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ANATOMY

The scleral stroma is composed of collagen bundles of varying size and shape that are not uniformly orientated as in the cornea, and so are not transparent. The inner layer of the sclera (lamina fusca) blends with the uveal tract. Anteriorly the episclera consists of a connective tissue layer between the superficial scleral stroma and Tenon capsule. There are three pre-equatorial vascular layers:

- ** Conjunctival vessels** are the most superficial; arteries are tortuous and veins straight.
- ** Superficial episcleral plexus** vessels are straight with a radial configuration. In episcleritis, maximal congestion occurs at this level (Fig. 8.1A). Topical phenylephrine 2.5% will also constrict the conjunctival and 10% also the superficial episcleral vessels.
- ** Deep vascular plexus** lies in the superficial part of the sclera and shows maximal congestion in scleritis (Fig. 8.1B); a purplish hue, best seen in daylight, is characteristic.

EPISCLERITIS

Episcleritis is a common, usually idiopathic and benign, recurrent and frequently bilateral condition. Females may be affected more commonly than males, except possibly in children, in whom episcleritis is rare; the average patient is middle-aged. It is typically self-limiting and tends to last from a few days up to 3 weeks, but rarely longer. Associated disease, either ocular (e.g. dry eye, rosacea, contact lens wear) or systemic (e.g. collagen vascular disorders such as rheumatoid arthritis, herpes zoster ophthalmicus, gout and others) has been identified in up to a third of patients seen at tertiary centres, with ocular disease the most common. Infectious causes are very rare but a wide range has been reported. Investigation of recurrent cases is as for scleritis (see later).

Simple episcleritis

Simple episcleritis accounts for 75% of cases. It has a tendency to recur (60%), decreasing in frequency with time. Features often peak within 24 hours, gradually fading over the next few days.

- ** Symptoms.** Redness; discomfort ranges from absent (up to 50%) to moderate and occasionally severe, when scleritis should be excluded. Grittiness is common, and photophobia may occur.
- ** Signs.** More than half of cases are simultaneously bilateral.
  - Visual acuity is almost always normal.
  - Redness may be sectoral (two-thirds – Fig. 8.2A) or diffuse (Fig. 8.2B). Often it has an interpalpebral distribution, in a triangular configuration with the base at the limbus.
  - Chemosis, ocular hypertension, anterior uveitis and keratitis are all rare.

- ** Treatment **
  - If mild, no treatment is required; cool compresses or refrigerated artificial tears may be helpful.
  - A weak topical steroid four times daily for 1–2 weeks is usually sufficient, though occasionally more intensive instillation is needed initially or a more potent preparation can be used with rapid tapering. A topical non-steroidal anti-inflammatory (NSAID) is an alternative, though may be less effective.
  - An oral NSAID is occasionally required (e.g. ibuprofen 200 mg three times daily, or occasionally a more potent agent such as indometacin). It is very rare for more aggressive systemic treatment to be required, and this is typically in patients with a known systemic association.

Nodular episcleritis

Nodular episcleritis also tends to affect females but has a less acute onset and a more prolonged course than the simple variant.

- ** Symptoms.** A red eye is typically first noted on waking. Over the next 2–3 days the area of redness enlarges and becomes more uncomfortable.
- ** Signs.** Attacks usually clear without treatment, but tend to last longer than simple episcleritis.
  - A tender red vascular nodule, almost always within the interpalpebral fissure (Fig. 8.3A). Occasionally more than one focus is present.
  - A slit lamp section shows an underlying flat anterior scleral surface, indicating the absence of scleritis (Fig. 8.3B).

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Fig. 8.1 (A) Diffuse episcleritis with maximal vascular congestion in the superficial episcleral plexus; (B) scleritis with congestion of the deep vascular plexus

(Courtesy of P Watson – fig. A; S Chen – fig. B)
can involve adjacent tissues and threaten vision. A classification of non-infectious scleritis is shown in Table 8.1; recurrences tend to be of the same type, though 10% progress to more aggressive disease.

Anterior non-necrotizing scleritis

Diffuse

Diffuse disease is slightly more common in females and usually presents in the fifth decade.

- Symptoms. Ocular redness progressing a few days later to pain that may radiate to the face and temple. The discomfort typically wakes the patient in the early hours of the morning and improves later in the day; it responds poorly to common analgesics.

- Intraocular pressure (IOP) is very occasionally elevated.
- An anterior chamber reaction may be present, but is uncommon (10%).
- After several episodes inflamed vessels may become permanently dilated.
- It is important to exclude other causes of a nodule such as phlyctenulosis (a phlycten is within rather than beneath the conjunctiva) or a conjunctival granuloma.

