Senile scleral plaques and senile scleromalacia

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SUMMARY A case of senile scleral plaques is reported in which expulsion of a calcified plaque anterior to the insertion of the medial horizontal rectus muscle gave rise to senile scleromalacia. Histopathological examination revealed a second calcified plaque at the site of the contralateral senile scleral plaque in this eye. This case indicates that senile scleromalacia is a scleral disease *sui generis* which occurs by expulsion of a calcified plaque in advanced cases of senile scleral plaques. Senile scleromalacia has not previously been described as a clinical entity.

Confusion exists in the literature concerning the nomenclature and nature of 2 apparently unrelated scleral lesions which develop with advanced age, namely, *senile scleral plaques* and *senile scleromalacia*. Senile scleral plaques are sharply demarcated slate-grey areas located anterior to the insertions of the horizontal rectus muscles. Senile scleromalacia describes spontaneously occurring scleral holes at the same site. Neither of these lesions is preceded or accompanied by inflammatory symptoms, and there is no association with rheumatoid arthritis or scleromalacia perforans.

The clinical and histopathological findings will be reported in a case of bilateral senile scleral plaques, in which sequestration and expulsion of a calcific plaque in one of the eyes gave rise to the development of senile scleromalacia.

Case report

In March 1973 a woman aged 83 years presented with burning and weeping eyes due to chronic catarrhal conjunctivitis. The right eye showed a sharply demarcated scleral defect anterior to the insertion of the medial rectus muscle in the shape of a vertical ellipse measuring 4×3 mm. A small slate-grey scleral area had developed anterior to the insertion of the lateral rectus muscle. Visual acuity was 4/60. An immature cataract prevented good visualisation of the fundus. The left eye showed a small slate-grey scleral lesion anterior to the insertion of the lateral rectus muscle (Fig. 1).

By October 1973 the nasal scleral defect had enlarged and its borders had become still more

sharply demarcated, giving a punched out appearance. Its temporal border (towards the cornea) was nearly perpendicular, and dark blue uveal tissue showed through the intact thin, normally vascularised conjunctiva over the most temporal part of the defect. The central and medial parts of the defect contained a glassy yellowish-brown transparent hardened plaque, the temporal part of which was elevated from the underlying tissue, while its nasal margin was still attached to the floor and the nasal border of the defect (Fig. 2).

In June 1974 a painful Mooren's ulcer was diagnosed in the nasal part of the cornea of the right eye. The hardened, glassy plaque had disappeared, leaving the scleral defect covered solely by intact conjunctiva through which dark blue uveal tissue was visible. The corneal ulcer progressed despite energetic therapy. Intensive general examination

Fig. 1 Senile scleral plaque in the left eye

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Fig. 2 Development of senile scleromalacia. The temporal part (towards the cornea) of a glassy plaque is elevated from the underlying tissue at the site of a previous senile scleral plaque (October 1973)

revealed no noteworthy abnormality. Neither the patient's history nor her present condition gave any indication of rheumatic disease. On 2 August 1974 the right eye was enucleated because of severe pain caused by the corneal ulcer.

Macroscopical examination showed a kidney-shaped, apparently total scleral defect immediately in front of the insertion of the medial rectus muscle (Fig. 3). Its vertical and horizontal diameters measured 6.5 mm and 4 mm respectively. The borders of the lesion were sharp, while its floor appeared to be formed by dark blue uveal tissue only. A greyish blue scleral area was present in front of the insertion of the lateral rectus muscle, with a vertical diameter of 3 mm and a horizontal width of 1.5 mm. The nasal part of the cornea

showed a large, vascularised thin area, the central margin of which appeared relatively thickened. Horizontal calottes were cut.

Microscopical examination showed 2 symmetrically located scleral lesions on both sides over the pars plana of the ciliary body and the peripheral retina (Fig. 4).

