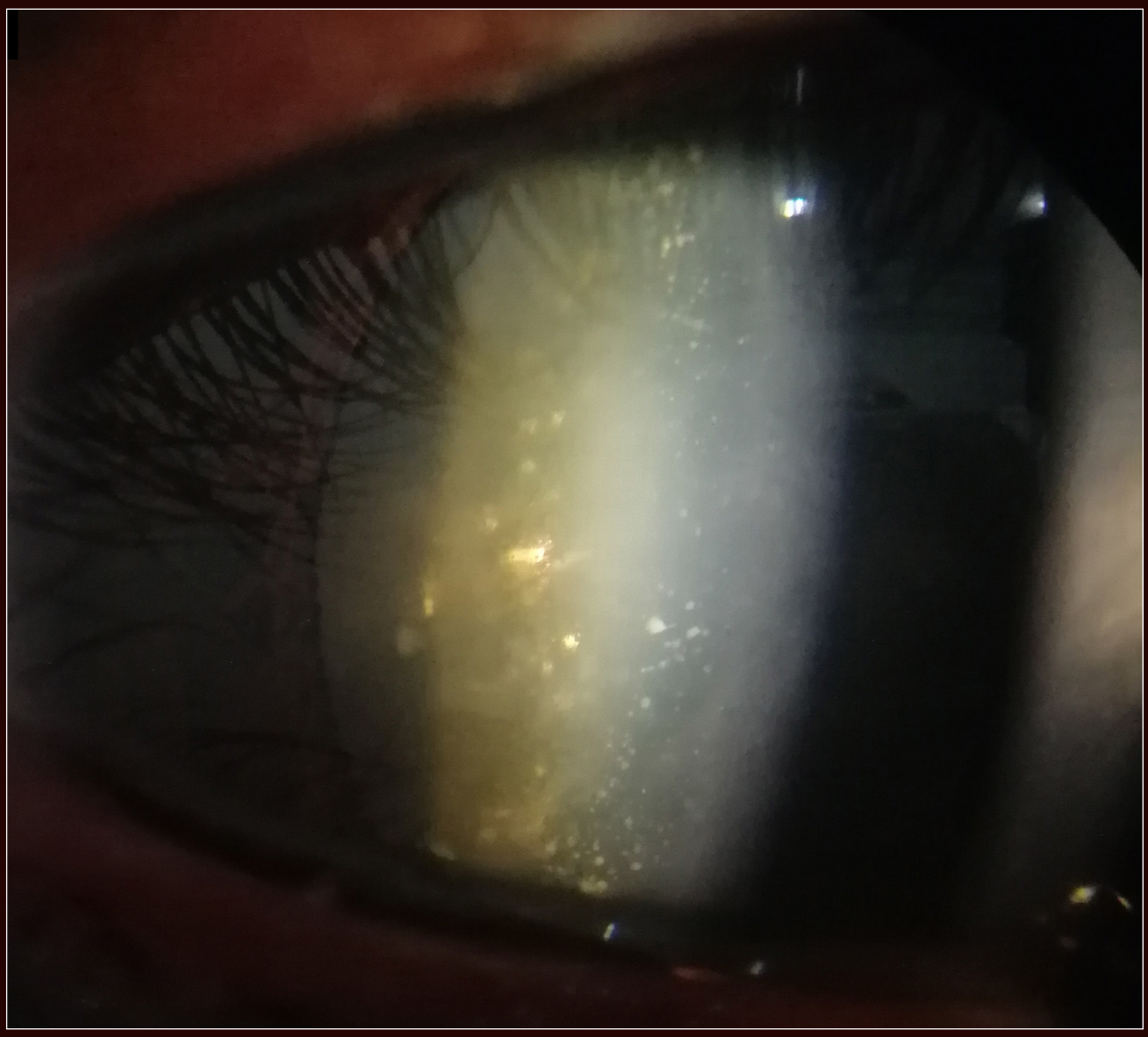
ISSN 1737-1104

Journal Tunisien d'Ophtalmologie

Organe d'expression de la Société Tunisienne d'Ophtalmologie



Novembre 2022 - Vol. 31

N°3

FAITS CLINIQUES

Christmas Tree Cataract: a shiny cataract

Cataracte en « Sapin de Noël » : une cataracte qui brille.

