Case study

Cataract in hypoparathyroidism: a case study.

ABSTRACT:
Ophthalmologic involvement in hypoparathyroidism can affect each of the components of the eye. However, it has a particular tropism for the lens, the prevalence of cataract reaches approximately 50% of hypoparathyroid subjects. Cataracts are sometimes indicative of these affections. Its detection is a constant concern in the monitoring of patients, because it constitutes a severity index of the disease, and is likely to benefit from a specific management.
In this case, we want to raise awareness of this etiology of cataracts, rarely encountered in ophthalmology, describe its pathogenesis and report the other ophthalmological manifestations linked to hypoparathyroidism which reflect a certain element of severity in order to detect and prevent them.

Keywords : Cataract, hypoparathyroidism, hypocalcemia.

1. INTRODUCTION :

Insufficient production or action of parathyroid hormone determines a situation of chronic calcipenia which is clinically expressed in the form of acute or chronic signs of neuromuscular hyperexcitability or tetany.
The ophthalmologic involvement of hypoparathyroidism is likely to alter each of the constituent parts of the eye. However, it has a particular tropism for the lens, the prevalence of cataract reaches approximately 50% of hypoparathyroid subjects **(1).** Cataracts are sometimes indicative of these affections. Its detection is a constant concern in the monitoring of patients, because it constitutes a severity index of the disease, and is likely to benefit from a specific management. Great questions persist as to its pathogenesis. Big questions persist as to its pathogenesis.

2. CASE REPORT:

We report the case of a 37-year-old patient followed for a familial hypercalciuric hypocalcemia as well as for a renal lithiasis and who consults for a progressive decrease in visual acuity in his right eye.
On physical examination, the patient had stable vital signs. The ophthalmological examination revealed a visual acuity of 6/10 in his right eye increasing to 10/10 after correction, and a visual acuity of 10/10 in the left eye.
The Intraocular pressure is normal in both eyes and the oculomotor examination is without abnormality.
The slit lamp examination is normal in the left eye and revealed in the right eye:
In diffused lighting: a clear cornea, a normal anterior chamber and a dusty cataract (**Figure 1**). In section: the opacities are radiary in an arc of a circle and sit at the level of the epinuclei (**Figure 2**). The dilated fundus is normal in both eyes.

Considering his good corrected visual acuity, the patient is adapted in corrective glasses. Phacoemulsification surgery with placement of an intraocular implant will be considered depending on the evolution of the lens, vision and functional impairment.

3. DISCUSSION:

Hypoparathyroidism is a deficiency of parathyroid hormone often caused by autoimmune disease, genetics, iatrogenic damage or removal of glands during thyroidectomy or parathyroidectomy, but the cause is often still imperfectly determined **(1).** Measurement of parathyroid hormone (PTH) levels is necessary for diagnosis. Treatment includes calcium and vitamin D supplementation.

Familial hypercalciuric hypocalcemia or Autosomal dominant hypocalcemia (ADH) is most often due to activating mutations of the CaSR (calcium-sensing receptor) gene encoding the calcium-sensitive receptor leading to hypocalcaemia associated with an unsuitable parathyroid hormone level (low or normal), as during hypoparathyroidism, but also an abnormally high calciuria due to hypocalcemia.

The most common ophthalmologic consequence of hypoparathyroidism is cataracts. although this mechanism remains obscure **(2).** Cataract appears to be authentically linked to chronic calcipenia, but also undoubtedly involves, as in diabetes mellitus, disorders of the hydration of the crystalline lenses.
Experimentally as early as 1926 had been obtained in rats subjected to phases of hypo- and normocalcemia, concentric layers of opacities and clarities **(3).** Lens opacities have also been obtained in rats on a calcium and hyperphosphorus diet **(4).**

Cataract lesions therefore appear frequent in adults with genetic hypoparathyroidism (like our case) that were recognized late. Conversely, compared to 2064 controlled subjects, the risk of cataract and its age of onset did not differ significantly in the 688 subjects who developed post-surgical hypoparathyroidism and subjected to vitamin-calcium treatment **(5),** even though traditionally the prevalence of cataract affects approximately 50% of hypoparathyroid patients **(1).**
This endocrine cataract can be unilateral or bilateral and also nuclear and / or cortical and / or subcapsular **(6).** The evolution of cataracts is typically slow in cases of idiopathic hypoparathyroidism like our case and more rapid in cases of acute hypoparathyroidism **(7).**
The severity of cataracts appears to be related to both the severity and the duration of the hypocalcemia with hyperphosphatemia **(8).** Cataracts have been shown to occur in 28% of patients with hypocalcaemia for more than 4 years **(9).**

The discovery of cataract lesions in a subject with hypoparathyroidism is an indication of severity of the disease. It imposes:

- ✓ The search for other trophic disorders: evaluation of the dentition, dental enamel, search for calcifications or ossifications of the soft parts, detection of nephrocalcinosis and nephrolithiasis, calcifications of the central gray nuclei or more diffuse of the brain (Fahr syndrome), ideally by abdominal and brain computed tomography (CT) scans without injection.

