Long-term results following intraocular surgery in unsuspected retinoblastoma eyes

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Purpose: To evaluate clinical long term results with a main focus on patient management, metastatic disease and overall survival after vitrectomy in unsuspected retinoblastoma eyes.

Methods: Retrospective, noncomparative case series.

Results: We report on 8 patients referred to a national referral center after pars plana vitrectomy in unsuspected retinoblastoma (from 1992 to 2016). The main preoperative diagnosis included unilateral inflammation in 3 cases, vitreous bleeding after minor trauma in another 3 cases and retinal detachment with PFV and Coats’ disease in the remaining 2 cases. The mean patient age was 4 years (14 months to 6.5 years). The mean interval between vitrectomy and referral was 15 days. All patients were affected unilaterally. Histopathological examination showed an infiltration of the vitrectomy ports in 2 eyes. Treatment consisted of enucleation and systemic chemotherapy of 4 to 6 cycles in all 8 cases. 6 out of 8 patients were treated with adjuvant radiotherapy including EBRT and proton beam irradiation. Of the 8 patients none experienced an orbital recurrence or metastatic disease during a mean follow up of 6.9 years.

Conclusion: Retinoblastoma may present as masquerade syndrome and vitreoretinal surgeons not trained in ocular oncology may mistake retinoblastoma as benign conditions like Coats’ disease or endophthalmitis. If this happens aggressive treatment with enucleation, systemic chemotherapy and/or radiotherapy is the treatment of choice to prevent systemic tumor dissemination but results in severe systemic and orbital morbidity.