pancytopenia (white-cell count, 1690/mm³; red blood cell count, 2.69/mm³; platelet count, 24 000/mm³) and splenomegaly. Electrolytes were normal. Uric acid, lipid profile, renal function, and liver function were normal. Urine for Bence-Jones protein was negative. Bone marrow biopsy showed increased histiocytes. She underwent splenectomy in July 1999. During the ensuing years, the crystalline deposits increased progressively and became polychromatic. Serum immunoglobulin levels were checked in June 2001 and showed an elevation of IgA to 1400 mg/dl (normal 158–358 mg/dl). The IgG level was 911 mg/dl (normal 1188–1800 mg/dl). The IgM level was 39.5 mg/dl (normal 72–216 mg/dl). Skull and thoracic spinal X-rays revealed no osteolytic lesions. Urinalysis for Bence-Jones proteins was now positive. Serum immunoelectrophoresis revealed a monoclonal gammopathy involving IgA-kappa light chain. A repeat bone marrow biopsy demonstrated increased plasma cells to 15%. The diagnosis of IgA-kappa multiple myeloma was made in November 2001. Systemic therapy was initiated with melphalan and prednisolone. Serum IgA/IgG/IgM levels were 1210/818/25.4 mg/dl, respectively, 13 months after chemotherapy. Her best-corrected vision remained at 20/25 OU. The corneal crystalline deposits persisted. She rejected a biopsy of the cornea because her vision was not affected.

Discussion

Corneal crystalline deposits associated with multiple myeloma were first described by Burki in 1958.⁴ Since then, a variety of immunoprotein deposits have been described in the cornea, among which, IgG-kappa light-chain deposition is most frequently reported. Only one case of IgA-kappa chain monoclonal gammopathy associated with deposition has been reported previously.² That patient's best-corrected vision was reduced to counting fingers in the right eye and 20/80 in the left eye because of macular drusen in both eyes. The authors suggested a specific association between IgA monoclonal gammopathy and macular drusen, because drusen were not found in IgG monoclonal gammopathy. In our case, however, we did not observe macular drusen.

Presenting initially as crystalline keratopathy has also been reported to occur in patients of IgG benign monoclonal gammopathy.⁵ One study showed that 19% of 241 patients with a diagnosis of benign monoclonal gammopathy developed myeloma, or a related disorder when followed for 10 years or more.⁶ Morphological changes of the corneal crystals were noted previously in a patient of IgG-kappa monoclonal gammopathy.⁷ The deposits changed from dot-like opacities to polychromatic crystals, at which time multiple myeloma

was diagnosed. Similar alteration of the corneal crystals and malignant transformation of the underlying disease were observed in our patient. The corneal crystals in the previously reported case of IgA benign monoclonal gammopathy appeared colourless to slightly white,² which perhaps indicated an early stage of the disease process. Ophthalmologists should be aware of these manifestations and the potential for developing malignancies.

References

- 1 Krachmer JH, Mannis MJ, Holland EJ *Cornea*, Vol II. 977. Mosby-year Book, Inc. : 1997, St. Louis, Missouri.
- 2 Nik NA, Martin NF. Corneal crystalline deposits and drusenosis associated with IgA kappa chain monoclonal gammopathy. *Ann Ophthalmol* 1985; **17**(5): 303–307.
- 3 Budde M, Gusek-Schneider GC, Mayer U, Seitz B. Annular crystalline keratopathy in association with immunoglobulin therapy for pyoderma gangrenosum. *Cornea* 2003; **22**(1): 82–85.
- 4 Burki E. Über Hornhautveränderungen bei einem Fall von multiple Myelom (Plasmocytoma). *Ophthalmologica* 1958; **135**: 565–572.
- 5 Hutchinson K, Dal Pra M, Apel A. Immunoglobulin G crystalline keratopathy associated with monoclonal gammopathy. *Aust N Z J Ophthalmol* 1998; **26**(2): 177–179.
- 6 Kyle RA. 'Benign' monoclonal gammopathy: a misnomer? *JAMA* 1984; **13**(251): 1849–1854.
- 7 Perry HD, Donnenfeld ED, Font RL. Intraepithelial corneal immunoglobulin crystals in IgG kappa multiple myeloma. *Cornea* 1993; **12**(5): 448–450.

C-M Hsueh, P-Y Lin, S-M Lee, W-M Hsu

Department of Ophthalmology
Taipei Veterans General Hospital
National Yang Ming University
No. 201, Sec 2
Shih-Pai Road Taipei 11217
Taiwan

Correspondence: P-Y Lin
Tel: +886-2-28757325
Fax: +886-2-28761351
E-mail: pylin@vghtpe.gov.tw

Sir,

The varix of angular vein

Eye (2004) **18**, 645–647. doi: 10.1038/sj.eye.6700679

These cases describe an unusual cause for a paranasal swelling, the lesion mimicking a lacrimal sac mucocoele,

and illustrate the importance of thorough examination in an apparently routine case.

Case report

Case one

A 59-year-old Caucasian woman was referred with a diagnosis of possible left lacrimal sac mucocoele. She gave a 6-month history of subcutaneous paranasal swelling overlying the left medial canthal tendon, but with no associated epiphora, discharge, pain, or headache. There had been no change in the mass and she was otherwise asymptomatic with no previous ocular history or past medical history.

