myocardial injury include increased sympathetic tone causing coronary arterial spasms, altered β-adrenergic regulation with enhanced responsiveness to exogenous catecholamine, and direct myocardial injury caused by oxygen-derived free radicals. 5

A critical incident analysis of the event revealed that a drug concentration error had occurred that resulted in the injection of a local anaesthetic mixture containing bupivacaine 0.5% and epinephrine 1:2000 in the block instead of the intended bupivacaine 0.5% with epinephrine 1:2000.

In conclusion, we present a case of severe left ventricular dysfunction caused by an epinephrine overdose during a retrobulbar block. Our experience raises the case for withdrawal from the operating room of high concentration epinephrine (1:1000), which was added to the local anaesthetic used for the block and also questions the necessity of administration of epinephrine with ophthalmic local anaesthetic blocks.

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Clinical and in vivo confocal microscopic findings of a patient with ocular ochronosis

Alkaptonuria is a rare autosomal recessive metabolic disorder of phenylalanine/tyrosine metabolism due to a defect in the enzyme homogentisate 1,2-dioxygenase (HGA) to maleylacetoacetate in the catabolism of phenylalanine and tyrosine. 1 This metabolic defect results in the accumulation of oxidized HGA polymers in collagenous structures and various tissues. 2 High HGA levels cause massive excretion of HGA in the urine and as it is oxidized, the urine turns dark, thus establishing the pathognomonic feature of alkaptonuria. 3 Pigment deposition eventually results in blue-black pigmentation of cartilage and skin, spondylarthropathy, large-joint arthropathy, and cardiac valvular disease. 2,4 The most important complications of alkaptonuric ochronosis are related to deposition of ochronotic pigment in target organs such as cartilage, tendon, conjunctiva, cornea, and sclera.

A 25-year-old male diagnosed with ochronosis was referred from the endocrinology department for further clinical and laboratory evaluation. His family history was negative and he was not taking any medication. Systemic examination revealed a reduction in movement in both knees and generalized bluish pigmentation on the skin. Discolouration of the ears and an irregular thickening of auricular cartilage were not observed. Cardiovascular examination revealed mild diastolic bruit at the base of the heart. The urinalysis showed calcium oxalate crystals. The urinary HGA concentration was markedly elevated and showed a level greater than 100 mmol HGA per millimole of creatinine by gas chromatography-mass spectrometry analysis. Transesophageal echocardiography disclosed tricuspid aortic valve. Radiologic evaluation revealed vacuum phenomenon at multiple levels of inter-vertebral joints indicative of nitrogen deposition, which is a marker for lumbar degenerative disc disease and concomitant joint space narrowing. On ophthalmologic examination, his uncorrected visual acuity was 20/20 OD and 1183 μm OS. Slit-lamp examination demonstrated bilateral brown pigmentation in the interpalpebral bulbar conjunctiva (Fig. 1A) and diffuse annular brownish gold corneal deposits (Fig. 1B). Fundus examination was within normal limits. Corneal pachymetry disclosed central corneal thickness of 526 μm OD and 508 μm OS. Peripheral corneal thickness measurements revealed 1187 μm OD and 1183 μm OS.

In vivo confocal microscopy (IVCM; Confoscan 3.0, Vigonza, Italy) demonstrated an accumulation of hyper-
reflective crystalline deposits at the level of the Descemet membrane in the form of a homogenous acellular band (Fig. 2A). More superficially, there were scattered hyper-reflective microdeposits in the appearance of arborizing lines in between the epithelium and anterior stromal layer (Fig. 2B). No sign of inflammation was observed at both locations. The other corneal layers appeared to be uninvolved. Throughout the stroma, keratocytes appeared normal in morphology, and no intracellular deposits were noted.

