

LETTERS TO THE EDITOR

Microcystic adnexal carcinoma masquerading as a chalazion

EDITOR.—Microcystic adnexal carcinoma (MAC) is a locally aggressive cutaneous tumour first described by Goldstein *et al* in 1982.¹ This tumour has a predilection for the face^{2,3} with only a few cases reported involving the periocular region.⁴⁻⁶ It is a low grade tumour of eccrine sweat gland derivation which behaves aggressively in its growth pattern and recurrence rate. It has also been known as a sclerosing sweat duct carcinoma (SSDC). We report a case of MAC initially masquerading as a benign chalazion.

CASE REPORT

A 66 year old white woman attended the accident and emergency department at the Western Eye Hospital with a 2 week history of red sore eyes. Bilateral blepharitis was diagnosed and a chalazion noted at the medial end of the left lower eyelid, adjacent to the punctum. The only unusual comment made was that the chalazion had "granulation tissue" on its conjunctival surface.

The blepharitis was treated with topical antibiotic and eyelid hygiene and she was listed for minor surgery. However, she had incision of her chalazion elsewhere and did not attend the accident and emergency department for another 18 months when she returned with recurrent symptoms of blepharitis.

On examination, a distorted reddened lower eyelid lump was observed in the exact location of her previous "chalazion" which had apparently remained despite incision. There was some puckering of the anterior lamella with radiating telangiectatic vessels suggesting that it was not a benign granuloma (Fig 1). It was nodular and fibrotic, affecting mainly the posterior lamella, and measuring 8 mm horizontally and 5 mm vertically. The tumour had surrounded the lower punctum and extended towards the medial canthal angle. There were no cilia overlying the tumour. There was no regional lymphadenopathy or periorbital anaesthesia.

An incisional biopsy (6 × 2 × 2 mm) of the involved posterior lamella (tarsal plate and conjunctiva) revealed a MAC (Fig 2). The tumour was subsequently excised completely by two layer Mohs' micrographic surgery, which necessitated excision of the medial half of the lower eyelid, from the medial canthal angle. Reconstruction was by a Hughes' tarsoconjunctival graft, with a free skin graft, with second stage division of the flap and reconstruction of the medial canthal angle 6 weeks later. The final eyelid appearance was satisfactory (Fig 3) and she was not concerned about epiphora, although the lower canaliculus had been lost.

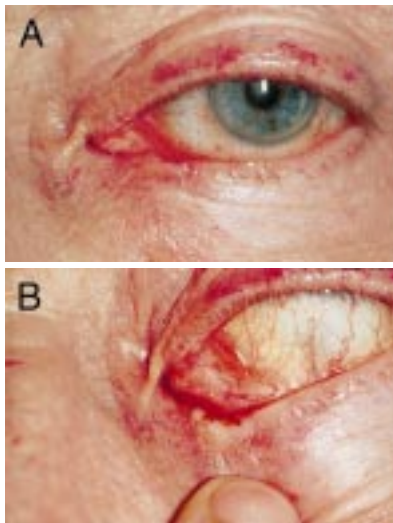


Figure 1 (A) Appearance of left lower eyelid before incisional biopsy. The contour is a distorted by the fibrotic lesion at the medial end of the lid. It is pale with telangiectatic vessels and has indistinct margins. The tumour has no cilia on it. (B) The tumour involves the full thickness of the lid including the posterior lamella. It surrounds the lower punctum and has obliterated its lumen.

The patient's past medical history was notable for systemic lupus erythematosus complicated by renal failure necessitating allograft renal transplantation with prednisolone and cyclosporine immunosuppression for 18 months before her initial presentation with an apparent chalazion.

HISTOPATHOLOGY

The tumour was composed of narrow cords of cells with round or oval vesicular nuclei containing small eosinophilic nucleoli associated with a fairly dense fibrous stroma. Focal ductal differentiation was evident with occasional keratocysts present.

