

Papillitis as the prominent ocular sign in Acquired Immune Deficiency Syndrome (AIDS)

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SUMMARY

A 29-year old homosexual presented with clinical symptoms and an immunological picture of AIDS syndrome.

Ocular involvement started in August 1986 with reduction of visual acuity in the right eye rapidly progressing to amaurosis. The most prominent ophthalmoscopic sign was of papillitis which had, in the beginning, the characteristics of an ischaemic optic neuropathy. Besides this, cotton-wool spots, retinal haemorrhages and limited areas of Cytomegalovirus (CMV) retinitis were found. Choroid was also involved with secondary CMV retinitis. On the other hand, sheathing of retinal vessels and Roth's spots were absent.

Although papilloedema, haemorrhages, cotton-wool exudates and CMV retinitis completely disappeared by October 1986, the general condition aggravated and the patient finally succumbed.

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INTRODUCTION

Acquired Immune Deficiency Syndrome (AIDS) represents a serious epidemiological problem on the increase. The risk factors and characteristics of this syndrome of diseases are well known and shall not be repeated here. This syndrome may appear also in heterosexual populations, as in Africans^{1 2 3}.

Reported ocular manifestations of AIDS vary according to the literature⁴. Generally the eye is involved in 55 to 100% of cases.

The most signs are cotton-wool spots (30 - 92%), Roth spots and haemorrhages (8 - 53%), perivascular sheathing (3 - 15%), and cytomegalovirus infection (0 - 46%). Occasionally a macular cherry-red spot is noticed. Kaposi's sarcoma of the conjunctiva has also been reported (0 - 10%). Cytomegalovirus may also cause acute retinal necrosis⁵.

Cotton-wool spots and haemorrhages represent the non infectious signs of AIDS retinopathy⁶.

The choroid is involved secondary to CMV retinitis⁷, but if the anterior segment of the uveal tract is rarely involved.

Bilateral closure glaucoma may appear in AIDS, the aetiology is a choroidal effusion with secondary anterior rotation of the ciliary body⁸.

Toxoplasmosis of the central nervous system may produce third and sixth nerve palsies⁵.

It is important to note that fundus findings and the patient's general condition do not correlate^{5 6}.

Human Immunodeficiency Virus (HIV), was isolated from two corneal specimens taken from carriers of the disease. This may prove to be a possible medium for transmission in keratoplasties^{9 10}.

Oedema of the optic nerve together with other fundus changes have been mentioned¹¹ however papillitis, as the most prominent fundus sign from the beginning, has not emphasised. It is for this reasons that our patient is of particular interest.

CASE PRESENTATION

A.B. born in 1957 had his first homosexual

relationship at 16 years of age. Between 1981 and 1983 he lived in Australia. There he had many homosexual partners, one of whom was his steady partner. When he left Australia he was in perfect health. In 1985 he was informed that his steady partner had died from AIDS. This brought him for a check up and he was found to be HIV antibody positive.

A.B. was asymptomatic up to March 1986. His initial complaint was dryness of the skin followed by sore throat, dysphagia, cough, diarrhoea, low grade fever and loss of weight — 10kg. in 3 - 4 months.

Microbiological examination performed in March 1986 showed oesophageal candidiasis and salmonella gastroenteritis. Dermatological manifestations included acquired ichthyosis. He also developed generalised lymphadenopathy but had no splenomegaly.

Antifungal therapy with ketoconazole was initiated and he recovered from his oesophagitis. He continued to lose weight and the skin became dry.

A reddish palatal plaque was biopsied in July 1986 and was histologically diagnosed as Kaposi's sarcoma. (Fig. 1.)

Colonoscopy performed in August 1986, because of his enteritis, showed granulomatous inflammation with acid fast bacilli which were most likely atypical mycobacteria. (Fig 2.)

LABORATORY INVESTIGATIONS:

— Stool culture: positive for Salmonella

— Chest X-Ray, mid stream urine for culture and sensitivity - all within normal values. ▶

- ▶ — Blood: Haemoglobin 7,6 g/dl., white blood cells count $3,3 \times 10^9/L$ (leucopenia). Platelets $189 \times 10^9/L$. Neutrophils 50% (neutropenia). lymphocytes 44% (relative lymphocytosis). Coagulation screen, liver profile within normal limits.

SEROLOGICAL INVESTIGATIONS

- HTV-III positive since 1985.
- Cytomegalovirus (CMV): positive — IGM antibody titre
- Paul Bunnell test for infectious mononucleosis: negative
- Hepatitis B surface antigen (HBsAg): negative
- Skin test with tuberculin (P.P.D.): negative — anergy

Previously of known perfect eyesight, A.B. presented in August 1986 with rapidly progressing blurring of vision from the right eye. In a period of a few days his visual acuity was reduced to counting fingers at 0.5m.

Ocular examination on 16 September 1986 showed his right eye was quiet with a normal anterior segment and clear media. The fundus, (fig. 3), showed an oedematous optic disc with a 2 dioptre prominence. It was surrounded by haemorrhages superimposed on the retinal vessels. Three cotton-wool spots covered the vessels superior to the optic disc. In the papillo-macular area was a diffuse whitish retinal oedema (CMV retinitis).

Small white spots in the deeper layers of the retina were seen in the area intervening between the temporal vascular arcade. These did not have an exudative appearance. Except for three cotton wool spots the retinal periphery was normal and no sheathing of the retinal vasculature could be seen. The macula was without oedema.

The left eye was completely normal with normal visual acuity.

Visual field on the right eye could not be correctly determined, due to poor vision, while that of the left eye was normal.