- Treatment is similar to that of simple episcleritis but is more commonly indicated.

Immune-mediated scleritis

Scleritis is an uncommon condition characterized by oedema and cellular infiltration of the entire thickness of the sclera. Immune-mediated (non-infectious) scleritis is the most common type, and is frequently associated with an underlying systemic inflammatory condition, of which it may be the first manifestation. Scleritis is much less common than episcleritis and comprises a spectrum from trivial and self-limiting disease to a necrotizing process that

Fig. 8.2 Simple episcleritis. (A) Sectoral; (B) diffuse
(Courtesy of JH Krachmer, MJ Mannis and EJ Holland, from Cornea, Mosby 2005 – fig. B)

○ Intraocular pressure (IOP) is very occasionally elevated.
○ An anterior chamber reaction may be present, but is uncommon (10%).
○ After several episodes inflamed vessels may become permanently dilated.
○ It is important to exclude other causes of a nodule such as phlyctenulosis (a phlycten is within rather than beneath the conjunctiva) or a conjunctival granuloma.

- Treatment is similar to that of simple episcleritis but is more commonly indicated.

Fig. 8.3 (A) Nodular episcleritis; (B) slit illumination shows that the deep beam is not displaced above the scleral surface
○ The duration of the disease is similar to diffuse scleritis.
○ More than 10% of patients with nodular scleritis develop necrotizing disease, but if treatment is instituted early superficial necrosis does not occur and the nodule heals from the centre leaving a small atrophic scar.

Anterior necrotizing scleritis with inflammation

Necrotizing disease is the aggressive form of scleritis. The age at onset is later than that of non-necrotizing scleritis, averaging 60 years. The condition is bilateral in 60% of patients and unless

**Signs**
- Vascular congestion and dilatation associated with oedema. If treatment is started early, which rarely happens, the disease can be completely inhibited.
- The redness may be generalized (Fig. 8.4A) or localized to one quadrant. If confined to the area under the upper eyelid the diagnosis may be missed.
- Secondary features can include chemosis, eyelid swelling, anterior uveitis and raised IOP.
- As the oedema resolves, the affected area often takes on a slight grey/blue appearance because of increased scleral translucency (Fig. 8.4B); this is due to rearrangement of scleral fibres rather than a decrease in scleral thickness.
- Recurrences at the same location are common unless an underlying cause is treated.

**Prognosis.** The average duration of disease is around 6 years, with the frequency of recurrences decreasing after the first 18 months. The long-term visual prognosis is very good.

**Nodular**

The incidence of nodular and diffuse anterior scleritis is the same but a disproportionately large number of those with nodular disease have had a previous attack of herpes zoster ophthalmicus. The age of onset is similar to that of diffuse scleritis.

**Symptoms.** The insidious onset of pain followed by increasing redness, tenderness of the globe and the appearance of a scleral nodule.

**Signs**
- Scleral nodules may be single or multiple and most frequently develop in the interpalpebral region close to the limbus (Fig. 8.5A). They have a deeper blue-red colour than episcleral nodules and are immobile.
- In contrast to episcleritis, a slit lamp beam shows an elevated anterior scleral surface (Fig. 8.5B).
- Multiple nodules may expand and coalesce if treatment is delayed.
- Instillation of 10% phenylephrine drops will constrict the conjunctival and superficial episcleral vasculature but not the deep plexus overlying the nodule.
- As the inflammation in the nodule subsides, increased translucency of the sclera becomes apparent.

**Table 8.1 Classification of immune-mediated (non-infectious) scleritis**

<table>
<thead>
<tr>
<th>Anterior</th>
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<tbody>
<tr>
<td>Non-necrotizing</td>
</tr>
<tr>
<td>Diffuse</td>
</tr>
<tr>
<td>Nodular</td>
</tr>
<tr>
<td>Necrotizing with inflammation</td>
</tr>
<tr>
<td>Vaso-occlusive</td>
</tr>
<tr>
<td>Granulomatous</td>
</tr>
<tr>
<td>Surgically induced (can also be infective)</td>
</tr>
<tr>
<td>Scleromalacia perforans (necrotizing without inflammation)</td>
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</tbody>
</table>

**Posterior**
appropriately treated, especially in its early stages, may result in severe visual morbidity and even loss of the eye.