- ✓ The intensification of therapeutic management to obtain a metabolic balance as perfect as possible, normalizing calcium levels, phosphatemia, phosphocalcic blood product, calciuria, at the cost of vitamin-calcium therapies, sometimes thiazides, or even recombinant PTH.

Other ocular manifestations related to hypoparathyroidism have been described in the literature:

- ✓ Papillary edema: Much less often reported, papillary edema appears mainly in children and is observed in both hypo- and pseudo-hypoparathyroidism **(7- 9).** Bilateral, often revealing, it is actually reversible with correction of hypocalcemia. Papillary edema is associated with alterations in axoplasmic conduction associated with hypocalcemia and stasis **(10 - 11).**
- ✓ Weiss's sign and spasm of the oculomotor muscles: Neuromuscular hyperexcitability linked to hypocalcemia is traditionally demonstrated by Weiss's sign: the percussion of the external angle of the orbit determines contracture of the orbicularis and of fact a zipper of the eyelids. In a 13-year-old adolescent girl with pseudo-hypoparathyroidism, irregular spasmodic eye movements, predominantly horizontally, were also reported during cataract surgery. The signs were precipitated by a state of respiratory alkalosis associated with hypocalcemia, and corrected with continuous infusion of calcium gluconate **(12).**
- ✓ Palpebral, orbital and sclera calcifications or ossifications: They have also been reported, forming part of the calcium deposits or ossifications of the soft tissues, characteristic of these situations **(13).**
- ✓ Keratoconjunctivitis: An exemplary inventory of the ocular signs of APECED syndrome or Autoimmune Polyendocrinopathy Candidiasis Ectodermal Dystrophia (a condition, transmitted in an autosomal recessive manner which is typically characterized by Whitaker's triad: hypoparathyroidism, adrenal insufficiency, recurrent mucocutaneous candidiasis), evolution and prognosis was carried out in Helsinki, reported in 2000. In a series of 69 patients, keratoconjunctivitis was present in 17 subjects (25% of cases) **(14).**
- ✓ More rarely: sicca syndrome, iridocyclitis, severe retinal sufferings and optic atrophies **(15).**

4. CONCLUSION:

By this case, we want to raise awareness about this etiology of cataracts, rarely seen in ophthalmology.

Diversity of expressions and mechanisms characterize the ophthalmologic complications associated with hypoparathyroidism. These manifestations reflect a certain element of severity. It is important to detect and prevent them.

FIGURES:

