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Neurofibromatosis Type 1 Revealed By A Retinal Detachment Caused By A Spontaneous Desinsertion At The Oraserrata

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I. Introduction

Neurofibromatosis type 1 (NF1) or Von Recklinghausen disease is the most common phacomatosis. It is an autosomal dominant condition. His diagnosis is clinical with defined criteria. The cutaneous manifestations come to the foreground with the café au laitspots, the other diagnostic criteria includegliomaof the optic pathways, Lisch nodules, sphenoid dysplasia… The originality of our observation is in the atypical association of Neurofibromatosis type1 and upper retinal detachment further to spontaneous desinsertion to the orra.

II. Materials And Methods

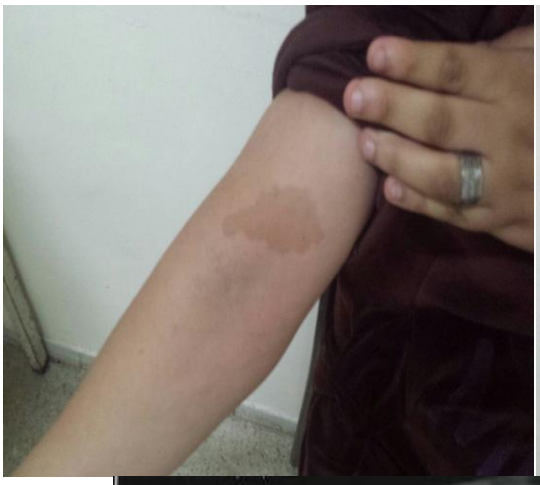
We report the case of a 15-year-old girl, with no significant pathological and traumatic history, who presented to ophthalmic emergencies for a progressive decrease in visual acuity of the left eye. On the general examination, we note the presence in the whole body of cutaneous spots of café-au-lait .Ophtalmic exam shows in the right eye: VA at 3/10 corrected at 10/10, an exophthalmia and bruise of the lower eyelid that go back to childhood, and three nodules of Lisch from the nasal side of iris. At the left eye, the VA was at 1/10 poorly improved, the biomicroscopic examination of the anterior segment found four Lisch nodules on the temporal side of the iris, the examination of the posterior segment visualized a fibrillar vitreous with presence of apigmented Tyndall, and the examination of the fundus and the retinal periphery show an upper retinal detachment from 8h to 3h passing through 12h, a desinsertion to the oraserrata from 10h to 12h, with curled edges, and a macula off. The results of the orbital-cerebral CT was requested which showed exophthalmia grade 2 secondary to the presence of four plexiform neuromas in the optic nerve, as well as the presence of low grade glial tumors (pilocytic astrocytoma) in frontal, inter ventricular septal, and left parietal. The PEV and ERG were altered on the right eye, and abolished on the left. The urgent surgical indication was taken, the patient benefited from a total vitrectomy with cryoapplication of the dehiscence and injection of silicone with simple operative follow-ups.

