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Congenital Naevus: Case Report of an Atypical Atrial Location

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Abstract: Congenital naevi are a benign proliferation of melanocytes. Present at birth or appearing in the first weeks of life, they have the appearance of light to dark brown lesions, sometimes bluish, more or less hairy. It is a non-hereditary mosaic pathology. They are due to postzygotic somatic mutations involving key proteins in the mitogen-activated protein kinase pathway, mainly NRAS and BRAF. Its prevalence is estimated to be between 1 and 3% of newborns. This disease can affect any part of the skin. There are 3 types of naevi: small, medium and large naevi then called giant. The objective was to discuss the clinical and therapeutic aspect of a congenital auricular naevus of medium size. We report the case of an 18 years old girl presenting a sessile tumefaction at the level of the concha of the right auricle that appeared from birth, firm, rough, unique, about 5 cm in diameter, homogeneous in color, pigmented, hairy, painless to palpation, apyretic sometimes pruritic without any other associated sign. The diagnosis of benign congenital naevus was clinically made after confirmation by the dermatologist. Surgical removal was performed. The postoperative course was simple with complete healing after one month. The middle congenital naevus is a benign tumor linked to a genetic variation of low incidence, which is all the more rare as it is located on the ear, thus having a significant psychological impact due to its often unsightly appearance. The diagnosis of this genodermatosis is essentially clinical. When it is an isolated nodule, as in our study, total surgical removal is a good alternative with simple after-effects and an almost non-existent risk of malignant transformation.

Keywords: Congenital Naevus, Auricular Nodule, Psychological Impact, Genodermatosis, Medium Naevi, Surgical Excision, BRAF Mutations

1. Introduction

Congenital naevi are benign proliferations of melanocytes. Present at birth or appearing in the first weeks of life, they have the appearance of light to dark brown lesions, sometimes bluish, more or less hairy. Their size varies and their growth is proportional to that of the child. It is established that congenital naevi are the result of a genetic variation that occurred in utero, after fertilization and the first cell divisions. They are due to postzygotic somatic mutations involving key proteins in the mitogen-activated protein kinase pathway, mainly NRAS and BRAF. It is a non-hereditary mosaic disease. No risk factors favoring the occurrence of such a variation have been identified to date [1-3]. They are frequent: although the prevalence is not precisely known, it is estimated to be between 1 and 3% of newborns. The incidence of large or giant congenital nevi is lower, ranging from 1/20,000 to 1/500,000 births [1]. This disease can affect any part of the skin. It can also exist on

non-visible parts (inner membranes). When it is present in the nervous system, it is called neuromeningeal melanosis [4]. Congenital atrial melanocytic naevi are rare and only a few cases have been reported in the literature. Indeed, in a study of the clinical analysis of benign auricular masses over a period of 41 years, only one case of congenital ear naevus was found [5]. The naevi are classified into 3 types: small, medium and large naevi, then called giant. Indeed, depending on the size and location, the presence of a naevus can be a source of psychological stress due to its unsightly appearance.

The objective was to discuss the clinical and therapeutic aspect of a congenital nevus of atypical auricular location of medium size.

2. Observation

We report the case of an 18-year-old female patient admitted to the department for right atrial swelling. The appearance of the mass dated from birth according to the patient painless, apyretic, firm associated sometimes with itching having progressively increased in volume in early childhood. We note an important psychological impact: the patient complained about its unaesthetic aspect and covering the ear permanently.

Physical examination revealed a firm, rough, single mass about 5 cm in diameter, sessile at the level of the concha of the right auricle, homogeneous in color, heterogeneous in texture, pigmented, hairy, painless to palpation [figure 1].

The diagnosis of medium congenital naevus was made after confirmation by the dermatologist in front of a pigmented, rough, heterogeneous, hairy swelling between 1.5 and 20 cm in diameter. We performed the surgical excision under local anesthesia, the removal of the nodule was complete with closure by direct suture, simple and without distortion of adjacent tissues. The postoperative course was simple with complete healing after two weeks and disappearance of the pigmentary trace after one month. We did not lose sight of the patient during the long term.

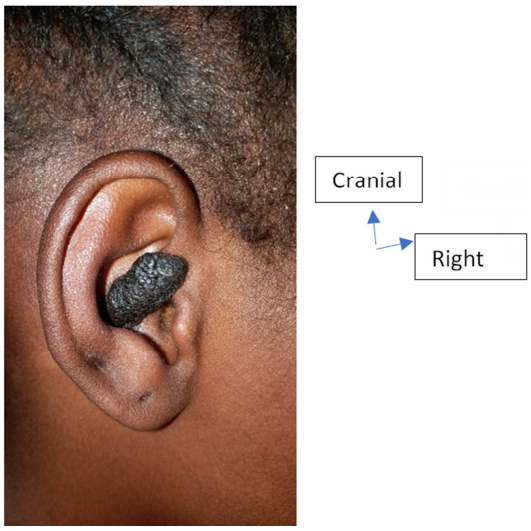

Figure 1. *Auricular Congenital Naevus.*

3. Discussion

For three quarters of a century, knowledge of congenital naevi has not been revolutionized, and many uncertainties remain. These cutaneous tumors, poorly understood in their ontogeny, have a prognosis that remains unclear [1].