On the nasal side a scleral defect extended from 2.5 mm in front of the ora serrata to 2.5 mm behind the ora (Fig. 5). The smooth floor of the defect was covered by epithelium and was formed by the deepest scleral layers which showed a normal fibrillar structure and birefringence without any sign of hyalinisation or necrosis. The thickness of the defect measured about one-third to one-sixth of the intact sclera. The sclera forming the floor of the defect and its margins showed a total loss of cellularity with a little infiltration by lymphocytes and

Fig. 3 Kidney-shaped, apparently total scleral defect at the site of the lesion represented in Fig. 2

Fig. 4 (a) senile scleromalacia represented in Figs. 2, 3, and 5; (b) site of senile scleral plaque represented in Fig. 6; (c) Mooren's ulcer. (H and E $\times 5$)

Fig. 5 Higher magnification of lesion (a) in Fig. 4. The floor is formed by the deepest scleral layers and is covered by epithelium. No hyalinisation or necrosis is present. (H and E $\times 21$)

Fig. 6 Higher magnification of lesion (b) in Fig. 4. The senile scleral plaque contains a calcified centre, and there is loss of cellularity in the surrounding tissue. (H and E $\times 23$)

plasma cells. The peripheral part of the scleral floor of the defect was covered by scar tissue, while the epithelium had formed a proliferative fold between the scar tissue and the remaining sclera.

The site of the scleral lesion in the temporal part of the sclera corresponded to that of the scleral defect on the nasal side. In the sections its horizontal diameter was about 4 mm with its major part located in front of the ora serrata. The affected sclera showed no thinning, but its centre contained a calcific plaque (Fig. 6), which, despite repeated decalcifying procedures, stained strongly by the von Kossa method and with alizarin red. The scleral tissue all around the calcific plaque showed a total loss of cellularity, normal birefringence, and no hyalinisation.

The nasal half of the cornea contained a large ulcer. Part of its peripheral slope was rather steep but elsewhere it shelved more gradually, while its central margin showed the characteristic overhanging lip of a Mooren's ulcer. The thickness of the overhanging lip was about half that of the cornea, while the lamellar cleft between the overhanging lip and the floor of the ulcer was partially filled by proliferating squamous epithelium in which mitotic figures were present. At one site the stromal necrosis had extended to Descemet's membrane, but generally the membrane was covered by a few stromal lamellae, a layer of heavily vascularised scar tissue of variable thickness, and a rather thick layer of epithelium.

The iris stroma and the ciliary body contained mononuclear inflammatory cells. The lens cortex showed vacuolisation. The posterior segment was not remarkable except for a diffuse intrascleral scattering of calcific granules of the type not infrequently encountered in elderly people.

Discussion

Senile scleral plaques have been described by a great variety of terms namely, 'localised areas of calcareous degeneration in the sclera' (Katz, 1929), 'senile degeneration of the sclera' (Pillat, 1933), 'senile thinning of the sclera' (Kiss, 1934; Graves, 1937, 1939, 1941), 'circumscribed scleromalacia at high age' (Kyrieles, 1939), 'scleral plaques' (Culler, 1939), 'hyaline scleral plaques' (Boshoff, 1942), 'senile hyaline scleral plaques' (Roper, 1945), and 'focal senile translucency of the sclera' (Cogan and Kuwabara, 1959). Since advanced age appears to be the most important predisposing factor and hyalinisation of the scleral tissue does not occur, the designation *senile scleral plaques* seems to be the most appropriate.

Senile scleral plaques are not rare but may escape notice through absence of subjective symptoms. The plaques appear as symmetrical, sharply demarcated, glassy slate-grey coloured areas just anterior to the insertions of the horizontal rectus muscles. Only in one instance has a different site been recorded, Gasteiger (1937) describing a third narrow

plaque in front of the insertion of the inferior rectus muscle. Their shape is that of a vertical angular ellipse, with an average horizontal width of 2 mm and an average vertical height of 5 to 6 mm. The overlying conjunctiva is thin but appears normal for the patient's age. The plaques are translucent, as previously mentioned by Rolandi (1915), Pillat (1933), Gasteiger (1937), and Kyrieleis (1939). This translucency was emphasised by Cogan and Kuwabara (1959), who coined the name 'focal senile translucency of the sclera'. The fact that senile scleral plaques are restricted to the region immediately in front of the insertion of the horizontal rectus muscles is thought to be due to the constant stress and strain on the scleral fibres exerted by muscular action. One of the causes of the transparency of the plaques may be local dehydration (Fischer, 1926).

