McCune-Albright Syndrome revealed by maxillary pain in adulthood: a review and case report

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Case Report

ABSTRACT

McCune - Albright syndrome is a rare sporadic disease defined by the triad: café-au-lait spots, fibrous bone dysplasia and endocrinopathy We report the case of a McCune - Albright syndrome of fortuitous discovery in a 70-year-old patient referred to the OMFS department for the management of chronic right maxillary pain, associated with headache, homolateral lacrimation and nasal obstruction. The examination of our patient reveals a facial asymmetry due to a maxillary tumefaction, a median mass mobile on swallowing, and cafe-au-lait spots of right scapular and abdominal regions. Facial CT scan shows an osteolytic and osteocondensing process of the right maxilla with extension to the nasal cavity and the hard palate. Cervical ultrasound found a multinodular goiter, with subclinical hyperthyroidism on thyroid checkup. The association of FD, hyperthyroidism, and cafe-au-lait spots classified the patient as a carrier of McCune-Albright syndrome. We opted for maxillary shaping surgery to meet the aesthetic demand and to improve the respiratory function of our patient. Histological examination confirmed the diagnosis of fibrous dysplasia. Follow-up of the patient after 2 years showed a disappearance of the maxillary pain and nasal obstruction with restoration of facial symmetry.

Key Words : McCune-Albright Syndrome; fibrous dysplasia ; bone remodelling

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INTRODUCTION

Fibrous dysplasia (FD) of bone is a benign pathology characterized by a medullary proliferation of fibrous tissue containing immature osteogenesis, sometimes causing pain, fractures, or even bone deformities. The lesion may be unique (monostotic form), multiple (polyostotic forms), or associated with other lesions forming the McCune-Albright syndrome (MAS) with a triad classically associating fibrous bone dysplasia, cafe-au-lait spots and endocrinopathy. We report the case of a McCune-Albright syndrome discovered following a maxillary pain in an elderly patient in order to recall the clinical, histological, therapeutic and evolutionary characteristics of this pathology.

CASE REPORT:

The patient is a 70-years-old woman referred to the oral and maxillofacial surgery department by her dentist for management of chronic right maxillary pain that had been evolving for 3 years and was improving little under analgesic and anti-inflammatory treatment. Our patient had undergone multiple avulsions of maxillary and mandibular teeth over the past 3 years. Moreover, the patient declared the appearance of a nasal obstruction and a right lacrimation evolving in the past 6 months, associated with frontal headaches.On examination, we noted a right maxillary swelling responsible for facial asymmetry with erasure of the right nasolabial fold (figure 1). This mass is integral with the maxillary bone, painful to palpation, hard in consistency, without associated redness or heat. Oral examination revealed a partially edentulous maxilla and mandible and a right maxillary mass with a buccal filling extending from tooth N 1 to tooth N 8, and a bulging of the homolateral hard palate. Anterior rhinoscopy showed a budding process in the right nasal cavity

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Figure 1. Frontal view showing a right maxillary tumefaction with facial asymmetry

responsible for an obstruction with an inflammatory aspect of the nasal mucosa. At the cervical level, we found a median cervical mass mobile on swallowing, with the presence of a left nodule of about 4 cm long axis, painless, hard, without compressive signs or cervical lymph nodes (figure 2).

Figure 2. Frontal view showing a cervical median mass mobile on swallowing

Finally, dermatological examination found cafe au lait spots at the scapular level of 3 cm long axis and abdominal level of 5 cm long axis on the right side, with irregular borders (figure 3).

Figure 3. Cafe au lait spots at the right scapular level measuring 3 cm

To explore the maxillary mass, our patient underwent a CT scan that revealed a tissue tumor process centered on the right maxillary, measuring 45x40x42 mm, of mixed density, both osteolytic and osteocondensing. This process invades the internal wall of the right maxillary sinus with extension in the right nasal cavity and septum, and the ipsilateral bony palate (figure 4).

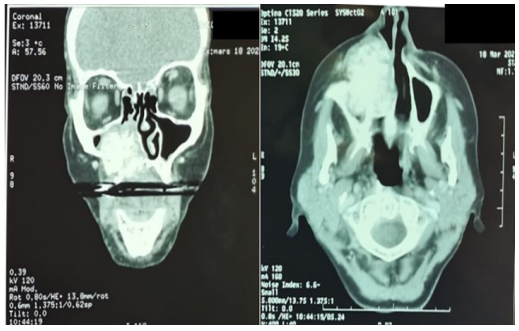

Figure 4. Coronal and axial CT scan showing a proliferation right maxillary process with extension into right maxillary sinus and right nasal cavity

A biopsy was performed and was in favor of fibrous dysplasia. Moreover, cervical ultrasound found a multi hetero nodular goiter classified TI-RADS III, the largest nodule is left lobar measuring 50 mm in diameter, while the thyroid workup showed a low TSH and normal level of thyroid hormones signing a subclinical hyperthyroidism. Based on the clinical, radiological and histological data, the decision was made to perform a remodeling conservative surgery of the maxilla in order to meet the patient's aes thetic request, but also to treat the rhinological symptoms that were progressively setting in. The surgical exploration allowed the resection of a friable bone developed at the expense of the right maxillary sinus and the right nasal fossae essentially. Histological examination of the resection specimen showed fibrous dysplasia. Follow-up of the patient after 2 years showed a disappearance of the maxillary pain and nasal obstruction with restoration of facial symmetry, with persistence of palatal bulging.