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Introduction

Christmas tree cataract (CTC) is a rare type of lenticular opacity. The terminology emerged from clinical picture. Indeed, Brightness and variety of colors refer to the ornaments that commonly decorate the Christmas tree [1]. It is characterized by polychromatic needle-shaped deposits in the deep cortex and nucleus [2]. CTC can be associated to cataractous changes. Unilateral presentation is common, though asymmetrical bilateral form is not rare [3]. There is an established association between CTC with myotonic dystrophy (MD). Most studies claim that CTC could represent an early manifestation of the disease [4].

Case report

A 55-year-old man complained of progressive bilateral blurred vision with no relevant medical history. His best-corrected visual acuity was limited to 20/200 in the right eye (RE) and 20/100 in the left eye (LE) (Snellen scale). Ophthalmologic exam was quite difficult since the patient was photophobic.

Slit-lamp examination after pupillary dilatation revealed polychromatic and highly reflective, needle-shaped opacities in both lenses. A magnified view using direct illumination revealed a spectacular array of polychromatic needle shaped crystals traversing all directions and cutting across the natural limits of the lens fibers, located in the deep cortex and anterior nucleus of the lens, giving the appearance of colored lights decorating the branches as on a Christmas tree. The colors of the crystals varied according to the angle of the incident light (**Figure 1 A, B**). Otherwise, ocular exam was unremarkable.
Furthermore, we performed an Ultrasound Biomicroscopy (UBM) which showed multiple vacuolar dark spots in the cortex and hyperreflective core within the lens matter presenting with back shadows in the RE (**Figure 2**).
The patient didn't have any history of systemic nor ocular illness and no prior trauma nor surgery. He didn't complain of any muscle weakness and had no history of cardiac illness. Following a physician consult, routine blood investigations (serum cholesterol, lipids and calcium), electrocardiogram (ECG) and electromyogram were found to be within normal limits. No positive family history was elicited. The diagnosis of idiopathic bilateral Christmas

