LOCALIZED INTRA-ORBITAL AMYLOID DISEASE

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AMYLOID disease or amyloid degeneration used to be met with much more commonly before the present era of medicine, in patients suffering from prolonged toxic states, particularly syphilis, tuberculosis and chronic sepsis. Chronicavitaminosis, mainly deficiency in the vitamin B complex, has been held by many to be a factor in the production of this degeneration.

With the present advance in prevention and treatment of such prolonged toxic states and with the routine administration of vitamins to such patients, this form of degenerative condition has greatly diminished and is now much more rarely seen.

It occurs both in a generalized and in a localized form. In the generalized form more than one organ is involved, usually together with the liver, spleen, and kidneys, while in the localized form, the disease is limited to one organ and is occasionally given the name “amyloid tumour”. The latter variety is known to occur as a primary process in the tissues of the larynx and also as a secondary degeneration in a pre-existing tumour; it may also occur in areas of chronic inflammation.

Case Report

The following is an instance of amyloid degeneration in an inflammatory mass in the orbit, causing proptosis.

A man, aged 45, complained of left proptosis of 13 months’ duration (Fig. 1). He gave a history of an attack of inflammation of both eyes 3 years previously which left him with blurred vision; 13 months before he was seen by me, he became conscious of a painless mass in the inner part of the left orbit and since then noticed that his left eye was gradually protruding.

During the last 4 years he had had dental sepsis because of which several teeth were extracted, and he was left with a discharging oro-maxillary fistula in the right upper jaw.

He had no history of syphilis, and had not suffered from diplopia.

Examination.—The left eye-ball was completely fixed and protruded forwards and outwards. A soft mass was seen and felt in the upper and inner quadrant of the left orbit, distending the upper eye-lid in that region and appearing to be the cause of the proptosis. It was not tender and the skin moved freely over it, but the mass was fixed to the bone and neighbouring soft structures.

Fig. 1.—Patient’s appearance when first examined.
INTRA-ORBITAL AMYLOID DISEASE

The bony edges of the orbit were intact. There was a nebula on each cornea; the pupils were regular and active.

Exophthalmic Measurements: Right 22, Left 30.
Ocular Tension: Normal.
Visual Acuity: Right—Counting fingers at 2 m.
Left—Counting fingers at 1 m.
Fundi: Not easily seen because of the corneal opacities.

There was marked chronic right maxillary sinusitis associated with a large right oromaxillary fistula; the nasal passages were free, the ethmoid regions appeared clear on both sides.

General examination revealed no abnormality. The urine and blood pictures were within normal limits. Serological tests for syphilis were normal before and after provocation.

Radiological study of the sinuses and orbits revealed:
(1) Opaque right maxillary antrum, communicating with the mouth cavity by a fistula (oromaxillary fistula.)
(2) Generalized widening of the left orbit (Left primary orbital dilatation); the orbital margins being intact.

On exploration, the intra-orbital mass could not be freed from the neighbouring structures, and gave the impression of being malignant. A piece was removed for histopathological examination, and the late Prof. Bernard Shaw reported as follows:

The histological sections, many of which have been examined, show no evidence of neoplasia. The tissues present a very unusual and unfamiliar appearance. There is a curious hyalinosis and swelling of the collagen around the blood vessels in the orbital fibro-adipose tissue (Fig. 2) and this is associated with aggregations of lymphocytes. Occasional foreign body giant cells are seen attacking the hyaline material. The appearances are those of localized amyloidosis, commonly known as "amyloid tumour".

With this finding, I was content to treat the septic foci, leaving the mass untouched.

![Fig. 2.—Photo-micrograph of biopsy. The thick-walled blood vessels show marked homogenous hyalinosis with narrowing of the lumina, due to amyloid infiltration. ×114](image)

Comment

This most unusual finding stresses the value of routine histopathological study in intra-orbital surgery. This "tumour" could easily have been mistaken for malignant disease, and the treatment given would have meant complete loss of the left eye, which could ill be spared.

We are much indebted to the late Prof. Bernard Shaw for the care he used to take in the study of our biopsies. Our thanks are also due to Dr. Marey for his radiological study of this case.