The patient was re-examined twice on the 11th and subsequently on the 16th October 1986. During his last ocular examination the findings were: The right eye was quiet. Dust-like opacities were noticed in the vitreous. The fundus (fig. 4) showed a complete disappearance of the papilloedema. The disc however was pale, indicating optic atrophy. The cotton-wool spots and retinal haemorrhages also disappeared. Superior to the optic disc a zone of chorioretinal scarring scattered with pigment dots remained.

The papillo-macular area which was previously oedematous was now of a greyish colour. The whitish spots located in deeper retinal layers, between the temporal vascular arcade remained unchanged.

This eye was amaurotic. The left eye remained normal.

Although the evolutive pathologic signs on the fundus had completely regressed the general condition of the patient deteriorated rapidly and he succumbed to his illness a few days later. A post mortem was not performed.

DISCUSSION

A.B. was definitely in the AIDS risk group. He was a carrier of HIV as detected in 1985 by a positive HIV — antibody titre. In March 1986 he developed symptoms of his illness.

The most prominent ocular sign was his papilloedema with rapidly deteriorating sight, indicating papillitis. The oedema looked like ischaemic oedema of optic neuropathy.

Haemorrhages and cotton-wool spots, present in up to 92% of AIDS patients' were

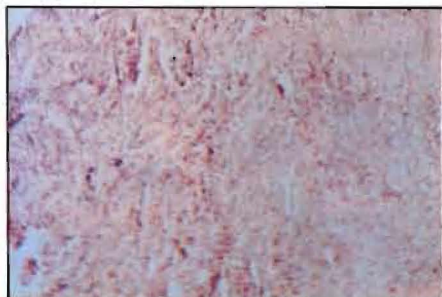


Fig. 1. Kaposi's sarcoma of the palate. Staining Haematoxylin - Eosin. Magnification x 320. (By courtesy of the Pathology Department).

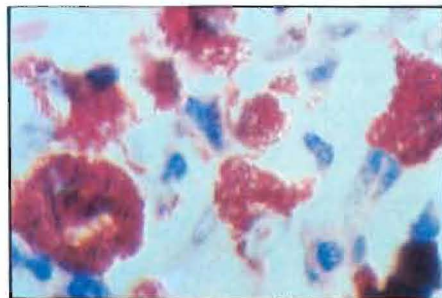


Fig. 2. High power view showing acid cast bacilli of atypical Mycobacterium from the granulomatous inflammation of the colon. Gram Staining. (By courtesy of the Pathology Department).

present but these disappeared without trace. Roth's spots were absent. Although CMV retinitis is said to be the most prominent ocular manifestation, in our patient, this was not as pronounced as the retinal oedema was limited to the peripapillary and interpapillomacular area. The retinal oedema had disappeared in the end leaving an atrophic area with irregular pigmentation.

Ophthalmoscopically no sheathing was visible but this does not imply that small retinal capillaries and those around the disc were not involved. The poor general health did not allow for a fluorescein angiogram by which other authors¹² have demonstrated non-perfusion areas.

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Fig. 3. Papillitis with "cotton-wool" spots, haemorrhages and CMV retinitis in interpapillomacular area. Note the absence of perivascular sheathing.

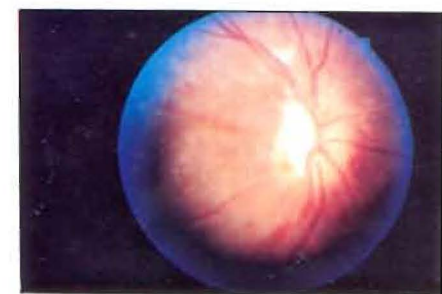


Fig. 4. Optic atrophy one Month after disappearance, of papilloedema, "cotton-wool" spots, haemorrhages, and retinal oedema. No signs of perivascular sheathing.

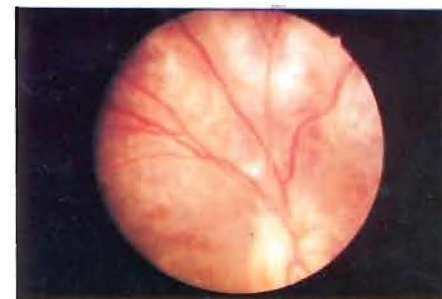


Fig. 5. Chorioretinal atrophic scars after complete disappearance of the retinal oedema indicating the involvement of the choroid secondary to the CMV-retinitis.

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As a result of the altered immunological status it is supposed that the immune complexes may affect small vessels. In our case we suppose that the small vessels of Haller's ring, around the optic disc, are affected. Fluorescein angiography is reported to have shown¹² focal non perfusion and microvascular changes in the retina before the involvement of the optic disc and the retina by CMV.

Sheathing of large retinal vessels probably represents a secondary manifestation of CMV retinitis. Its absence in our patient could be explained by the CMV retinitis being localised to a relatively small area.

Changes in the vitreous occurred at a later date and were not very pronounced.

Involvement of the uveal tract in AIDS is of special importance. In our patient the choroid was involved secondarily. When the CMV retinitis disappeared chorioretinal scars were noted indicating that both retina and choroid were involved (fig. 5.) Some have found by histological examination that the subjacent choroid may be infiltrated with both acute and chronic inflammatory cells⁷.

CONCLUSION

In many publications the frequency of cotton-wool spots, retinal haemorrhages, Roth's spots and CMV retinitis in AIDS have been pointed out. In our patient, however, papillitis was from the beginning the prominent manifestation in the fundus besides CMV retinitis. Initially the optic

disc had the characteristics of ischaemic optic neuropathy. For this reason we may suppose that the primary lesions were located in the small vessels supplying the optic disc.

There are no correlation between fundus changes and the general condition, as evidenced at the end when the former signs regressed whilst the patient deteriorated.

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