Melanocytic naevi are congenital or acquired benign proliferations of cells of melanocytic origin. Congenital naevi are the result of a genetic variation that occurred in utero, after fertilization and the first cell divisions. [6] They can be isolated or multiple and there is a risk of developing a melanoma, particularly in the case of giant naevi. The risk is extremely rare before puberty and is estimated at 1% [6, 7, 8]. Congenital nevi are clinically diagnosed [1, 6] and are usually asymptomatic, with itching, dryness and local fragility of the skin [4]. In our case, we could only note the presence of itching.

Indeed, extensive and giant naevi are unexpected birth events that are very shocking for the parents and family, and the morbidity associated with the heavy surgical programs that are proposed can affect their lives.

Our patient presented with a single well-limited nodule of average size, whereas in the literature, the size and number of naevi can be very variable, ranging from a single lesion of a few millimeters to several tens of centimeters, to innumerable lesions, with or without large lesions (multiple congenital naevi). In the most severe forms, up to 80% of the skin surface may be affected. [9].

Large and multiple naevi are predominantly related to NRAS mutations, whereas small to medium naevi are preferentially the result of BRAF mutations [10]. In our case, it is most likely a BRAF codon V600E mutation. It accounts for 5-15% of them [2, 6, 11, 12].

This pathology affects all populations and both sexes, with a slight female predominance [13].

The incidence of medium-sized naevi is 0.67% [14]. The diagnosis of congenital naevi is clinical [1, 6]. The Krengel classification [10] is a descriptive clinical classification that allows the categorization of congenital nevi according to their clinical characteristics: topography, number of naevi at the age of 1 year, degree of roughness, color heterogeneity, hypertrichosis, presence of proliferating nodules, projected adult size when the diagnosis is made in early childhood. This classification allowed us to make the diagnosis of medium congenital naevus in front of the pigmented, rough, heterogeneous, hairy tumefaction between 1.5 and 20 cm in diameter in our female patient.

When they are of medium size, as in our study, the main location is either on the face, the limbs or the abdomen. The particularity of our case lies in its auricular localization, which is rather rare in the literature [5]. The ear is indeed [15].

The congenital naevus is an apparent congenital skin feature. The awareness of having a congenital naevus is not innate, but is acquired and reinforced or weakened by situations and events. Throughout childhood, the child may face mockery, exclusion and stigmatization [16].

Although the naevus was of average size, our patient, just

out of adolescence, was strongly marked by it.

The management of nevi is most often surgical under local anesthesia, it can also be done using laser to treat pigmented lesions (CO_2, Ruby, Yag, alexandrite) [16, 17, 18].

In the neonatal period, curettage (Moss technique) can also be applied with good results. Therapeutic abstention can be defended when the after-effects of a major surgery and its scarring are negatively weighed, without ignoring the psychological impact of medicalization and repeated hospitalizations, especially in cases of extensive naevi. For small lesions, abstention is the basic course of action, but a personal request for aesthetic convenience, after explanations on the scarring risk, deserves to be considered [1].

In our case, the surgical removal was performed at the patient's request, as is most often the case in the literature. It was the reason for consultation, essentially for aesthetic reasons. Indeed, the presence of a naevus can have an important psychosocial impact on the patient and her entourage because of its unsightly appearance [19, 20]. The location of the ear is all the more sensitive as the appearance of the ear in an 18 years old girl is part of the beauty criteria: the ear is an even and symmetrical organ, it is located laterally on each side of the skull in the temporal bone [15]. It plays an important role in the harmony of the face and consequently in the aesthetics of the face, especially in young girls. It is usually accessorized and highlighted by the female gender. Adults with congenital naevi should also have the opportunity to receive advice, and possibly treatment, from experienced dermatologists and surgeons in other countries as was the case for our patient. In the past, the fear of a transformation into melanoma guided this choice. However, for some years now, we have known that this risk is less and is no longer the primary reason for the indication for surgery, thus putting the "aesthetic" problem in the foreground [6].

Apart from the surgeon's monitoring of the quality of the scar in the first year, there is no particular monitoring for naevi operated and completely excised. For naevi left in place, there are no validated recommendations. It may be useful to suggest self-monitoring to the adult patient and to ask for a consultation if the lesion changes. In the older child or adult, systematic monitoring does not appear to be useful, although the risk of symptomatic involvement persists [1]. Our patient was lost to follow-up after one month of treatment, but self-monitoring was recommended.