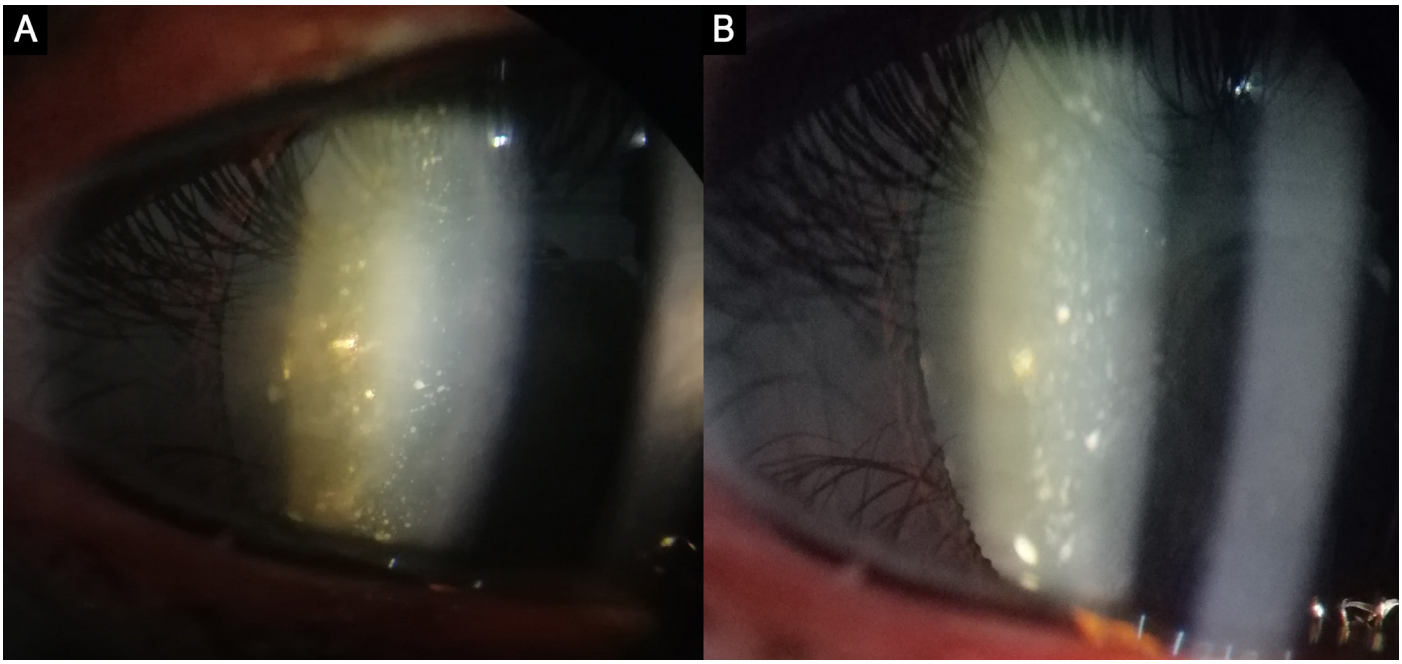

Figure 1. Lens photography showing a spectacular array of polychromatic needle shaped crystals traversing all directions and cutting across the natural limits of the lens fibers, located in the deep cortex and anterior nucleus of the lens, giving the appearance of colored lights decorating the branches as on a Christmas tree. (A: right eye; B: left eye).

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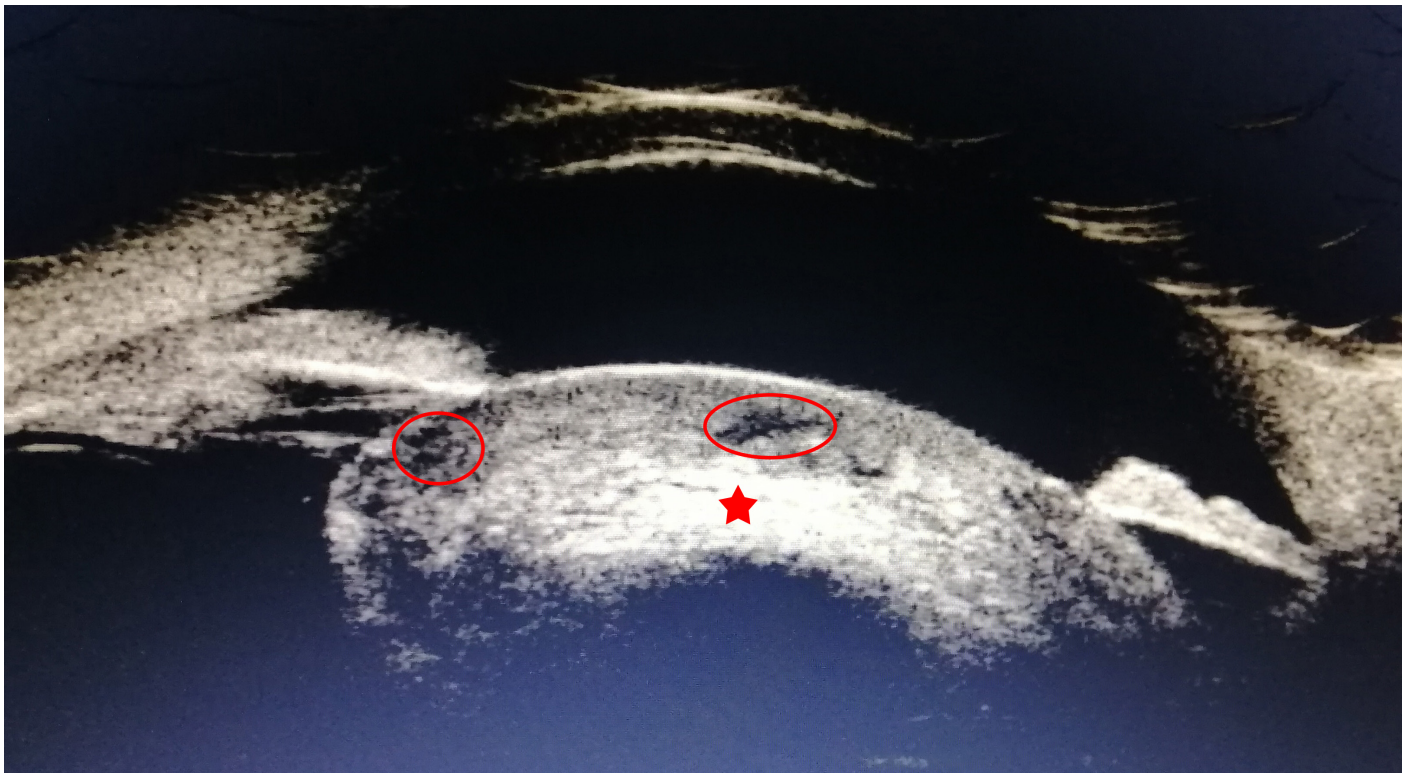