In ocular ochronosis, the pigment is localized to the sclera, conjunctiva, and limbus. Often, pigmentation of the ocular surface is the initial manifestation of the disease. Deposits associated with ocular ochronosis may involve the conjunctiva, and thus should be considered in the differential diagnosis of pigmented lesions of the conjunctiva, among which extraocular extension of uveal melanoma is also included. Extraocular extension of uveal melanoma typically causes single or multiple homogeneously dark nodular episcleral masses underneath the conjunctiva. In contrast, pigmentation of the ocular surface creates flat lesions, may be limited only to the conjunctiva, and may not be sharply circumscribed or homogenous in colour.

Bilateral pigment accumulation in the conjunctiva and sclera is more frequently reported than corneal involvement in the literature. Carlson et al. reported an accumulation of amber-coloured pigmentation at the level of Bowman layer, a finding also supported by our observations. Nasally and temporally located perilimbal ochronotic scleral lesions
were found to be associated with progressive peripheral corneal thinning and astigmatism in the axis of the lesions.\textsuperscript{9} Ring pigmentation in the peripheral cornea may also arise secondary to Wilson disease, non-Wilsonian chronic liver disease, hypercarotenemia (hypervitaminosis A), or ocular chrysiasis.\textsuperscript{6–12} Typically, hypercarotenemia presents with circumferential involvement of the peripheral deep stroma with yellow-brown pigment.\textsuperscript{11} Corneal involvement in Wilson disease is characterized by bilateral peripheral golden-brown, blue-green, or ruby red homogeneous deposits at the level of Descemet membrane.\textsuperscript{10} Impaired copper metabolism may also lead to similar deposits in non-Wilson hepatic disease.\textsuperscript{10} In contrast, diffusely scattered dotlike deposits in peripheral and central stroma is observed in ocular chrysiasis.\textsuperscript{13} By providing microscopic details of corneal deposition, IVCM may be helpful in the differential diagnosis of such disorders.

Our case describes a patient with bilateral conjunctival pigmentation and annular corneal pigmentation in addition to arthropathic changes and skin pigmentation. Using IVCM, we were able to demonstrate the extent, level, and nature of corneal involvement in ochronosis. IVCM revealed highly reflective, homogeneous deposits most dense at the level of Descemet membrane and scattered microdeposits at Bowman layer with sparing of remaining corneal layers, including epithelium, stroma, and endothelium.

To the best of our knowledge, our report is the first one in the literature to detail the in vivo ultrastructural corneal characteristics of ochronosis using IVCM. Because ocular involvement may be the first sign of alkaptonuric ochronosis, it is important to make the differentiation from other pigmented lesions of the eye. In these settings, IVCM is a valuable diagnostic tool that may assist in detection and exact localization of pathologic deposits in the cornea and help to establish the correct diagnosis in such patients.

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REFERENCES


\textbf{Intraosseous orbital hydatid cyst: report of a rare case}

A 69-year-old female came to Oculoplastic Clinic, Farabi Eye Hospital, Tehran, Iran, with the complaint of new-onset proptosis and loss of vision in her left eye from 4 months ago. She had no history of any trauma.

On examination, there was no visible skin scar of any previous surgery around the orbit. The right eye had best corrected visual acuity (BCVA) of 9/10, and slit-lamp examination of its anterior segment and funduscopy showed no pathology except mild cataract. In her left eye, she had BCVA of 2/10, mild conjunctival injection and chemosis, severe proptosis (10-mm difference in exophthalmometry; Fig. 1A), lateral gaze limitation, and a reactive pupil with 2+ relative afferent pupillary defect.

The left eye had also mild cataract, and its funduscopy showed no significant abnormal finding.

Radiologic evaluation for the eye was started with CT scan (Fig. 1B). CT scan revealed a well-defined, dumbbell-shaped cystic lesion—internal density similar to water, delineated by a perceptible hyperdense rim—in lateral wall of orbit, with destruction of the bone and extension into the orbit, compressing orbital contents and pushing the globe forward.

For further evaluation of the lesion, magnetic resonance imaging (MRI) was performed (Fig. 1C, 1F). MRI showed a multiloculated cystic lesion with better depicting peripheral fibrous capsule and internal septations. The lesion had low signal intensity in T1-weighted imaging without enhancement and high signal intensity in T2-weighted images.