4. Conclusion

The middle congenital naevus is a benign tumor linked to a genetic variation of low incidence, all the more rare as it is located at the level of the ear having thus a significant psychological impact due to its often unsightly aspect. Its diagnosis is entirely clinical. When it is an isolated nodule, as in our study, total surgical removal is a good alternative with simple after-effects and an almost non-existent risk of malignant transformation.

References

[1] Taieb A., Vergnes P. Naevus congénitaux Points saillants. 2005 https://www.chu-bordeaux.fr.

[2] Price HN. Congenital melanocytic nevi: update in genetics and management. Curr Opin Pediatr. 2016; 28 (4): 476-482.

[3] Aubert H., Scard C., Barbarot S. Naevus congénitaux, Rev Prat Med Gen 2020; 34 (1042); 411-415.

[4] Alexandra A. Naevus géant congénital. Santé sur le net. https://www.sante-sur-le-net.com/sante-enfant/naevus-geant-congenital. 11 September 2018.

[5] Jung SY, Kim MG, Boo SH, et al. Analyse clinique des masses bénignes auriculaires. Coréen J Audiol. 2012; 16 (1): 10-13.

[6] Protocole National de Diagnostic et de Soins (PNDS) Naevus congénital, FIMARAD NC / Septembre 2021. https://www.has-sante.fr

[7] Navarro-Fernandez IN, Mahabal GD. Congenital Nevus. In: StatPearls. StatPearls Publishing, Treasure Island (FL); 2021.

[8] Koot HM, de Waard-van der Spek F, Peer CD, Mulder PG, Oranje AP. Psychosocial sequelae in 29 children with giant congenital melanocytic naevi. Clin Exp Dermatol. 2000; 25: 589-593.

[9] Kinsler VA, O'Hare P, Bulstrode N, Calonje JE, Chong WK, Hargrave D, et al. Melanoma in congenital melanocytic naevi. Br J Dermatol. mai 2017; 176 (5): 1131-1143.

[10] Krengel S, Scope A, Dusza SW, Vonthein R, Marghoob AA. New recommendations for the categorization of cutaneous features of congenital melanocytic nevi. J Am Acad Dermatol. mars 2013; 68 (3): 441-451.

[11] Salgado CM, Basu D, Nikiforova M, Bauer BS, Johnson D, Rundell V, et al. BRAF mutations are also associated with neurocutaneous melanocytosis and large/giant congenital melanocytic nevi. Pediatr Dev Pathol Off J Soc Pediatr Pathol. févr 2015; 18 (1): 1-9.

[12] Polubothu S, McGuire N, Al-Olabi L, Baird W, Bulstrode N, Chalker J, et al. Does the gene matter? Genotype-phenotype and genotype-outcome associations in congenital melanocytic naevi. Br J Dermatol. 2020; 182 (2): 434-443.

[13] Merkel EA, Mohan LS, Shi K, Panah E, Zhang B, Gerami P. Paediatric melanoma: clinical update, genetic basis, and advances in diagnosis. Lancet Child Adolesc Health. 1 Sept. 2019; 3 (9): 646-654.

[14] Captier G. Les nævus melanocytaires de la face chez l'enfant: quoi de neuf? Rev Stomatol Chir Maxillofac Chir Orale 2015; 116: 187-199.

[15] Houari S., Aderdour L., Raji A. Anatomie tridimensionnelle de l'oreille [Thèse de médecine], 2013. 4p.

[16] Chong SJ, Jeong E, Park HJ, Lee JY, Cho BK. Treatment of congenital nevomelanocytic nevi with the CO_2 and Q-switched alexandrite lasers. Dermatol Surg. 2005; 31: 518-521.

[17] Kono T, Ercocen AR, Nozaki M. Treatment of congenital melanocytic nevi using the combined (normal-mode plus Q-switched) ruby laser in Asians: clinical response in relation to histological type. Ann Plast Surg. 2005; 54: 494-501.

[18] Noordzij MJ, van den Broecke DG, Alting MC, Kon M. Ruby laser treatment of congenital melanocytic nevi: a review of the literature and report of our own experience. Plast Reconstr Surg. 2004; 114: 660-667.

[19] Vries H. L' Analyse Comportementale, La Conceptualisation des Cas en Thérapie Comportementale et Cognitive. Rev Société Algér Psychiatr. déc 2010; (10): 5-10.

[20] Viana, Ana Carolina L. et al. "Naevus mélanocytaire congénital géant." Anais brasileiros de dermatologie vol. 88, 6 (2013): 863-878.