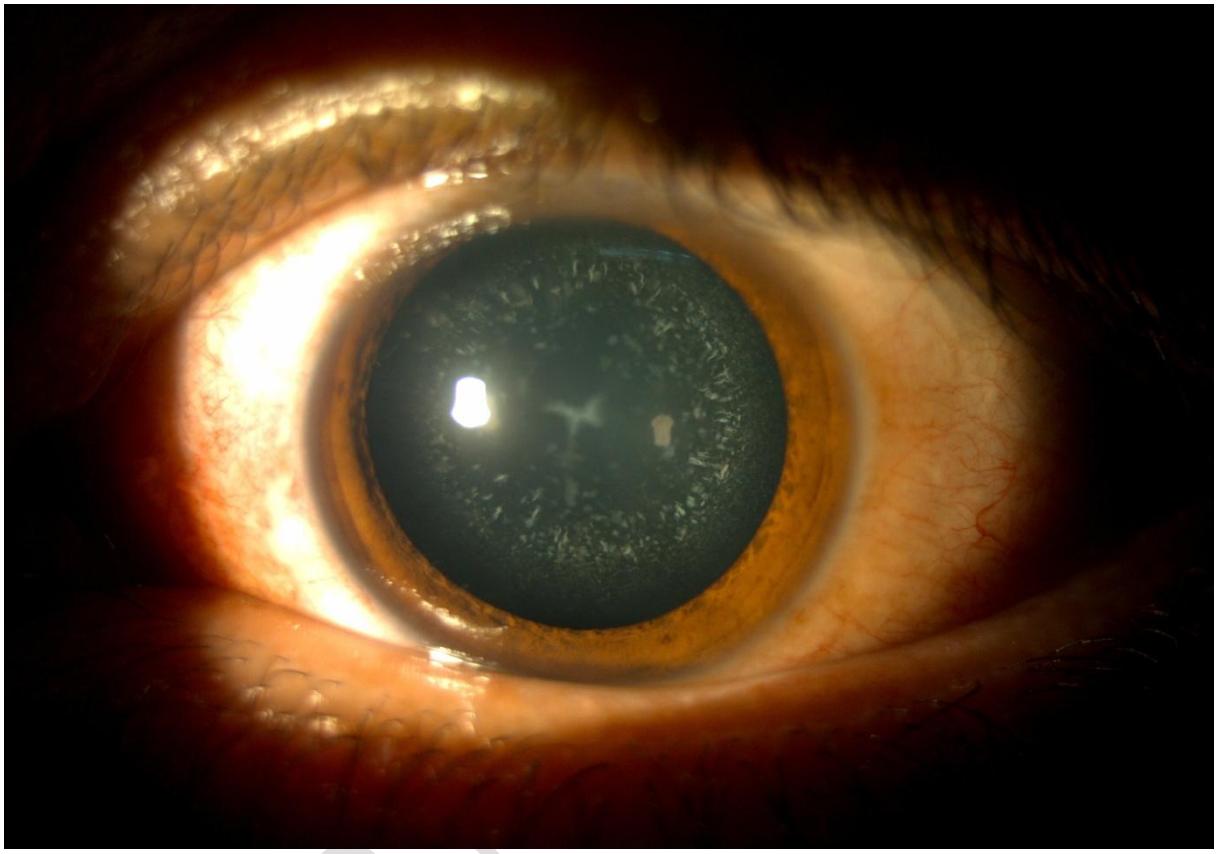

Figure 1: Slit lamp examination of the right eye showing a dusty cataract in diffused lighting.

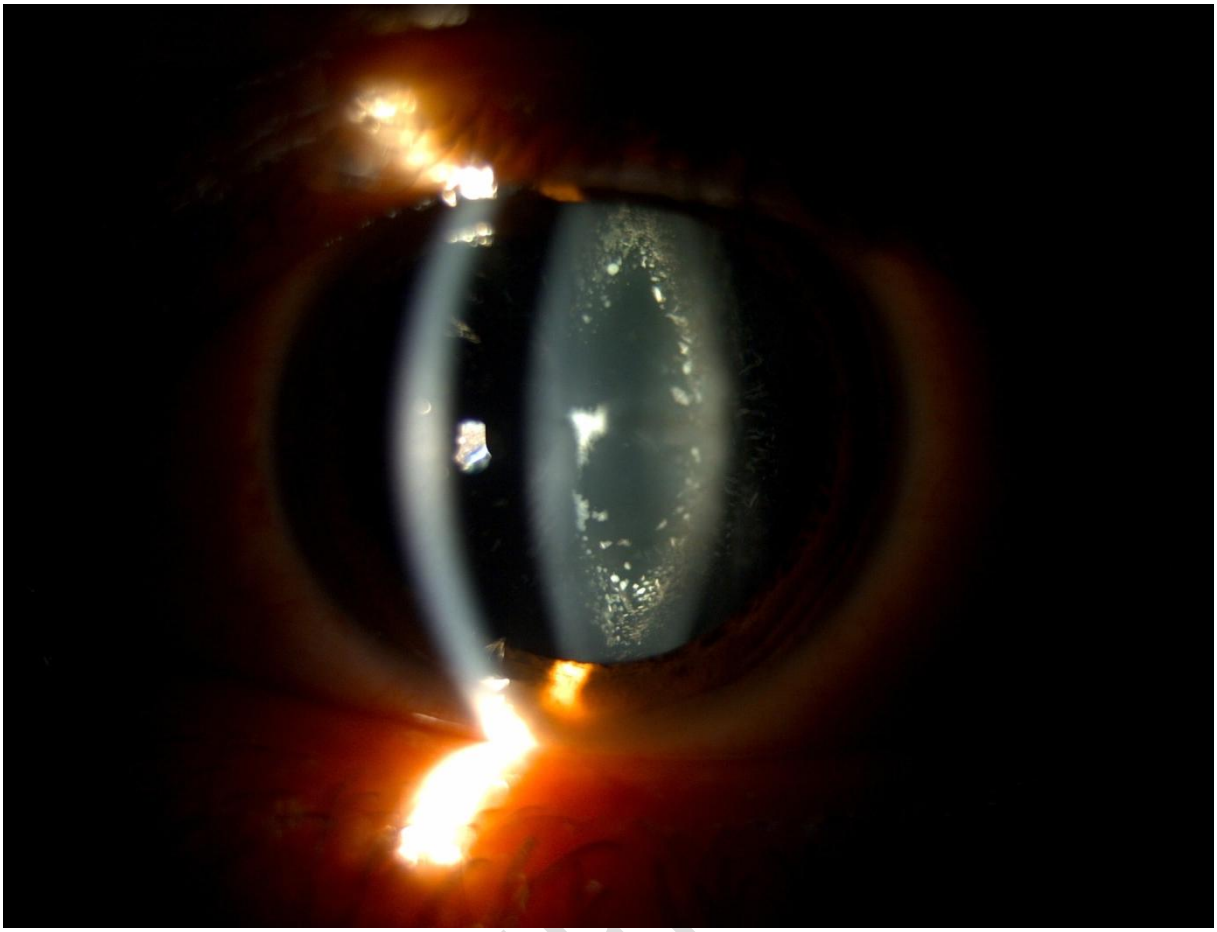

Figure 2: In section: the opacities are radiary in an arc of a circle and sit at the level of the epinuclei.

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