**Clinical features**

- **Symptoms.** Gradual onset of pain that becomes severe and persistent and radiates to the temple, brow or jaw; it frequently interferes with sleep and responds poorly to analgesia.

- **Signs** vary according to the following three types of necrotizing disease.
  - Vaso-occlusive is commonly associated with rheumatoid arthritis. Isolated patches of scleral oedema with overlying non-perfused episclera and conjunctiva are seen (Fig 8.6A). The patches coalesce, and if unchecked rapidly proceed to scleral necrosis (Fig. 8.6B).
  - Granulomatous may occur in conjunction with conditions such as granulomatosis or polyarteritis nodosa. The disease typically starts with injection adjacent to the limbus and then extends posteriorly. Within 24 hours, the sclera, episclera, conjunctiva and adjacent cornea become irregularly raised and oedematous (Fig. 8.7).
  - Surgically induced scleritis typically starts within 3 weeks of a procedure, though much longer intervals have been reported. It may be induced by any type of surgery including strabismus repair, trabeculectomy (Fig. 8.8) and scleral buckling, and excision of pterygium with adjunctive mitomycin C. The necrotizing process starts at the site of surgery and extends outwards, but tends to remain localized to one sector.

**Investigations**

- **Laboratory.** These should be employed as adjuncts to clinical assessment, and evaluation by a general physician or rheumatologist should be considered. Specific tests may
include erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), full blood count (e.g. anaemia related to inflammatory connective tissue disease, eosinophilia for polyarteritis nodosa, atopy or Churg–Strauss syndrome), rheumatoid factor, antinuclear antibodies (ANA), antineutrophil cytoplasmic antibodies (ANCA) and anti-cyclic citrullinated peptide (CCP) antibodies, serum uric acid, syphilis serology, Lyme serology, hepatitis B surface antigen (polyarteritis nodosa) and antiphospholipid antibodies. Investigation for tuberculosis, sarcoidosis or ankylosing spondylitis may be appropriate (see Ch. 11).

- **Radiological imaging.** Chest, sinus, joint and other imaging may be indicated in the investigation of a range of conditions such as tuberculosis, sarcoidosis, Churg–Strauss syndrome, Wegener granulomatosis, ankylosing spondylitis and other conditions.

- **Angiography.** Fluorescein angiography of the anterior segment helps to distinguish necrotizing disease by the presence of non-perfusion, and can be used for monitoring; occlusion is predominantly venular in inflammatory disease, and mainly arteriolar in scleromalacia perforans (see below). Indocyanine green is a more accurate indicator of disease activity.

- **Ultrasonography** can help to detect associated posterior scleritis (see below).

- **Biopsy.** This may be considered in resistant cases, especially if infection is suspected.

### Complications of anterior scleritis

- **Acute infiltrative stromal keratitis** may be localized or diffuse.

- **Sclerosing keratitis,** characterized by chronic thinning and opacification in which the peripheral cornea adjacent to the site of scleritis resembles sclera.

- **Peripheral ulcerative keratitis** is characterized by progressive melting and ulceration (Fig. 8.9), and may constitute a severe risk to the integrity of the eye. In granulomatous scleritis the destruction extends directly from the sclera into the limbus and cornea; this characteristic pattern is seen in Wegener granulomatosis, polyarteritis nodosa and relapsing polychondritis. Peripheral corneal ulceration can occur at any stage of a necrotizing scleritis and, in rare cases, precede its onset. (See also Ch. 6.)

- **Uveitis,** if severe, may denote aggressive scleritis.

- **Glaucoma** is the most common cause of eventual loss of vision. The intraocular pressure can be very difficult to control in the presence of active scleritis.

- **Hypotony** (rarely phthisis) may be the result of ciliary body detachment, inflammatory damage or ischaemia.

- **Perforation** of the sclera as a result of the inflammatory process alone is extremely rare.
Scleromalacia perforans

