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Congenital Agenesis of Columella, Associated with an External Auditory Canal Atresia: Case Report

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Abstract

Background: Congenital agenesis of columella is an abnormality extremely rare. Only five cases have been reported in the literature to 1999, a fifth case accompanying a facial haemangioma was presented in 2004, and a seventh case was described in 2019, the etiology is still unclear.

Case presentation: We present a case of congenital aplasia of the nasal columella. The patient is a 5-month-old female infant, and we show that this rare malformation has not only aesthetic, but also serious functional repercussions, which may necessitate early surgical intervention. We also highlight the incidental discovery on Tomodensitometry of the face and petrosal bone, of an associated atresia of the external auditory canal not evident at the clinical examination, an association never reported, or perhaps not sought in the observations hitherto available.

Conclusion: The association observed in this case report with an atresia of the external auditory canal shows the importance of requesting tomodensitometry of the face and petrosal bone, in similar cases.

Key Words: Agenesis, Columella, Congenital, External Auditory Canal.

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1- INTRODUCTION

Agenesis of nasal columella is the absence of a tissue that connects the nasal tip to the nasal base and separates the nares (1). For the infant, the small size of the nose makes the anomaly not very bad on the aesthetic plan, which could be the argument of the surgeons to defer the intervention to a more advanced age, but we want with this work to show the functional importance of this small structure, at the inferior margin of the nasal septum, through the observations on a 5-month-old female infant. This work also stresses the importance of the search for other malformations not always obvious, at the clinical examination, such as atresia of the external auditory canal found in this infant.

2- CASE PRESENTATION

A five-month-old female infant was admitted to our hospital with respiratory problems. A three-day period prior to admission, she had clear rhinorrhea complicated by breathing difficulty and refused the baby bottle. At admission, the infant was febrile, tonic, reactive, and had a rosy complexion. However, there was a slight tugging at the subcostal area. Facial examination revealed columellar agenesis; mouth, and lips were normal. The examination of both ear pavilions finds normal ear pavilions with presence of damp appearance in the external orifice of the right ear making difficult an examination at the otoscope and evoking an otitis (Fig. 1).

The same day of his admission, the child was put under amoxicillin and clavulanic acid, and there was permeable choanae, normotensive anterior fontanelle, abdomen supple, no abnormality on cardiac auscultation, and discreet rhonchi on pleuropulmonary auscultation. The patient weighed 4Kg100g (-3DS) and weighed 59cm (-3DS).

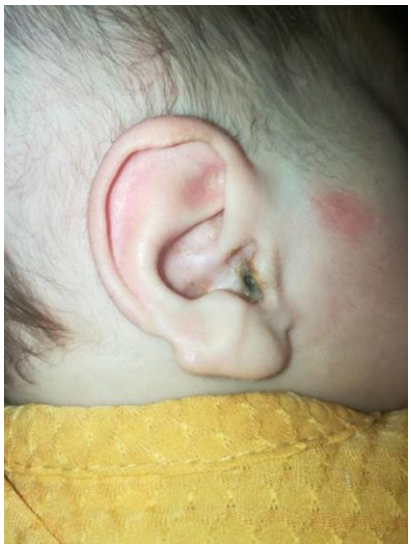

Fig. 1: Right ear pavilion

The biochemistry and radiologic tests conducted at admission were all normal, as were urea, creatinine, C- reactive protein, and lung x-rays. More respiratory comfort was noted after nasal unblocking. And we noticed that the nostrils were directly connected, without columella, collapsed at inspiration, and this hindered breathing. The nasal orifice was flat and narrow even with a clean nose (Fig. 2), and the obstruction increased with rhinorrhea on inspiration.When we attempted to maintain the nostrils open, we found the device shown below (Fig. 3), but its use wasn't feasible at this age, due to the high risk of ingestion and false routes.