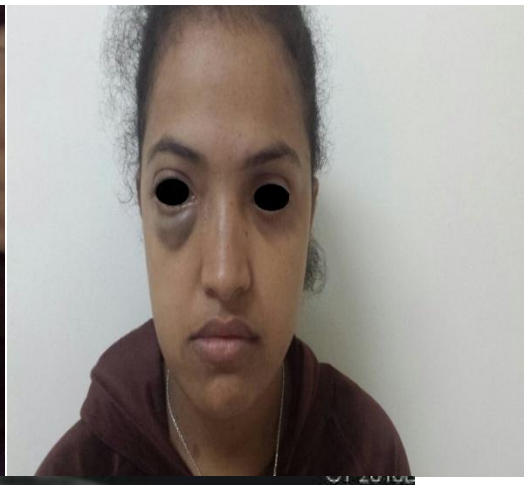
III. Discussion

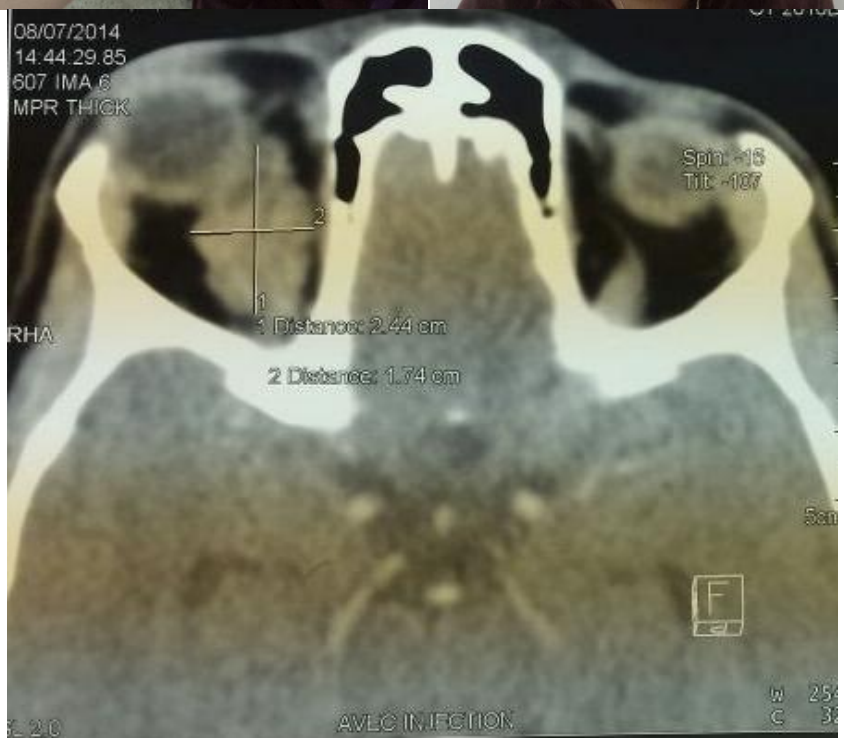
The results of the general and ophthalmological examination made it possible to diagnose in our patient a Neurofibromatosis type 1. It is a frequent dominant autosomal hereditary phacomatosis, due to a mutation of the chromosome 17. The number and the location of the neurofibromas are extremely variable causing polyviscular manifestations, including ophthalmological manifestations, namely Lisch nodules, pleoformneurofibromas, choroidalhamartomas, gliomas of the optic pathway ... To our knowledge, the case we report is the first observation concerning the association Neurofibromatosis type 1 and detachment of retina by spontaneous disinsertion to the ora, since the data of the literature on this subject are non-existent until now

Conclusion

Although the association Neurofibromatosis type 1 and retinal detachment is atypical, it is useful to know it, and to carry out a close examination of the retinal periphery in front of this type of phacomatosis.



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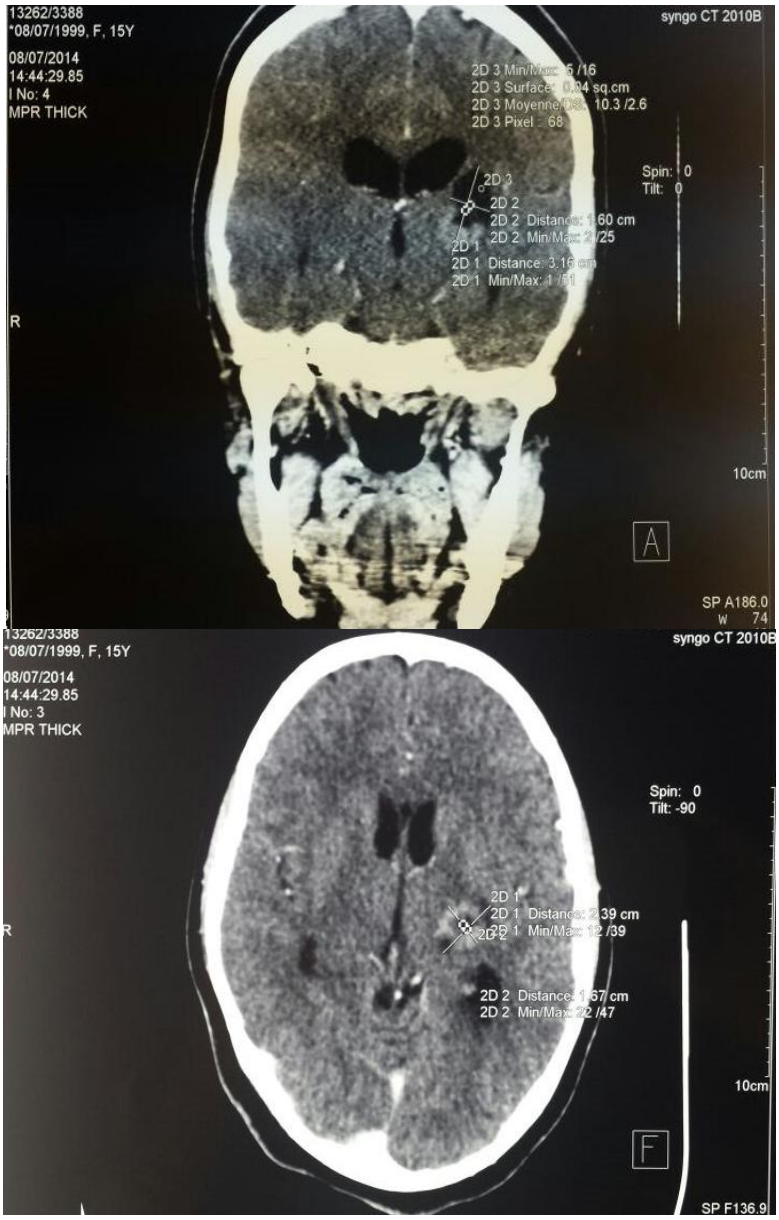

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