganciclovir 5 mg/kg BID was started. After a few days, the parotid swelling disappeared and the patient could eat without pain. The therapy was continued for three weeks. Viraemia and viruria disappeared, and ophthalmological lesions showed progressive healing so that only one 5 mg/kg perfusion was continued when he was ambulatory. This same treatment was still being administered, five months after the acute phase of the CMV infection. His general status was good and the symptoms of dry mouth and eyes were no longer reported by the patient. Didanosine therapy was not changed.

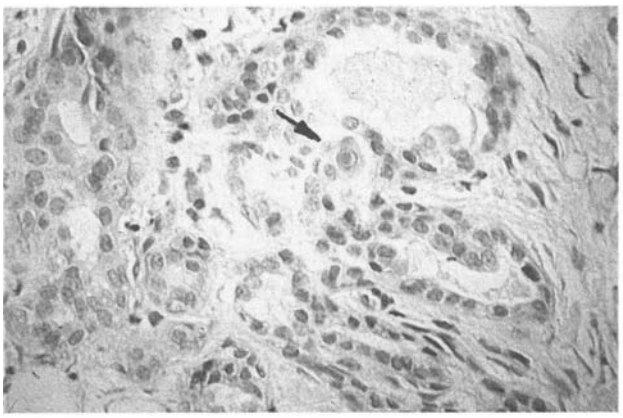

Fig. 1
Biopsy specimen from right parotid gland. Intracellular inclusion (arrowed) is characteristic of CMV involvement.

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Discussion

HIV-infected patients sometimes complain of parotid gland enlargement and associated xerostomia. The causes of these symptoms, much more common in children than in adults, remain unknown. CMV, EBV or HIV infections have been suspected, but anatomopathological clues of specific viral involvement have until now never been obtained. In the presence of a Sjögren-like condition, serological markers are usually positive in true Sjögren disease like rheumatoid factor and antinuclear antibodies are frequently absent in HIV-associated cases. Patients on didanosine therapy sometimes complain of a dry mouth. Neoplastic lesions such as lymphoma and Kaposi's sarcoma are rare (Itin *et al.*, 1993). In this report, considering the acute form of the parotid disease, a bacterial infection was first suggested. The absence of response to antibiotherapy and the rapid alteration of the patient's general status led us to perform a biopsy of the parotid gland. CMV involvement was assessed by histological examination and culture. The diagnosis was further confirmed by observing the complete relief of the Sjögren-like syndrome symptoms after only a few days of ganciclovir treatment.

CMV inclusions are frequently observed on histopathological examination of salivary glands in autopsies of unselected non-immunodeficient young children, parotid glands being mostly involved. By contrast in subjects older than 10 years, the presence of CMV is not demonstrated even by culture. These data suggest that active CMV infection of the salivary glands develops early in infancy. The infection becomes latent but CMV persists in many organs including possibly the parotid tissues. This infectious cycle has been studied in animal models showing that immunodepression results in reactivation of the virus present in the reservoir organs (Ho, 1991). Most of the AIDS patients have serologic markers for past CMV infection and the frequent isolation of CMV from organs or blood cells reflects reactivation of the latent virus (Drew, 1988). Parotid infection when present is reported to provoke an asymptomatic excretion of CMV in the saliva (Griffiths *et al.*, 1988). Pialoux *et al.* (1991) reported a generalized CMV infection in an AIDS patient who presented a symptomatic CMV involvement of the submandibular gland. We have demonstrated that CMV can also cause acute reversible parotid and lacrymal damage.

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