ORO-DENTAL ASPECTS IN A PEDIATRIC CASE WITH TYPE 1 FAMILY NEUROFIBROMATOSIS WITH NEPHROGENIC DIABETES INSIPIDUS

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Images of: Patient, female, ages 7, weight 21 kg and 112,5 cm height - under the normal height average for this age (122.52), presented at the pediatric medical consultation at the age of 2 and a half, when the mother noted the appearance of some spots, axillary ephalides, and in the left eye an important tumefaction. Based on the anamnesis, clinical examination and magnetic resonance imaging - cerebral MRI - left retro-orbital neurofibroma diagnosis (Fig. 1), type 1 family neurofibromatosis (genetic disease with dominant autosomal transmission)

Figure 1. Patient, female, 7 years old - facial appearance.

diagnosis is established, and blood tests confirm the suspicion of nephrogenic diabetes insipidus. Our hypothesis on etiology of nephrogenic diabetes insipidus in this patient is that it could be hereditary; the maternal grandmother and the patient's brother being diagnosed with insipidus diabetes.

General clinical examination: reveals numerous café-au-lait spots with different localizations (face, bilateral armpits, abdomen and posterior thorax) (Fig. 2). In the left eye there is a retro-orbital tumor, a neurofibroma that produces ocular globule occlusion and left palpebral paresis. Skeletal abnormalities are present, patient had scoliosis and bowing of the lower legs. Also the patient showed cognitive impairment, attention deficit hyperactivity disorder (ADHD) and learning disorders.

Facial examination: reveals a facial accentuated asymmetry on the left side consecutive to the neurofibroma paresis of the left hemiface (Fig. 1). At the level of the left genian region, there are café-au-lait spots.

Functional examination reveals functional disorders of mastication, swallowing and physiognomy. The patient has sialorrhea.

Figure 2. Patient, female, 7 years old - trunk appearance.

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Figure 3. Patient, female, 7 years old - oro-dental appearance.

At the endobuccal level (Fig. 3), there are dental abnormalities of number, shape and rash. The patient has mixed dentition - dental status (Fig. 4) consistent with age. The crowns of the superior central incisors (1.1 and 2.1) are more bulky - blocking the access in the oral cavity of the lateral incisors (1.2 and 2.2). The absence of the first lower deciduous premolars (7.4 and 8.4) is noted. Carious dental morbidity is moderate (Fig. 3).

		Right										Left											
		O.D.	-	-	-	X	P					P	X	-	-	-	O.D.						
1.8	1.7	1.6	1.5	1.4	1.3	1.2	1.1	2.1	2.2	2.3	2.4	2.5	2.6	2.7	2.8								
Maxillary		P	P	P	-	-				-	P	P	P					Maxillary					
		5.5	5.4	5.3	5.2	5.1	6.1	6.2	6.3	6.4	6.5												
Mandible		8.5	8.4	8.3	8.2	8.1	7.1	7.2	7.3	7.4	7.5					Mandible							
		M.D	X	P	-	-			-	P	X	O.D.											
4.8	4.7	4.6	4.5	4.4	4.3	4.2	4.1	3.1	3.2	3.3	3.4	3.5	3.6	3.7	3.8								
		O.D	-	-	-	P	P	P	P	-	-	-	O.D										
		Right										Left											

Figure 4. Patient, female, 7 years old – dental status (P=present; O.D.=occlusal decay; M.D. mesial decay; X=missing teeth).

The lower dental arcade shows inconsistency between the volume of the mandibular alveolar bone and the dental volume, thus inducing a dental-alveolar crowding disharmony.

Gingival bleeding when brushing is noted.

Conflict of interest

The authors declare that they have no conflict of interest.