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“MACULAR PUCKER” AS THE LEADING SYMPTOM OF CHRRPE IN YOUNG PATIENTS: DIAGNOSIS AND THERAPEUTICAL CONSIDERATIONS.

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Introduction:

In young patients that present with an opaque epiretinal membrane, the usual diagnosis is “idiopathic” or a “secondary” macular pucker (if other indicative findings are present). Differential diagnosis also includes a combined hamartoma of the retina and RPE (CHRRPE), a disease that is often overlooked. However, the diagnosis is crucial for decision making; in particular, if surgery may be useful or not.

Materials and methods:

With 2 exemplary cases we present characteristic initial findings and postoperative results after vitrectomy/peeling. In both cases only the postoperative workup has led to the correct diagnosis of a CHRRPE in the macula.

Results:

Typical preoperative findings indicative of CHRRPE were present, such as thickened hyperreflective inner retina, resolution of the horizontal retinal layers, saw-tooth inner and outer mini-peaks, gross inner retinal folding (omega sign) and epiretinal membraneous tissue.

Both patients (4 years old girl and 20 years old female) experienced uneventful surgeries and postoperative courses with good anatomic improvement, but stable visual function.

Conclusions:

Rationale for the current presentation is: 1. To present the broad spectrum of fundus changes in CHRRPE that usually is unrecognized. 2. To show the new insights of the disease which are based on modern imaging with HD-OCT and OCTA and that allow a more reliable diagnosis. 3. To continue the controversial discussion, if - in face of the primary dysplastic process - there is a role for vitrectomy in special cases.