Her Snellen visual acuities were 6/6 in each eye, with no intraocular disease. There was an uninflamed mass, measuring 4 mm × 5 mm, overlying the left medial canthal tendon and although resembling a lacrimal mucocoele, it could be emptied by firm pressure and readily refilled (Figure 1); it did not increase in size with a Valsalva manoeuvre, although refilling was faster. There was no evidence of an orbital mass or restriction of eye movements. The dye disappearance test was normal and both canaliculi were freely patent to syringing. A clinical diagnosis of angular vein varix was made, with a recommendation of no active intervention.

Case two

A 65-year-old Caucasian woman presented with a right paranasal lump, which had slowly increased in size over 4 years. Although she had childhood pulmonary tuberculosis, there was no previous ocular history.

The paranasal lump was nontender, purple, mobile in the subcutaneous space and found to lie in the line of the angular vein. There was a nearby scar where a previous removal had been attempted. As the lesion was not compressible, a diagnosis of thrombosed angular vein was made. It is possible that previous surgery had caused retrograde thrombosis of the angular vein up to the union of the frontal and superior ophthalmic veins. CT scan confirmed there was no associated intranasal or intraorbital anomaly and the patient elected for no treatment.

Case three

A 61-year-old Sri Lankan woman presented with a left paranasal lump that was initially painful. It had been present for several months. She had a history of blepharitis and keratoconjunctivitis sicca secondary to glutaraldehyde exposure. She had no history of trauma to the area. Her history was otherwise unremarkable. The paranasal lump was nontender, mobile in the subcutaneous space and found to lie in the line of the angular vein (Figure 2). The lesion was not compressible and a diagnosis of spontaneous thrombosis of an angular

Figure 1 The paranasal mass (a) is easily compressible with a finger (b) and starts to refill rapidly even before capillary return is complete (c); this confirms the diagnosis of an angular vein varix, rather than a lacrimal sac mucocoele.

vein varix was made without further investigation. The patient elected for no treatment.

Comment

A lacrimal sac mucocoele typically presents as a paranasal swelling centred just below the medial canthal

Figure 2 Left thrombosed angular vein varix (arrow).

tendon and the patient will usually complain of epiphora. Pressure will deflate the sac if open to the canaliculi or nasolacrimal duct, but otherwise the sac may become distended with mucus and assume a blue appearance. Dacryocystorhinostomy is to be recommended to prevent dacryocystitis and relieve epiphora.

A varix of the angular vein may simulate a lacrimal sac mucocoele, but tends to be anterior to the medial canthal tendon. The angular vein is formed by the junction of the supra-trochlear and supra-orbital veins that runs obliquely downwards on to the side of the root of the nose, to the level of the lower margin of the orbit where it becomes the anterior facial vein. It communicates with the cavernous sinus by draining into the superior ophthalmic vein.

The aetiology of the condition is unclear. Varices usually occur at other sites because of chronic obstruction to flow,¹ but could also be due to abnormalities of connective tissue,² following trauma³ or due to hereditary predispositions.⁴ There was no clear aetiology in any of our cases.

There has been one prior English language report of angular vein varix,⁵ where the lesion was excised for cosmetic reasons. The diagnosis is a clinical one, but could be confirmed by phlebography or Doppler ultrasonography. None of our cases justified surgical excision with the small, perhaps entirely theoretical, risk of intracranial air embolus via the superior ophthalmic vein. Sclerotherapy and ligation have been used with some success for oesophageal varices,⁶ but we have no experience of their use in this situation.

References

- 1 Sarin SK, Agarwal SR. Extrahepatic portal vein obstruction. *Semin Liver Dis* 2002; **22**: 43–58.
- 2 Chun JY, Dillon WP, Berger MS. Symptomatic enlarged cervical anterior epidural venous plexus in a patient with Marfan syndrome. *Am J Neuroradiol* 2002; **23**: 622–624.
- 3 Cesarone MR, Belcaro G, Nicolaides AN, Geroulakos G, Griffin M, Incandela L *et al*. 'Real' epidemiology of varicose veins and chronic venous diseases: the San Valentino Vascular Screening Project. *Angiology* 2002; **53**: 119–130.
- 4 Laurikka JO, Sisto T, Tarkka MR, Auvinen O, Hakama M. Risk indicators for varicose veins in forty- to sixty-year-olds in the Tampere varicose vein study. *World J Surg* 2002; **26**: 648–651.
- 5 Nasr AM, Huaman AM. Anterior orbital varix presenting as a lacrimal sac mucocoele. *Ophthalmic Plast Reconstr Surg* 1998; **14**: 193–197.
- 6 Bosch J, Garcia-Pagan JC. Prevention of variceal rebleeding. *Lancet* 2003; **361**: 952–954.

SR Khan, BJL Burton, M Beaconsfield and GE Rose

Moorfields Eye Hospital, London EC1V 2PD, UK

Correspondence: GE Rose

Tel: +44 20 7253 3411 (x2034)

Fax: +44 20 7566 2019

E-mail: Geoff.rose@moorfields.nhs.uk

Sir,

Onset of Charles Bonnet syndrome (formed visual hallucinations) following bilateral laser peripheral iridotomies

Eye (2004) **18**, 647–649. doi:10.1038/sj.eye.6700719

Formed visual hallucinations in patients with normal cognition and insight (Charles Bonnet Syndrome (CBS)) have been reported after certain ophthalmic procedures, including macular laser photocoagulation¹ and macular translocation.² It is important for clinicians to recognise this condition as it is frequently misdiagnosed³ and the symptoms may bother some patients.⁴ We report a patient who developed formed visual hallucinations following bilateral laser peripheral iridotomies for angle-closure glaucoma.

Case report

A 90-year-old Chinese woman presented with blurring of vision secondary to bilateral nuclear sclerotic cataracts and chronic angle-closure glaucoma. Her best-corrected