Giant neurofibroma: a localization palpebral

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A 4-year-old child with asymptomatic upper right eyelid mass, observed by the mother since the age of one year, having progressively increased in size. A physical examination revealed a mass of 4 cm taking the upper right eyelid (Fig. 1), of soft consistency with ptosis (Fig. 2). Moreover, the child presented more than 6 coffee milk spots > 5 mm long axis, axillary lentigines without palpable adenopathies (Fig. 3). had confirmed that it was a plexiform neurofibroma associated with a dysplasty of the great right sphenoidal wing. the child was referred to the ophthalmology department for the management of his palpebral neurofibroma.

Neurofibroma is a manifestation of neurofibromatosis type 1 (NF1) or Von Recklinghausen disease, who is an oncogenic condition with an autosomal dominant inheritance pattern [1]. His incidence in children with NF1 is less than 10%. It is identified within the first few years of life. It follows the distribution of the trigeminal nerve [1]. It is manifested by a firm or soft palpebral mass with concomitant eyelid edema and it can lead to a ptosis or strabismus. Plexiform neurofibroma mostly occurs on the trunk and proximal extremities and presents as an occasionally pigmented, bag-like mass [2]. It is associated with pigmented spots (coffee coloured) in the skin, commonly seen on the back, abdomen and limbs (café au lait spots). Axillary freckling and lisch nodules may be present [3]. Magnetic resonance imaging (MRI) of the brain and orbits is needed to confirm diagnosis and to define its extent. The

Figure 1: A 4 cm mass involving the upper right eyelid.

Figure 2: Profile view revealing ptosis.
CONSENT

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

REFERENCES


Figure 2: Multiples coffee milk spots > 5 mm long axis.

treatment is mainly surgical, it must be practiced early in order to avoid intraorbital extension and esthetic damage in children.