COMMENT

Microcystic adnexal carcinoma is a rare skin tumour which is becoming increasingly recognised. It may be mistaken clinically and histologically for other benign and malignant tumours.² MAC is a malignant sweat gland tumour and has a variety of synonyms which are now in disuse; sclerosing carcinoma of sweat ducts, malignant syringoma, sweat gland carcinoma with syringomatous features, and sclerosing sweat duct (syringomatous) carcinoma (SSDC).⁷ Clinically it presents in the fourth to seventh decade of life with no sex predilection. Up to 90% of these tumours are facial.^{2,3}

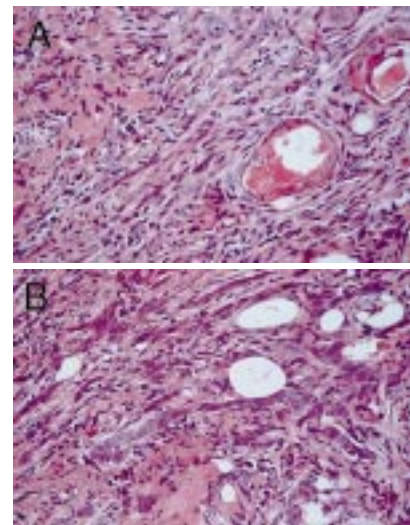


Figure 2 (A) In this field there are two keratocysts. Narrow strands of epithelium are embedded in a dense fibrous stroma (haematoxylin and eosin, original magnification × 25). (B) Ductal differentiation in the form of small cysts are present (haematoxylin and eosin, original magnification × 25).

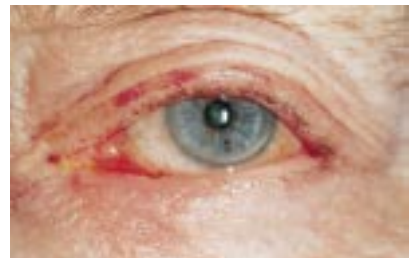


Figure 3 Satisfactory lower eyelid appearance 4 months after tumour excision and reconstruction with Hughes tarsoconjunctival flap and free skin graft to medial half of lower eyelid.

Wide excision by Mohs' micrographic surgery is indicated^{3,8} as the tumour is locally aggressive with perineural, intraneural, and direct spread into adjacent dermis, subcutaneous tissue, muscle, periosteum, and bone.^{5,9} Solid nests of dark or clear staining cells form nests, strands, or cords in a sclerotic stroma. Large biopsies are necessary for these features to be conserved for evaluation at low magnification; small biopsies have been shown to contribute to misdiagnosis.

The aggressive nature of MAC is not initially recognisable from its slow growth and benign clinical and histological appearance. It is more infiltrative with perineural invasion, than basal cell carcinoma.

Table 1 Clinical features of malignant eyelid tumours

Tumour	Characteristics	Treatment	Spread
Basal cell 90%	Nodular/morpheaform, ulcerates/bleeds, slow growth	Excision biopsy, Mohs' excision (2-3 mm margin), radiotherapy, cryotherapy	Local extension, neglected tumours may spread to sinuses and brain
Squamous cell <10%	Ulcerates/bleeds/horn, vascular/velvety surface, rapid growth	Mohs' excision for best cure (3-4 mm margin), relatively radioresistant	Local extension to orbit and sinuses
Sebaceous cell 1%	Firm nodular or pagetoid mass, inflamed eyelid, slow growth	Wide excision (6 mm margin), exenteration if eye involved	Local and distant to lymph nodes, lung and liver
Malignant melanoma <1%	Lentigo maligna or nodular melanoma, bleeds, medium growth	Wide excision (6 mm margin), prognosis depends on depth of lesion, may have skip lesions	Distant spread to lymph nodes, lung, liver, brain

Simple excision alone is insufficient as local recurrences are common (recurrence rate over 40%).^{2,3,7} Late recurrence has been reported, even 30 years after simple excision. Recurrences are greatly reduced with augmented excision and Mohs' micrographic surgery.

This case of MAC differs from others in that it was mistaken clinically by several ophthalmologists for a chalazion. In addition, our patient had been immunosuppressed following renal transplantation, and it is conceivable that this had a role in its pathogenesis. The association of radiotherapy at a distant site and the development of MAC has been reported.¹⁰

In the past MAC has variably been thought to be derived from the hair follicle, the eccrine sweat gland, or even both. Nowadays, however, the tumour is firmly classified within the spectrum of eccrine sweat gland malignancies. A constant finding in this tumour is the presence of ductal differentiation which can be made more obvious with the periodic acid Schiff reaction or by EMA/CEA immunohistochemistry.¹¹

Other malignant tumours occurring around the eyelids are summarised in Table 1. MAC is a rare tumour which may be differentiated clinically from other eyelid tumours by its slow growth, dense fibrous appearance, and slightly indistinct margin without skin ulceration. It is less common than sebaceous cell carcinoma which may also masquerade as a chalazion. Breast carcinoma metastases do occur in the eyelid and should be considered as a clinical differential diagnosis; however, the histological differential diagnosis does not include breast sweat gland carcinoma metastasis.