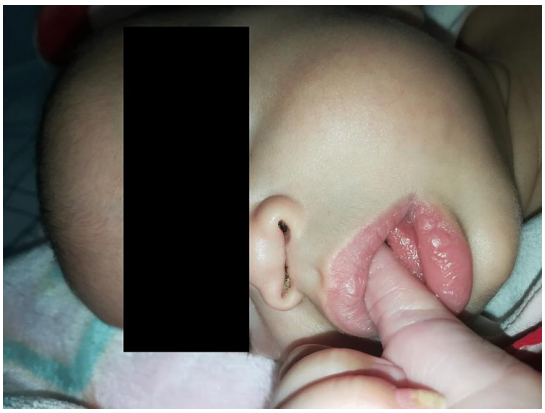

Fig. 2: The nasal orifice was flat and narrow even with a clean nose, we note the impact of the absence of the columella as a support to the nasal tip

Fig. 3: Not used due to the risk of introducing it in the mouth and false routes

We then adapted oxygen goggles by opening them with scissors to allow the infant to breathe easily in ambient air (Fig. 4).

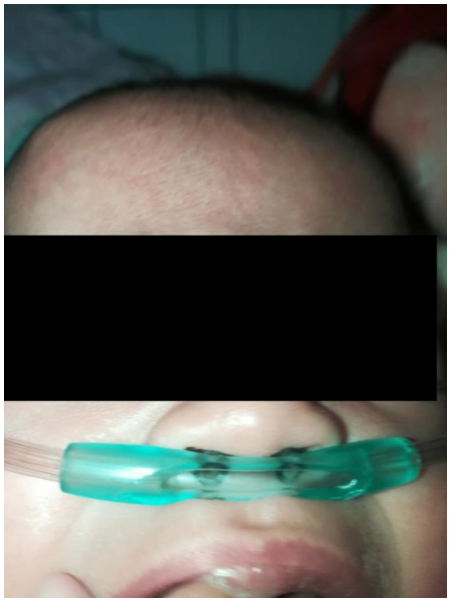

Fig. 4: Oxygen goggles opened with scissors so as to allow the infant to breathe easily

Just the installation of this device made it possible to eliminate respiratory fights and increase oxygen saturation from 95% to 98%, without connection to an oxygen source. The infant could then breathe freely through his nose, and drink his milk in a bottle without difficulty. A simple tool used for keeping the nostrils open had helped the infant well in respiring, but it needed to be changed to prevent its obstruction. After the infection episode, the inspiration's collapsing effect reduced. Clinical improvement was noticed after three days in hospital (Fig 5 and Fig. 6).

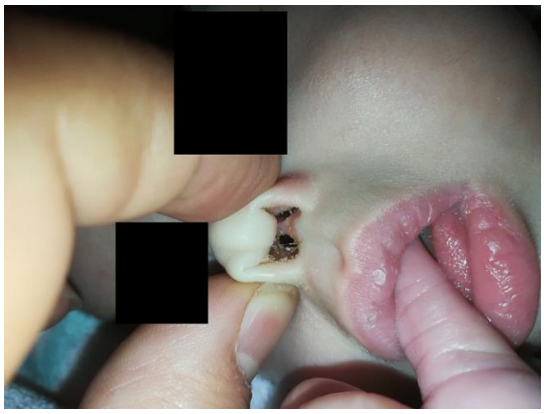

Fig. 5: After three days in hospital

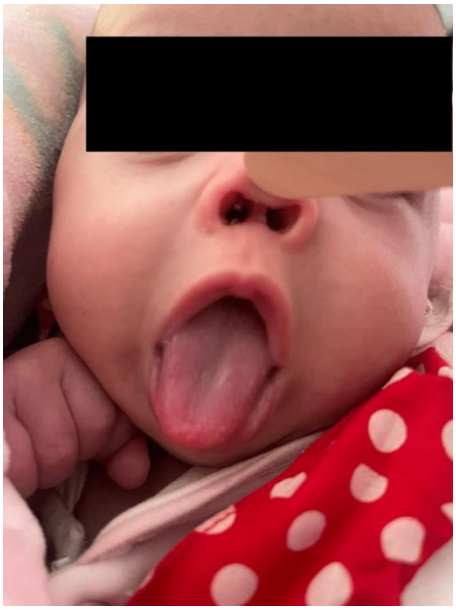

Fig. 6: After six days in hospital

A goggles was removed, and regular nasal cleansing was recommended. We did a malformation assessment; heart

ultrasound, transfontanellar ultrasound and abdominal ultrasound returned normal. We were reluctant to irradiate infants with tomodensitometry of the face and petrosal bone, especially after the clinical improvement; but we finally demanded it, especially in the vision to present a satisfactory medical record to the surgeon. We were surprised by the results referring to an atresia of the right external auditory canal, the ossicular chains and the elements of the inner ear were normal (Fig. 7).