Histopathology. Histopathological findings in single cases have been reported by Urrets Zavalía *et al.* (1937), Culler (1939), and Kyrieleis (1939). A detailed study of 30 specimens by Cogan and Kuwabara (1959) showed that, contrary to previous reports, the thickness of the sclera at the site of the senile plaques is either normal or slightly increased. Furthermore the plaques were seen to be characterised by early loss of cellularity, as Kyrieleis had already noted, although the collagenous fibres had a normal fibrillar structure and birefringence with no sign of hyalinisation.

Another characteristic feature is calcification in the centres of the translucent areas. Calcified plaques were found in less than half the cases studied, their prevalence in lesions with extensive areas of translucency suggesting that calcification is probably a sequel to the loss of cellularity. In all recorded cases the calcified plaques overlay the pars plana of the ciliary body and were usually composed of calcium phosphate, although in an exceptional case Cogan and Kuwabara (1959) demonstrated calcium sulphate. Findings suggestive of calcification had previously been reported by a number of observers (Rolandi, 1915; Urrets Zavalía *et al.*, 1937; Culler, 1939; Kyrieleis, 1939).

As commented by Cogan and Kuwabara (1959), calcification could be a secondary, and probably late, sequel to the loss of cellularity. My own case demonstrates that sometimes this stage is followed by a further complication, namely, sequestration and expulsion of the calcified plaque, leaving a scleral hole which mimics the clinical picture of scleromalacia perforans. The expulsion of the calcified plaque in the present case was documented by photography. It is interesting that Kyrieleis (1939) was of the opinion that expulsion of the almost completely sequestered intrascleral calcific plaque

in his patient would almost certainly have caused a scleromalacia 'perforans'. The fact that Cogan and Kuwabara could not find scleral indentations or abnormal scleral thinning in any of the cases in their series indicates that expulsion of the calcified plaques is rare. In none of their cases had the senile scleral plaque progressed to the ultimate stage of senile scleromalacia.

Senile scleromalacia is essentially different from scleromalacia perforans, being characterised by (1) advanced age of the patient, (2) location just anterior to the insertions of the horizontal rectus muscles alone, (3) a vertical, irregular, oval or kidney-shaped punched out scleral defect, covered by thin conjunctival tissue and possibly having a glassy, yellowish-grey hardened plaque in the floor, (4) the presence of senile scleral plaques in the ipsilateral and contralateral eye, (5) development by expulsion of a sequestered calcified plaque, (6) no evidence of (rheumatoid) scleral nodules or rheumatic disease, and (7) histopathological finding of a flat intact thin scleral floor covered by a continuous healthy layer of epithelium, and having smooth sharp margins and minimal inflammatory reaction with absence of necrosis. The floor and the surrounding scleral tissue show normal fibrillar architecture and birefringence with loss of cellularity and absence of hyalinisation. In most cases a senile scleral plaque is present anterior to the insertion of the opposite horizontal rectus muscle.

Scleromalacia perforans (van der Hoeve, 1934), on the contrary, is characterised by (1) less advanced age of the patient, (2) location anywhere in the anterior sclera, (3) the emergence of large holes in the sclera at the base of which the uvea appears to be bare, (4) absence of senile scleral plaques, (5) gradual development from a necrobiotic rheumatoid nodule, and (6) characteristic histopathology (Verhoeff and King, 1938; Ashton and Hobbs, 1952; Anderson and Margolis, 1952): the entire thickness of the affected sclera is necrotic so that the defect is devoid of a floor and the margins are ill defined. The necrotic granulomatous inflammation often extends into the underlying uvea and the surrounding sclera. Rheumatoid nodules may be found elsewhere in the sclera.

Despite these differences the early clinical manifestations of senile scleromalacia and scleromalacia perforans may be confusingly similar, both being initially painless with minimal inflammatory reaction. It is probable that many cases of senile scleromalacia have been misdiagnosed as scleromalacia perforans, if only because senile scleromalacia has never been described as a separate entity, despite some authors having stressed the essential differences between senile scleral plaques and sclero-

malacia perforans (Roper, 1945; Anderson and Margolis, 1952).

The present case, however, lends strong support to the prediction made by Kyrieleis almost 40 years ago that senile scleromalacia is a disease sui generis which occurs by expulsion of a calcified plaque in advanced cases of senile scleral plaques.

So far the simultaneous occurrence of senile scleral plaques and Mooren's ulcer has not been reported and is unexplained. On the basis of a single case it is probably safest to refrain from speculation.

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