DISCUSSION:

Fibrous dysplasia is a benign bone disorder characterized by the replacement of normal bone tissue with fibrous-like tissue. It remains a rare condition, with an estimated prevalence of less than 1 in 2,000, accounting for approximately 2.5% of all bone lesions and 7% of benign tumor-like bone lesions [1,2]. Although it can be diagnosed at any age, the majority of cases (90%) are identified before the age of 40. Late diagnoses are more frequent in women and are almost exclusively observed in those over 50 years of age [3]. An even rarer form, McCune-Albright syndrome (MAS), is estimated to affect between 1 in 100,000 and 1 in 1,000,000 individuals [4]. At the craniofacial level, fibrous dysplasia can lead to bone pain and pathological fractures [5],

The feeding duration and amount per feeding were used in order to calculate the outcome measure, feeding efficiency (mL/min). Feeding efficiency was calculated by the investigator for each appliance. The measure was defined as the volume of fluid transferred per unit time, in milliliters per minute (mL/min). Evaluation The feeding efficiency (mL/min) was calculated for each baby. Babies' visits due to discomfort necessitating adjustments to their feeding appliances were counted.These adjustments were essential to relieve pain caused by the appliance.

Results

The Acrylic feeding plate demonstrates significantly higher feeding efficiency compared to both the Flexible feeding plate and the Special feeding bottle. There is no statistically significant difference in feeding efficiency between the Flexible feeding plate and the Special feeding bottle (Table 1-2) The Flexible feeding appliance was associated with a significantly higher number of adjustment visits compared to both the acrylic feeding plate and the Special feeding bottle. There is no significant difference between the acrylic feeding plate and the Special feeding bottle in terms of adjustment visits (Table 1-2)

Table1: Comparisons of Feeding Methods for Feeding Efficiency and Adjustment Visits

		N	Mean	Std. Deviation	Minimum	Maximum	median	F	Sig
feeding efficiency	Acrylic feeding plate	10	9.4300	1.66283	7.32	12.03	9.5	57.4	.001*
	flexible	10	5.2930	0.79720	4.28	6.54	5.3		
	Special feeding bottle	10	4.0390	0.86861	2.94	5.40	4.1		
adjustment visit	Acrylic feeding plate	10	1.6000	0.69921	1.00	3.00	1	39.1	.001*
	flexible	10	3.4000	0.51640	3.00	4.00	3		
	Special feeding bottle	10	1.3000	0.48305	1.00	2.00	1.5		

The mean difference is significant at the 0.05 level.

Table 2: Mean Differences and Significance from Post-Hoc Comparisons of Prosthetic Appliance Types on Feeding Efficiency and Adjustment Visits

			Mean Difference (I-J)	Std. Error	Sig.
Feeding efficiency	Acrylic feeding plate	Flexible feeding plate	4.13700*	0.52631	0.000*
	Acrylic feeding plate	Special feeding bottle	5.39100*	0.52631	0.000*
	Flexible feeding plate	Special feeding bottle	1.25400	0.52631	0.073
Adjustment visit	Acrylic feeding plate	Flexible feeding plate	-1.80000*	0.25676	0.000*
	Acrylic feeding plate	Special feeding bottle	0.30000	0.25676	0.759
	Flexible feeding plate	Special feeding bottle	2.10000*	0.25676	0.000*

The mean difference is significant at the 0.05 level.

Discussion

The present randomized clinical trial evaluated the feeding efficiency of three management strategies for neonates with complete unilateral cleft lip and palate:(A) acrylic rigid heat-cured polymethyl-methacrylate (PMMA) feeding plates,(F) vacuum-formed 1.5 mm thermoplastic feeding plates,and (T) no plate but a specialized silicone cleft teat. The primary outcomes were feeding efficiency and the number of adjustment visits.Acrylic feeding appliances offer superior pre-surgical feeding performance for these infants. The Flexible feeding appliance is associated with a significantly higher number of adjustment visits compared to both the acrylic feeding plate and the Special feeding bottle. Feeding plate aim to obturate the palatal defect to create negative intra-oral pressure, shield sensitive tissues from teat trauma, and guide tongue posture to facilitate suck–swallow–breath coordination [2]. Heat-cured PMMA remains the clinical gold standard because of its rigidity and dimensional stability. Vacuum-formed thermoplastic sheets are increasingly advocated because they are resilient and can be fabricated rapidly without polymerization monomer exposure [9,10]. The present study corroborated these advantages. Consistent with the observational study [9], the PMMA feeding plate significantly improved feeding efficiency relative to the no-plate special teat group.By sealing the oronasal communication, the feeding plate permits generation of intraoral suction pressures comparable to non-cleft neonates [8].

Notably, the acrylic plate delivered higher mean milk transfer rates than the flexible appliance The rigid plate supplies a firm, stable surface that lets the infant draw milk efficiently [12-14], whereas the flexible vinyl plate can flex, warp, or press into palatal undercuts, causing shifts and flexible tissue injuries that lead to more adjustment visits. The maintenance of the flexible vinyl appliance is particularly sensitive, as the use of warm or hot water during cleaning can result in distortion. This distortion may lead to increased flexible tissue injuries and necessitate more frequent adjustment visits. There are few clinical studies that have directly compared flexible vacuum-formed feeding plates with conventional rigid acrylic feeding plates in infants with cleft lip and palate; the published evidence on flexible plates usually consists of isolated case reports and tiny case series. [6,11.10] underscoring the need for the present randomized trial.

Conclusion

Babies generated better levels of feeding when they used an acrylic feeding appliance in comparison to the flexible appliance and specialized feeding teats.

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