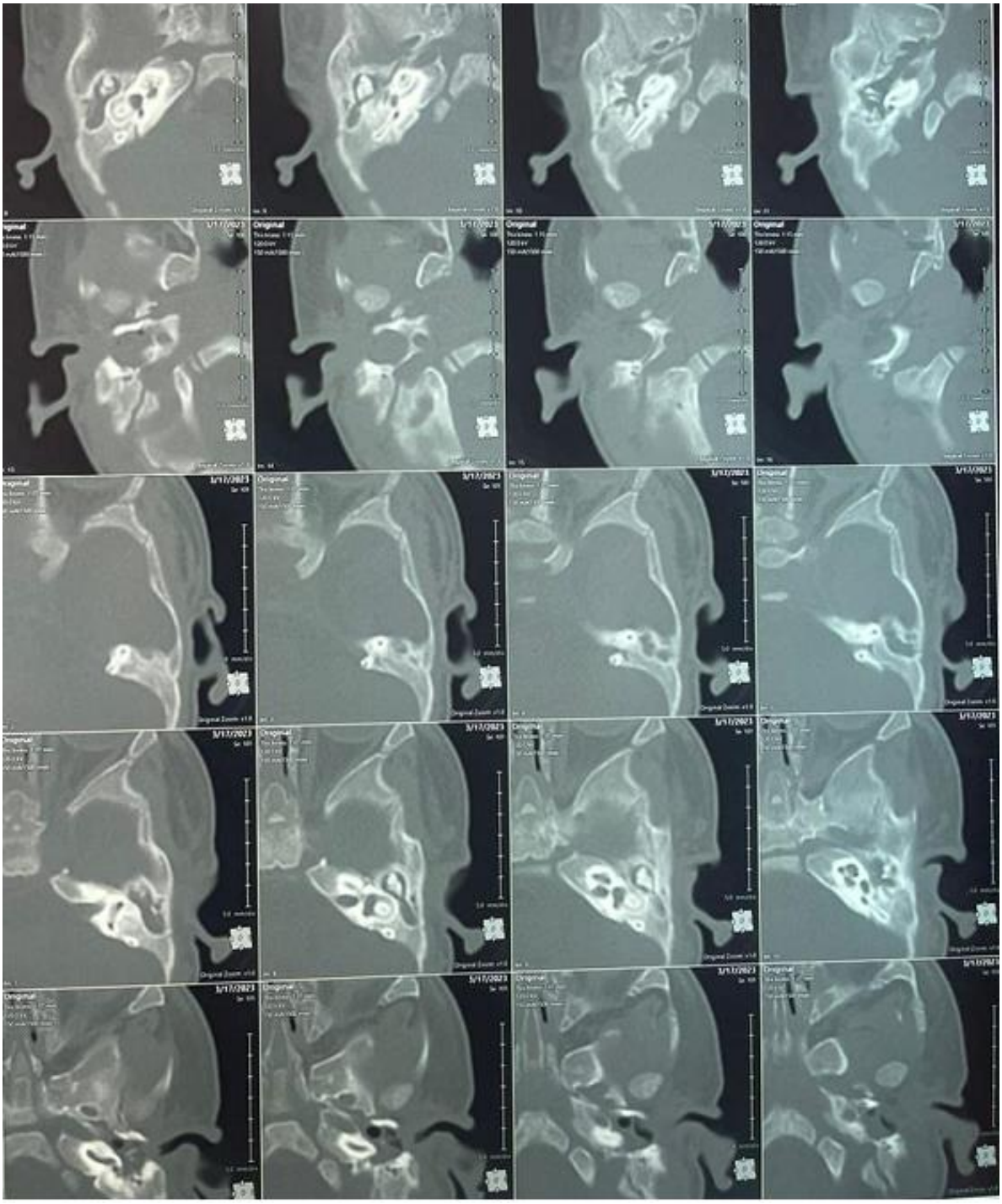

Fig. 7: Atresia of the right external auditory canal on Tomodensitometry

3- DISCUSSION

Congenital agenesis of columella is an extremely rare anomaly (2, 3). For the infant, the small nose size makes the anomaly not very ugly on the aesthetic plan. This could explain why parents often do not seek medical attention until late childhood or adolescence. The etiology of congenital absence of nasal columella is still unclear (2). The embryonic development of the nose takes place between the 3rd and 10th weeks of gestation (4). The medial nasal processes fuse in the midline with the frontal prominence, resulting in the formation of the frontonasal process which give origin to the columella (2, 4). This supports the assumption that teratogen introduced during columellar development might selectively arrest cellular penetration and impede chondrification of the nasal columella (2). The columella provides support to the nasal tip. Its absence in the case we report has aggravated an episode of upper respiratory infection by provoking a collapse of the nostril orifices at inspiration, in addition to an obstruction by nasal secretions. A simple tool used at keeping the nostrils open helped the infant breathe more easily. However, as described, occlusion of the nose in an infant may and can prove to be fatal (5); even claims have been made in the medical literature that infants breathe between six weeks and six months exclusively through their noses (6). The functional impact of columella absence may indicate an early surgical intervention. Surgical reconstruction of an absent nasal columella is challenging because of its complex anatomy. The columella is the subunit between the two nostrils that, along with the lower lateral cartilages and caudal septum, provides support and projection to the nasal tip. It plays a functional role in nostrils patency and nasal breathing (7). It has very specific characteristics in terms of texture, morphology and function. The missing columella cannot always easily find an ideal substitute that integrates perfectly with the neighboring integuments of the central part of the face (8).This necessitates a particular finesse in order to obtain optimal outcome, both functionally and aesthetically. Tomodensitometry of the face and petrosal bone was not systematically requested in all cases of congenital agenesis of columella reported in the literature, We have asked