



A Pediatric Case of Parotid Manifestation of Recklinghausen Neurofibromatosis

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Clinical Image

A 12-year-old girl is referred to our ORL Department. The disease started in childhood with the appearance of multiple hyper pigmented skin macules. Dermatological exam showed of hyper pigmented skin macules on the chest and back, multiple pigmented spots with diameter >1.5 cm (Figure 1), axillary freckling. During oral examination, we saw a formation of neurofibromatic mass on the left side of her face (Figure 2) mucous membranes was not affected. MRI showed the presence of a hypodense left intraparotidial process before and after injection of contrast medium, molding the intraparotidic vascular structure and extending along the facial vascular bundle from the mandibular homolateral angle below, and the region pretracheal at the top, with significant narrowing of the external auditory canal of the homolateral ear.

Neurofibromatosis is genetic diseases that predispose to the development of tumors in the nervous system. There are two essential forms: neurofibromatosis type 1 and neurofibromatosis type 2. This disease cannot be treated. Only complications are treated. The parotidofacial neurofibroma is a readily unique lesion, centered on the parotid region and more or less invading the jugular region. The parotid tissue is invaded by the neurofibroma and the position of the facial nerve is unpredictable, the neurofibroma can be developed from the facial nerve. We can also find a posterior tilt of the pavilion of the ear and occipital osteolytic lesions. Endo-oral extensions should be systematically sought [1].

Bone deformities of the upper and lower jaw are clearly associated with the presence of facial neurofibromas that invade the branches of the trigeminal. A progressive widening of the superior orbital fissure and the oval foramen. The mandible on the affected side is asymmetrical just as the adjacent part of the maxilla and the external wall of the orbit are destroyed [2]. A parotid swelling that can reflect a nervous attack, while thinking of the facial nerve even if its involvement is rare (0.5% to 1%), and studies, as far as possible, its path in front of any tumor in the region parotid [3]. Imaging plays an important role in positive diagnosis, prognostic evaluation, price orientation in therapeutic charge and progressive follow-up of lesions. Treatment is surgical but abstention and monitoring may be indicated because the progress is slow.

The particularity of our observation is that it a rare presentation of the disease in children

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Figure 1: Multiple hyper pigmented skin macules pour la plus grande measurement 7cm.



Figure 2: Neurofibromatic masse on the left side of her face.



Figure 3: Expanding left parotid process with partial agenesis of the large left sphenoid wing.

as well as the interest of imagery to evoke the diagnosis of parotid neurofibroma and assessment of locoregional extension and the follow-up of these tumors.

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