- Scleromalacia perforans (5% of scleritis) is a specific type of progressive scleral thinning without inflammation that typically affects elderly women with longstanding rheumatoid arthritis, but has also been described in association with other systemic disorders. Despite the nomenclature, perforation of the globe is extremely rare as integrity is maintained by a thin layer of fibrous tissue. Differential diagnosis is from the innocuous scleral hyaline plaque and senile scleromalacia (see below).
- **Symptoms.** Mild non-specific irritation; pain is absent and vision unaffected, and keratoconjunctivitis sicca may be suspected.
- **Signs**
  - Necrotic scleral plaques near the limbus without vascular congestion (Fig. 8.10A).
  - Coalescence and enlargement of necrotic areas.
  - Slow progression of scleral thinning with exposure of underlying uvea (Figs 8.10B and C).
- **Treatment** may be effective in patients with early disease but by the time of typical presentation, either no treatment is needed or progression has been marked.
  - Consistent benefit from any agent has not been demonstrated, though frequent lubricant instillation, local (including topical sodium versenate) or systemic anticollegenase agents, immunosuppressives (including topical and oral, but not periocular injection of, steroids, and topical ciclosporin) and biological blockers have been used.
  - Underlying systemic disease should be treated aggressively.
  - Protection from trauma is important.
  - Surgical repair of scleral perforation (e.g. patch grafting) is mandatory to prevent phthisis bulbi.

Posterior scleritis

Posterior scleritis is a potentially blinding condition in which diagnosis is commonly delayed, with an adverse prognostic effect. The inflammatory changes in posterior and anterior scleral disease are identical and can arise in both segments simultaneously or separately. The age at onset is often less than 40 years; young patients are usually otherwise healthy but about a third over the age of 55 have associated systemic disease.

**Diagnosis**

- **Symptoms.** Pain does not correlate well with the severity of inflammation but tends to be more severe in those with accompanying orbital myositis; photophobia is not a dominant feature.
- **Signs.** The disease is bilateral in 35%.
  - Choroidal folds (see Ch. 14) are usually confined to the posterior pole and orientated horizontally (Fig. 8.11A).
  - Exudative retinal detachment occurs in around 25%; yellowish-brown subretinal exudative material can be mistaken for a choroidal tumour.

**Fig. 8.10** Progression of scleromalacia perforans. (A) Asymptomatic necrotic patch; (B) moderate and (C) severe thinning and exposure of underlying uvea (Courtesy of R Bates – fig. A; C Barry – figs B and C)
• Ultrasonography may show increased scleral thickness, scleral nodules, separation of Tenon capsule from sclera, disc oedema, choroidal folds and retinal detachment. Fluid in the Tenon space may give a characteristic ‘T’ sign, the stem of the T being formed by the optic nerve and the cross bar by the fluid-containing gap (Fig. 8.12).
• MR and CT may show scleral thickening and proptosis.

Differential diagnosis
• Subretinal mass. Alternative lesions include miscellaneous granulomatous conditions and choroidal neoplasia.
• Choroidal folds, retinal striae and disc oedema may also occur in orbital tumours, orbital inflammatory disease, thyroid eye disease, papilloedema and hypotony.
• Orbital cellulitis may cause proptosis and periocular oedema but is associated with marked pyrexia.

Important systemic associations of scleritis

Rheumatoid arthritis

The autoimmune disease rheumatoid arthritis (RA) is the most common systemic association of scleritis, and is characterized by a symmetrical deforming inflammatory polyarthropathy, with a spectrum of possible extra-articular manifestations. Presentation is commonly in the third decade with joint swelling, usually of the hands (Fig. 8.13A). It is much more common in females than males. Rheumatoid factor autoantibodies are present in 80–90%. All forms of immune-mediated scleritis have been described in RA, and the clinical course is often more aggressive than when
Relapsing polychondritis

Relapsing polychondritis is a rare idiopathic condition characterized by small vessel vasculitis involving cartilage resulting in recurrent, often progressive, inflammatory episodes involving multiple organ systems such as the ears, respiratory system (Fig. 8.13C), heart and joints. Presentation is frequently in middle age. Scleritis is often intractable and may be necrotizing or non-necrotizing. Isolated anterior uveitis may also occur.

Polyarteritis nodosa

Polyarteritis nodosa (PAN) is an idiopathic aneurysmal vasculitis affecting medium-sized and small arteries, with a wide range of manifestations across multiple organ systems (Fig. 8.13D). Presentation is in the third to sixth decades, often with constitutional symptoms. The male:female ratio is about 3:1. Ocular involvement may precede the systemic manifestations by several years. About a third of patients have hepatitis B infection. Scleritis is
often aggressive and necrotizing. Peripheral ulcerative keratitis, orbital pseudotumour and occlusive retinal periarthritis are other reported ocular features.