for it especially in the vision to present a satisfactory medical record to the surgeon, and coincidently, we found an atresia of the right external auditory canal, We had already pointed out the rarity of the columellary agenesis in the literature, but concerning the association with an agenesis of a distal portion of the external auditory canal, we could not find such cases presented in the literature, from another point of view, as the etiology of columnar agenesis is still not precise pathogenetically. Its relation with an external auditory canal atresia remains even more ambiguous. Atresia of external auditory canal although frequently isolated (in cases which the abnormality is less severe), can be associated with a number of syndromes, including Crouzon syndrome, Treacher Collins syndrome, Goldenhar syndrome, and Pierre Robin syndrome (9). But our case didn’t fit any of these syndromes, hence showing the uniqueness of this observation. From another point of view, Tomodensitometry of the face and petrosal bone was not systematically requested in all cases of congenital agenesis of columella reported in the literature. Hence we were faced with the following questions: Were there similar undiagnosed associated malformations in the previously described observations? And must we systematically demand a tomodensitometry of the face and petrosal bone when we find a congenital columellar agenesis on clinical

examination? I think the questions deserve to be asked, in view of the risk of the missing case of deafness in the child, especially as neonatal screening for deafness is not yet widespread in all countries.

4- CONCLUSIONS

The functional impact of columellar agenesis could be more alarming for the infant than the aesthetic one. The surgical team of the hospital initially affirmed to have never seen such a malformation before; the decision was taken a month ago to delay the surgical intervention until preschool age; but with the description of the case as it was presented in our service with significant functional repercussions, they decided to coordinate with a team of maxillofacial surgery and otolaryngologists to discuss the surgical intervention in the shortest possible delay. The association in this case report with an atresia of the external auditory canal shows the importance of requesting tomodensitometry of the face and petrosal bone, in similar cases.

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