Clinicians should have a low threshold for submitting curettings for histopathology from any apparent chalazion which appears solid on incision. In particular, clinicians should be suspicious of solid recurrent chalazions, even if they have a benign clinical appearance which gives very little indication of the tumour's identity and aggressive behaviour. If in doubt, we recommend a large incisional biopsy for histopathological diagnosis; then wide excision biopsy with Mohs' micrographic surgery once the histopathology has been confirmed, to prevent recurrence.

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Optic disc dimensions, body length, and body weight

EDITOR,—In the optic disc, all visual afference originating in the photoreceptors of more than 1000 mm² retina is concentrated on an area of about 2 mm² to 3 mm². The retinal ganglion cell axons are nowhere else so densely and tightly packed as in the optic nerve head. It explains the importance of the optic disc for anomalies and diseases of the optic nerve. Regarding the marked interindividual variability of the size of the optic disc,¹ we undertook the present study to evaluate whether the dimensions of the optic disc are correlated with the length and weight of the whole body.

CASE REPORT

The study included 517 white subjects (243 women, 274 men) with a mean age of 46.6 (SD 13.0) years (range 8-87 years) and a mean refractive error of -0.91 (2.77) dioptres (range -24.0 to +7.0 dioptres). The subjects came to the eye hospital for diagnosis and treatment of glaucoma (n=244), or for diseases in the contralateral eye not included in the study (n=273). If both eyes had been examined, only one randomly selected eye per individual was considered for statistical analysis. For all subjects, colour stereo optic disc photographs had been taken. The diapositives were projected, the outlines of the optic disc were plotted on paper and morphometrically evaluated. The ocular and camera magnification was corrected according to the Littmann method taking into account the anterior corneal curvature and refractive error.^{2,3} Body length and body weight were additionally determined.

No statistically significant relation was found between area and diameters of the optic disc and length and weight of the whole body (p>0.50; Pearson's correlation coefficient R²<0.0005) (Fig 1). The same held true when

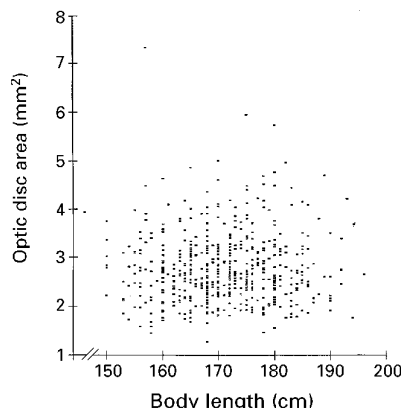


Figure 1 Scattergram showing the correlation between optic disc area and body length. $R = 0.0003$; $p = 0.69$.

only eyes with a myopic refractive error of less than 4 dioptres were taken into account, or when we considered only eyes with a myopic and hyperopic refractive error of less than 4 dioptres, or eyes with a myopic and hyperopic refractive error of less than 2 dioptres. Dividing the total study group into women and men, both sexes differed significantly ($p < 0.0001$; Mann-Whitney test) in body length and body weight. They did not vary significantly ($p = 0.45$) in optic disc area (2.74 (SD 0.72) mm² in women versus 2.80 (0.72) mm² in men).

COMMENT

The results suggest that, in white people, the size of the optic disc is independent of the dimensions of the whole body. Although the Littmann method may underestimate optic disc measurements in myopic eyes,⁴ one may arrive at this conclusion, since the correlations between disc area and body length and weight also remained statistically insignificant when only eyes with minor refractive errors were taken into account. The result of this study corresponds with the finding that women and men, although varying in body length and weight, did not differ in optic disc area. It agrees with other morphometric studies in which women and men did not vary significantly in retinal surface area and number of retinal photoreceptors and optic nerve fibres while optic disc area was significantly correlated with the count of optic nerve fibres and retinal photoreceptors and retinal surface area.⁵ The finding of the present study that the size of the optic disc is independent of the dimensions of the whole body may be important for optic nerve anomalies and diseases, such as optic disc drusen and non-arteritic anterior ischaemic optic neuropathy, the frequencies of which are correlated with optic disc size.⁵