Figure 2. The Ultrasound Biomicroscopy (UBM) of the right eye showing multiple vacuolar dark spots in the cortex (red circles) and hyperreflective core within the lens matter (red star) presenting with back shadows.

Tree Cataract was established. The patient had cataract surgery by phacoemulsification and foldable IOL implantation in the RE. Postoperatively, best corrected visual acuity improved to 20/20.

Discussion

Christmas tree cataract (CTC), also known as starry cataract, is a rare form of polychromatic lens opacity. It is characteristically described as refractile, needle-shaped, multicolored crystalline structures found in the cortex of the lens [4]. Its colors usually range from red to green depending on the incident beam and resemble the lights on a Christmas tree.

There are still many unresolved controversies regarding the optical properties of the highly refringent multi-colored needles [4]. There have been very few published articles describing this entity since 1982. Pau and Forster were the first ones to suggest that the opacities seen in CTC were cholesterol crystals [5]. Hayes and Fisher subsequently postulated the diffraction of light from parallel sided stacks of fused cell membranes to be the cause for the striking polychromatic luster [1]. As per Anders and Wollensak, the crystals were cholesterol in nature and were the result of lens metabolism [6]. According to Shun-Shin et al., CTC was most likely made of cystine due to an age-related aberrant change in crystalline fibers induced by elevated blood calcium levels [7]. Similarly, Obi and Weir postulated that CTC resulted from excess cystine concentration leading to growing crystals [8]. Reiter and Gramer found MD in 16.7% of patients with CTC [4]. Few other authors have also reported CTC but did not postulate any cause for its development [9-25]. Table I shows all CTC case reports so far from different parts of the world.

Recently, Yangzes et al [19] and Ramesh et al. [25] used the Pentacam Scheimpflug images and Anterior Segment optical coherence tomography (AS-OCT) to better explore CTC. Pentacam Scheimpflug imaging revealed hyper-reflective shadows in the cortical and nuclear lens matter. Cataractous material were seen as dark spots with few crystals presenting with back shadows. Besides, the highly refractive multi-colored needle material on slit-lamp examination was visualized as hyperreflective spots within the lens matter on AS-OCT

CTC is managed by phacoemulsification with foldable IOL implantation. The surgical procedure is standard and does not require specific measures. However, some authors reported an increased incidence of posterior capsular opacity and postoperative anterior capsule contraction [12]. Since the crystals are highly refringent and change considerably the viewing of the surgeon during the procedure, an slightly increased time of surgery may be expected [12].

To conclude, CTC is a rare cataract that may be found incidentally since it can be visually non-disturbing. Its spectacular display of brilliance is very striking. Therefore, a systemic evaluation, especially a neurological examination, must be performed in order to exclude serious diseases as MD. New imaging techniques as AS-OCT, UBM and Pentacam Scheimpflug can help better understanding this rare entity.

Competing interests

The authors have no commercial associations or sources of support that might pose a conflict of interest.

Funding

The authors declare that this study is not funded by any foundation or organisation.

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