Fuchs' heterochromic uveitis and sarcoidosis

Editor—Richard Goble and Philip Murray provide further interest in the possible cause(s) of Fuchs' heterochromic uveitis (FHU), in reporting five patients with suggested sarcoidosis, including four with raised serum angiotensin converting enzyme (ACE). In patients with sarcoidosis, increased production of ACE is attributed to activated cells within granuloma. In 'granulomatous' uveitis it is reasonable, even in the absence of systemic symptoms, to investigate for the possibility of sarcoidosis. However, it is rarely appreciated how occult the occurrence of this can be for this purpose. We therefore rely on indirect methods of diagnosis.

Weinreb measured ACE levels in normal controls and found that 4.2% had significantly raised levels. By comparison, of those with 'granulomatous' uveitis but without evidence of systemic sarcoidosis, 44% had raised levels. His conclusion, that 'ocular sarcoidosis' may be diagnosed in the absence of systemic evidence, is difficult, though unproved, and would explain a large subset of idiopathic uveitis. However, to extend this group to include forms of uveitis which are untypical of sarcoid related uveitis is mere speculation. Iris nodules and keratic precipitates of FHU, yet their appearance (small, dome-shaped, multiple, and translucent) and position (on the anterior iris surface, mostly peripupillary, scattered symmetrically) differentiate them from nodules seen in granulomatous disease (usually larger, fewer, often irregular in shape, sometimes buried within the stroma). Their presence in FHU cannot per se make a convincing case for a granulomatous process. However, though unproved, this would explain a large subset of idiopathic uveitis.

At some points in their disease each of our five patients showed the characteristic clinical features of FHU. All patients had uniformly distributed, stellate keratic precipitates (KPs), iris stromal atrophy with heterochromia, and no posterior synechiae. Four out of five patients had iris nodules and three had posterior subcapsular cataract. Although mutton fat KPs are not a recognised feature of FHU, very occasionally these patients can develop a super-added anterior uveitis resulting in the formation of atypical KPs.

Sarcoidosis can be difficult to diagnose particularly in the absence of extraculcular features. Although elevated angiotensin converting enzyme (ACE) levels are not specific for sarcoidosis the uveitis in these patients would be highly unlikely to result from any of the other causes of a raised ACE. Also, one patient was Kveim positive and another had chest x ray changes compatible with sarcoidosis.

We agree that a larger series of patients would be required in order to demonstrate any statistically significant association between raised ACE levels and FHU. Nevertheless, the finding of a raised ACE in patients with this condition that forms only 3% of all uveitis entities would appear to be more than just coincidental.

Ocular sarcoidosis may not always present with the typical textbook findings, an example of this would be those patients who have a fundal appearance similar to birdshod retinochoroidopathy but are HLA-A29 negative. The features of FHU seen in our patients may be another atypical presentation.

Although FHU has been reported in combination with numerous conditions, a possible association with sarcoidosis has not been previously described. We felt that this was an interesting new finding which would support the theory that FHU may be a secondary phenomenon or a clinical end stage of a number of conditions.

Determining the importance of eye diseases in Africa

Editor.—In Africa the public health importance of trachoma and xerophthalmia is often underestimated when based on routine surveillance data and even data from population based surveys of low vision and blindness. Surveillance data may under-represent occurrence because both diseases are prevalent in children who rarely complain of it and health personnel, at least in rural areas, if they do not have an eye complaint. Population based surveys may under-represent occurrence if cluster sampling is used as this is a weak technique for detecting diseases with focal distribution. These problems are highlighted below using experiences on estimating the importance of these diseases in Ethiopia.

In 1978–80 the Ethiopian Nutrition Institute and the WHO conducted a countrywide assessment on the reporting of xerophthalmia in health centres and hospitals. The study concluded that the condition was rarely recorded.1 In the early 1980s two foci of vitamin A deficiencies were detected in famine-free areas of Asa', Bale, and Gami Gofa provinces.2,3 Trachoma was also heavily under-reported. In a study of eye conditions at three health centres, where all children under 10 years of age attending the centres for any reason were examined for eye diseases, prevalence of trachoma was 10-fold higher than previously suggested by hospital records.

In 1981 a population based survey of blindness was undertaken.4 The survey found a blindness prevalence of between 1.35% and 1.5% and trachoma was found to be the major cause. However, the survey failed to detect a high prevalence of xerophthalmia in the country. By chance the xerophthalmia foci were not selected when the random sample of clusters were drawn.

Because of the limitation of the methods discussed above health authorities should identify areas where the population is likely to be at high risk, because of the presence of known risk factors for that condition, and then undertake a sample survey of the children within the high risk areas. This should provide a more realistic insight into the magnitude of the problem in specific high risk areas and direct planning for targeted intervention.

References


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G De Sole

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