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Orbital Wegener's granuloma resulting from direct extension of nasal disease through a surgical rhinostomy

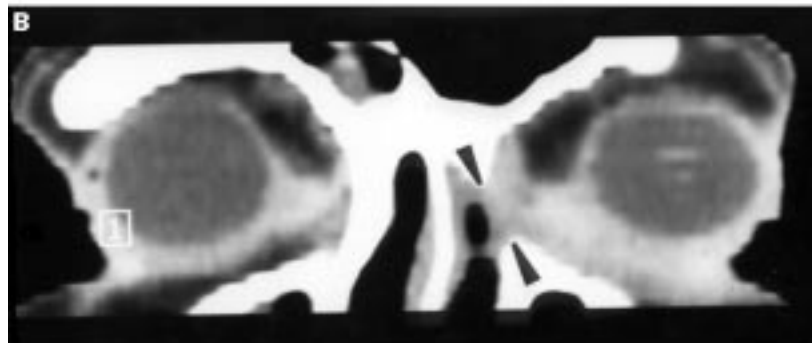
EDITOR,—Wegener's granulomatosis is a multi-system granulomatous disease of unknown aetiology but known to have numerous ocular complications including orbital inflammatory disease. We report a patient with "limited Wegener's granulomatosis" (Wegener's disease without renal involvement) who developed an orbital inflammatory mass by a direct extension of nasal inflammatory disease through a rhinostomy formed during lacrimal drainage surgery.

CASE REPORT

A 46 year old man, with a 7 year history of quiescent bilateral nasal sinus disease due to



Figure 1 Patient at 11 months after left dacryocystorhinostomy, with the left globe displaced anteriorly and superolaterally.



nasal mucosal biopsy proved Wegener's granulomatosis, was seen in the lacrimal clinic at Moorfields Eye Hospital. He was maintained on daily oral prednisolone 10 mg and azathioprine 150 mg. He had a 7 month history of bilateral epiphora and lacrimal irrigation demonstrated obstruction of the nasolacrimal ducts. Biopsy of the lacrimal sac mucosa at the time of left dacryocystorhinostomy was clear of granulomatous disease and he received no additional postoperative immunosuppression. Postoperatively all healed well and computed tomographic scan (CT scan) showed no orbital disease. At 11 months, while still on the original dose of immunosuppression, he presented with left retrobulbar ache. Visual acuity was unimpaired (Snellen 6/6 right eye, 6/9 left eye) and there was no relative afferent pupil defect. The left globe was displaced superolaterally by 2 mm and there was 3 mm of relative proptosis with no restriction of left ocular movements (Fig 1). An ill defined inferomedial orbital mass, of soft tissue attenuation and in continuity with the nasal cavity at the site of rhinostomy, was shown by CT scan (Fig 2). The proptosis and orbital signs resolved after a 6 month course of reducing dose of oral prednisolone 60 mg and cyclophosphamide 250 mg and the lacrimal drainage fistula remains patent. A right dacryocystorhinostomy was performed 3 months later, while the patient was on an increased maintenance dose of oral prednisolone 17.5 mg and cyclophosphamide 100 mg,

Figure 2 Axial (A) and coronal (B) CT scans at 11 months after surgery, showing a left inferomedial orbital mass extending into the orbit from the nose, through the site of rhinostomy (small arrows). The globe, medial rectus, and inferior rectus (large arrow, A) are displaced and surrounded by the mass (B, C).

and his postoperative course has been uneventful to 24 months of follow up.

COMMENT

Nasolacrimal duct obstruction has been reported in 7% of patients with Wegener's granulomatosis¹ and dacryocystorhinostomy in these patients may be associated with postoperative wound necrosis and the formation of nasocutaneous fistula.² Although dacryocystectomy has been suggested as a treatment for nasolacrimal duct obstruction in this disease,³ others have reported more encouraging results with dacryocystorhinostomy.^{4,5}

Extension of nasal Wegener's granulomatosis into the orbit after dacryocystorhinostomy has not previously been reported, although non-contiguous involvement, probably due to reactivated orbital disease, has been recorded at 1 month after a dacryocystorhinostomy in Wegener's granulomatosis.³ The disease is known to involve ocular and orbital tissues and several papers report the coexistence of orbital and nasosinus disease, with erosion of intervening bone.^{3,6,7} In the case reported here, orbital involvement was the direct result of the disease process passing through an iatrogenic bony opening created during dacryocystorhinostomy and, in view of this, we suggest that such patients should be monitored postoperatively for signs of orbital disease and be considered for additional perioperative systemic immunosuppression.

