

Giant plexiform neurofibroma of the eyelid

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A 3-year-old girl with a history of delayed acquisition consulted for an asymptomatic mass of the right upper eyelid, observed by the mother since the age of 4 months, which had progressively increased in size. Physical examination revealed a 3 cm mass taking the right upper eyelid, of soft consistency with ptosis (Fig. 1a). The child also presented multiple axillary lentiginosities and more than 6 café au lait spots exceeding 5 mm in long axis on the trunk and limbs (Fig. 1b). A cerebro-orbital MRI confirmed that it was a plexiform neurofibroma of the right eyelid associated with dysplasia of the large right sphenoidal wing and glioma of the optic pathways, then she was referred to the ophthalmology department for management.

Neurofibromatosis type 1 (NF-1) is an autosomal dominant neurocutaneous disease. Plexiform neurofibromas (PNs) is pathognomonic of NF-1 and represents a rare kind of this disease where the neurofibroma originates from nerve sheath cells or subcutaneous peripheral nerves [1]. They are often congenital, but they may not be evident immediately after birth, they can increase rapidly in size before the age of 8 years, and they present a risk of malignant transformation [2]. PNs mostly occurs on the trunk and proximal extremities [3], they may be seen on the eyelids but are usually small size. The palpebral mass can be either firm or soft, sometimes the palpation of the involved tissues is said to resemble a 'bag of worms' and can result in ptosis [2]. At this location they tend to infiltrate the orbit, leading to severe proptosis and facial deformity [1], hence the interest in performing a cerebral and orbital MRI when a PN is suspected [2]. Early surgical treatment before the age of 10 years may be necessary for a better functional and esthetic prognosis [2].



Figure 1: (a) A right upper eyelid mass with ptosis. (b) Multiple café au lait spots on the trunk.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

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