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Extranodal right-optic nerve rosai-dorfman disease: review and a new case

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Introduction: Rosai-Dorfman is a rare disease that usually occurs in young adults. It is characterized with massive painless cervical lymphadenopathy and histiocyte proliferation. Isolated intracranial involvement is extremely rare. Our aim is to present a new rare case of extranodal Rosai-Dorfman disease that involves right optic nerve in a four-year-old boy.

Methods: A four-year-old boy with right sided convergent strabismus and amblyopia lasting for one year was treated at the Department of pediatric ophthalmology. Initial optical fundus examination was normal. Examination repeated after one year noted the atrophy of the optic nerve papilla. Visual evoked potentials of the right eye showed normal findings of prechiasmatic visual pathway with severe dysfunction of the right optic nerve. MRI of the brain and orbits showed expansive changed and elongated right optic nerve with contrast enhancement, and smaller lesion in the right temporal operculum region visible in T2 and FLAIR sequence. Through small eyebrow „keyhole“ osteoplastic frontoorbital craniotomy the fusiform enlarged (to 2 cm) right optic nerve was identified, resected between eyeball and optic chiasm and transferred for pathohistological analysis.

Results: Postoperatively, the boy was placed in the ICU Department. Early postoperative course had no complications. Histological, immunohistochemical and ultra structural analyses revealed extranodal Rosai-Dorfman disease. Right periorbital edema was verified on seventh postoperative day and regressed to supportive therapy. Control MSCT of endocranium and orbits showed total tumor removal with no signs of complications. After obtaining histological findings and verification of no neurosurgical complications the patient was transferred to the pediatric Department of Hematology for further treatment.

Conclusion: Although rare, extra nodular intracranial Rosai-Dorfman disease should be taken into account in differential diagnosis of intracranial and intraorbital lesions, especially in pediatric age group.