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Rhodococcus keratitis

EDITOR,—Ocular infection with *Rhodococcus* is rare.¹⁻⁴ In previous reports *Rhodococcus* species caused endophthalmitis^{1,3} and keratitis which failed to respond to treatment.⁴ We present a case of *Rhodococcus* keratitis which was successfully treated.

CASE REPORT

Eleven weeks after simple pterygium excision, an otherwise healthy 56 year old woman developed a corneal ulcer. No organisms were identified from corneal scrapes. Resolution had not occurred after 5 weeks' treatment with topical cefuroxime, gentamicin, and econazole. The patient was referred to Moorfields

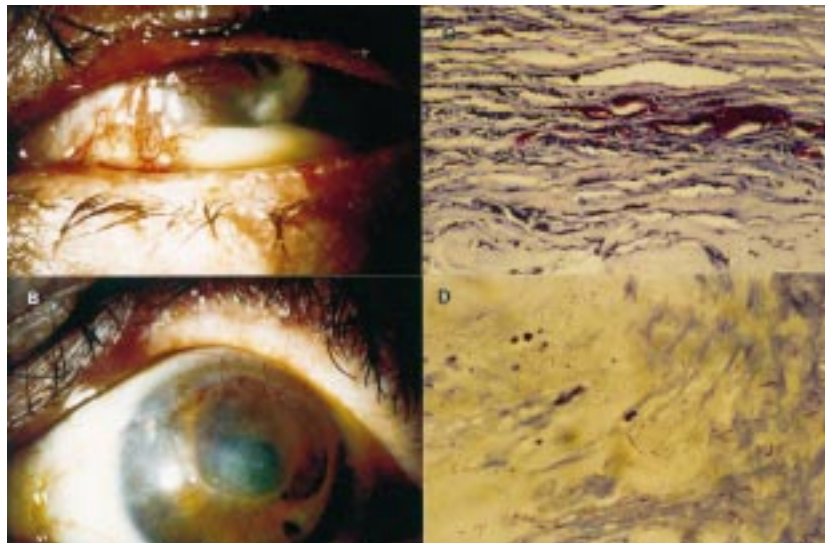


Figure 1 (A) The eye at the time of referral with the corneal infiltrate and associated hypopyon. (B) After 5 weeks of therapy with topical imipenem the epithelial defect had healed, the infiltrate resolved, and the neovascularisation had largely regressed. (C) and (D) The acid fast modified Ziehl-Neelsen staining response. Together with a weaker silver methenamine reaction this made a diagnosis of nocardial keratitis highly likely and *Rhodococcus* was confirmed serologically (C \times 235; D, \times 590).

Eye Hospital and treatment was stopped. On examination there was a central epithelial defect (1 \times 2 mm) associated with stromal thinning (60%), adjacent temporal multifocal infiltrates, nasal stromal neovascularisation, a 2 mm hypopyon, and scleritis (Fig 1A). After a further corneal scrape, treatment with topical amphotericin was commenced. Further deterioration occurred and a 6 mm diameter, 0.3 mm deep lamellar corneal biopsy was performed. The keratitis responded to this removal of infected tissue and treatment with hourly topical ofloxacin and oral ciprofloxacin. Histopathology suggested a nocardial infection (Fig 1C and D). The latter was confirmed by microbiological culture and the organism identified as a *Rhodococcus*. The organism was resistant to all 10 antibiotics tested except imipenem. Complete resolution of the keratitis occurred after 5 weeks' treatment with topical imipenem 0.5% (Fig 1B).