### Treatment of immune-mediated scleritis

- **Topical steroids** do not affect the natural history of the scleral inflammation, but may relieve symptoms and oedema in non-necrotizing disease.
- **Systemic NSAIDs** should be used alone only in non-necrotizing disease; it is often necessary to try a number of different drugs before finding one that provides adequate relief of symptoms. A cyclo-oxygenase (COX)-2 inhibitor may be optimal for elderly patients or if there is a history of peptic ulceration, noting concerns regarding cardiovascular adverse effects for these and some other NSAIDs such as diclofenac.
- **Periocular steroid injections** may be used in non-necrotizing disease but their effects are usually transient; some authorities view them as contraindicated in necrotizing scleritis.
- **Systemic steroids** (e.g. prednisolone is 1–1.5 mg/kg/day) are used when NSAIDs are inappropriate or inadequate (necrotizing disease). Intravenous methylprednisolone may be used for emergent cases.
- **Immunosuppressives** and/or **biological blockers** should be considered if control is incomplete with steroids alone, as a steroid-sparing measure in long-term treatment or for underlying systemic disease. A wide range of drugs may be utilized, including cytostatics (e.g. cyclophosphamide, azathioprine, methotrexate), drugs acting on immunophilins (e.g. ciclosporin, tacrolimus) and others; in necrotizing disease, rituximab may be particularly effective.

### Infectious Scleritis

Infectious scleritis is rare but may present diagnostic difficulty as the initial clinical features are similar to those of immune-mediated disease. In some cases infection may follow surgical or accidental trauma, endophthalmitis, or may occur as an extension of corneal infection.

#### Causes

- **Herpes zoster** is the most common infective cause. Necrotizing scleritis is extremely resistant to treatment and may result in a thinned or punched-out area (Fig. 8.14A).
- **Tuberculous** scleritis is rare and difficult to diagnose. The sclera may be infected by direct spread from a local conjunctival or choroidal lesion, or more commonly by haematogenous spread. Involvement may be nodular (Fig. 8.14B) or necrotizing.
- **Leptospirosis**. Recurrent necrotizing scleritis can occur, even after apparent systemic cure. Nodular disease may be seen in lepromatous leprosy.
- **Syphilis**. Diffuse anterior scleritis may occur in secondary syphilis, and occasionally scleral nodules may be a feature of tertiary syphilis.
- **Lyme disease**, Scleritis (Fig. 8.14C) is common but typically occurs long after initial infection.
- **Other causes** include fungi (Fig. 8.14D), *Pseudomonas aeruginosa* and *Nocardia*.

### Scleral Discoloration

#### Alkaptonuria

In this autosomal recessive condition a defect in homogentisic acid oxidase results in the accumulation of homogentisic acid in collagenous tissues such as cartilage and tendon (ochronosis). Systemic features include dark urine and arthropathy. Ocular manifestations include bluish-grey or black generalized pigmentation of the sclera and the tendons of horizontal recti associated with discrete pigmented globules (Fig. 8.15).

#### Haemochromatosis

The systemic features of haemochromatosis are caused by increased iron deposition in various tissues. Features may be more subtle than the classic triad of a bronze complexion, hepatomegaly and diabetes. Inheritance is autosomal recessive. Dry eye and rusty-brown perlimbal conjunctival and scleral discoloration may develop.

### Blue Sclera

Blue scleral discoloration is caused by thinning or transparency with resultant visualization of the underlying uvea (Fig. 8.16). Major associations are discussed below; rare associations include Marshall–Smith syndrome (accelerated prenatal skeletal maturation and growth), Russell–Silver syndrome (short stature and other features) and Hallermann–Streiff–François syndrome.

#### Osteogenesis imperfecta

Osteogenesis imperfecta is an inherited disease of connective tissue, usually caused by defects in the synthesis and structure of type 1 collagen. There are multiple types, at least two of which have ocular features.

- **Type I** is autosomal dominant. Patients suffer few fractures with little or no deformity, hyperextensible joints, dental hypoplasia, deafness and easy bruising. Possible ocular features include blue sclera, megalocornea and corneal arcus.
- **Type IIA** is either sporadic or inherited in an autosomal dominant manner. Systemic features include deafness, dental anomalies, multiple fractures (Fig. 8.17A) and short limbs, with death in infancy from respiratory infection. Ocular manifestations include blue sclera and shallow orbits.
Fig. 8.14 Infectious scleritis. (A) Focal necrosis due to herpes zoster; (B) nodular tuberculous disease; (C) nodular scleritis in Lyme disease; (D) fungal infection
(Courtesy of R Fogla – fig. B, P Watson – fig. C, C Barry – fig. D)

Fig. 8.15 Alkaptonuria – pigmentation (ochronosis) of the sclera and horizontal rectus tendons

Fig. 8.16 Blue sclera
(Courtesy of P Watson)
Ehlers–Danlos syndrome type VI

Ehlers–Danlos syndrome VI (ocular sclerotic) is an inherited disorder of collagen formation. Patients have thin and hyperelastic skin (Fig. 8.17B) that bruises easily and heals slowly; joints are hypermobile (Fig. 8.17C), which may lead to recurrent dislocation and falls. Cardiovascular disease can be severe, including a bleeding diathesis, dissecting aneurysms, spontaneous rupture of large blood vessels and mitral valve prolapse. There are six major types but type VI and, rarely, type IV, are associated with ocular features. As well as blue sclera, these may include scleral fragility (globe rupture may be caused by mild trauma), epicanthic folds, microcornea, keratoconus, keratoglobus, ectopia lentis, myopia and retinal detachment.

MISCELLANEOUS CONDITIONS

Congenital ocular melanocytosis

Congenital ocular melanocytosis is an uncommon condition characterized by an increase in number, size and pigmentation of melanocytes in the sclera and uvea. The periocular skin, orbit, meninges and soft palate may also be involved.

- **Ocular** melanocytosis, the least common, involves only the eye. Multifocal slate-grey pigmentation is seen within the sclera and episclera (Fig. 8.18A) – the process does not involve the overlying conjunctival layers, unless there is incidental conjunctival pigmentation. The overlying conjunctiva is mobile over the episcleral pigmentation but the pigmentation itself is intrinsic and cannot be moved over the globe. The peripheral cornea is occasionally involved.

- **Dermal** melanocytosis (one-third) involves only the skin.

- **Oculodermal** melanocytosis (naevus of Ota) is the most common type and involves both skin and eye. Naevus of Ota is bilateral in 5%; it occurs frequently in darker-complexioned races but is rare in Caucasians. There is deep bluish hyperpigmentation of the facial skin, most frequently in the distribution of the first and second trigeminal divisions (Fig. 8.18B). It may be subtle in pale-skinned individuals, when it is best detected under good lighting.

- **Ipsilateral associations**
  - Iris hyperchromia is common (Fig. 8.19A).
  - Iris mammillations are uncommon, tiny, regularly spaced villiform lesions (Fig. 8.19B). They may also be found in
Fig. 8.18 Congenital melanocytosis. (A) Episcleral melanocytosis; (B) cutaneous melanocytosis in naevus of Ota

Fig. 8.19 Ipsilateral associations of naevus of Ota. (A) Iris heterochromia (hyperchromia); (B) iris mammillations; (C) fundus hyperpigmentation; (D) trabecular hyperpigmentation

(Courtesy of B Gilli – fig. A; L MacKeen – fig. D)
Idiopathic sclerochoroidal calcification is an innocuous, age-related condition that usually involves both eyes of an affected older adult.

- **Signs.** Geographical yellow–white fundus lesions with ill-defined margins (Fig. 8.20A), often multiple and located in the superotemporal or inferotemporal mid-periphery associated with the vascular arcades (Fig. 8.20B).
- **Ultrasoundography** shows highly reflective choroidal plaque-like lesions with orbital shadowing (Fig. 8.20C).
- **Differential diagnosis** is mainly from osseous metaplasia associated with a choroidal haemangioma, and from neurofibromatosis type 1, Axenfeld–Rieger anomaly and Peters anomaly.

○ Fundus hyperpigmentation (Fig. 8.19C).

○ Trabecular hyperpigmentation (Fig. 8.19D); this is associated with glaucoma in about 10% of cases.

○ Uveal melanoma develops in a small minority of patients, and long-term anterior and posterior segment review is required.
choroidal osteoma, which is usually a single – though often large – lesion that usually (80–90%) involves only one eye.

**Scleral hyaline plaque and senile scleromalacia**

Scleral hyaline plaques are oval dark-greyish, generally sharply demarcated, areas located close to the insertion of the horizontal rectus muscles (Fig. 8.21). Senile scleromalacia refers to a spontaneously occurring irregular, oval or kidney-shaped partial-thickness scleral defect found at the same location, typically with one or more scleral hyaline plaques at the opposite location in the same eye or in the fellow eye; separation of a scleral hyaline plaque to leave an area of scleromalacia has been described. Both these entities typically affect elderly patients, are innocuous, and should not be confused with scleromalacia perforans (see above), which occurs in somewhat younger patients, may be located anywhere in the anterior sclera, is unassociated with hyaline plaques elsewhere and can progress to a full-thickness scleral defect with uveal exposure.