COMMENT

Rhodococcus species possess characteristics of both *Mycobacteria* and *Nocardia*, and exhibit pleomorphism growing as cocci, short rods, or branching filaments, although the *Rhodococcus* species have a less well developed mode of branching than the other species. The organism is ubiquitous and is a common cause of pneumonia in foals.⁴ Most reported *Rhodococcus* infections in humans have occurred in immunocompromised patients, including those with AIDS.⁵ The present case is of interest because of its rarity and because the patient was not immunocompromised. In addition, the infection responded to keratectomy and non-standard antibiotic therapy.

The authors are grateful to the Mycotic Reference Laboratory (Bristol) for typing the *Rhodococcus* organism.

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Anterior ischaemic optic neuropathy in a child with AS haemoglobinopathy and migraine

EDITOR.—Anterior ischaemic optic neuropathy (AION) is a disease that occurs most frequently in middle aged and elderly people resulting from vascular occlusive disorders or transient decreased blood flow in the territory of the short posterior ciliary arteries. Although rare in the young, it has been described in association with a number of conditions such as migraine, juvenile diabetes, collagen vascular diseases, haematological disorders, prothrombotic state, pre-eclampsia, hypertension, episodic hypotension, embolism, and even in the absence of other ocular or systemic condition.¹ We would like to report on a case of AION developing in a child with sickle cell trait and migraine. To our knowledge this association has not been reported so far as a cause of AION.

CASE REPORT

An 11 year old black boy was seen because of transient episodes of monocular loss of vision in both eyes. The patient had a long standing history of migraine usually triggered by fatigue, anxiety, hunger, and some foodstuffs. In most of the episodes he complained of severe throbbing hemicranial headache with no visual symptoms, but occasionally he noticed blurred vision in association with the pain. He used to take aspirin or paracetamol (acetaminophen) for pain relief. He also complained of recurring transient loss of vision in the left eye with no headache during intensive physical exercises such as playing soccer and doing gym at school. Four months earlier he had had an episode of headache in the morning followed by decreased sight in his left eye which persisted even after relief of the pain. He was seen at another hospital where a left optic disc oedema was observed and he was given prednisone 60 mg a day for 14 days with a partial recovery of vision.

Ophthalmic examination revealed his best corrected visual acuity to be 20/20 in the right and 20/60 in left eye. The right eye was unremarkable but the patient could read none of the Ishihara plates with the left eye, had a dense centrocaecal scotoma, and a pale optic disc in this eye, as well as a left relative afferent pupillary defect. There was no retinal abnormality.

Laboratory examination including complete blood cell count, blood chemistry, platelet count, prothrombin time, partial thromboplastin time, protein C, protein S, antithrombin III, and factor V activities were normal. Tests for anti-phospholipid antibodies and for collagen vascular diseases, echocardiogram, chest x rays, cranial magnetic resonance imaging, and cerebrospinal fluid analysis with immunoelectrophoresis were all negative. A visual evoked response study showed increased latency and reduced amplitude in the left eye. Haemoglobin electrophoresis revealed a double band of A and S mobility. The S haemoglobin was 38%, A haemoglobin 62%, and the red cells sickled under classic conditions. Careful systemic evaluation for possible complications of sickle cell trait was unrevealing.

COMMENT

Infarction in sickle cell disease is probably a very complex and multifactorial process. Its intricate mechanisms still remain to be elucidated but a number of disturbances such as intimal hyperplasia due to abnormal adhesive and procoagulant properties of sickled red blood cells, time to gelation, and platelet changes may play a role.^{2,3} Altered vascular reactivity and vasospasm may be also important.⁴ Although complications of sickle cell trait are rare they occur under extreme conditions such as vigorous exertion at high altitudes. Reported neurological complications of AS haemoglobinopathy include seizures¹ and spinal cord and cerebrovascular thrombosis.^{5,6} Migraine with resulting vasospasm may be a factor in triggering vascular occlusion in some of these patients.^{7,8} Complicated migraine may even be found in higher prevalence among people with sickle cell trait.⁷ Posterior ischaemic optic neuropathy has been reported in a patient with sickle cell anaemia and migraine,⁹ and in a case of SC haemoglobinopathy.¹⁰ Our patient presented loss of vision during a migraine attack. Extensive investigation did not suggest demyelinating disease or haematological disturbances

other than sickle cell trait. The present case is the first to be described with AION in the set of AS haemoglobinopathy and migraine. It is possible that sickle cell trait may increase the risk of AION in patients with migraine.

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Microcystic adnexal carcinoma masquerading as a chalazion

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