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Contribution of Medical Imaging in the Diagnosis of Epidermoid Cyst of the Anterior Fontanel in an Child: A Case at the "Mother-Child" University Hospital Center (Chu) Bamako Luxembourg

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Abstract Case Report

This study aims to identify the epidemiological, clinical, radiological, surgical and histological characteristics of epidermoid cysts. The epidermoid cyst of the anterior fontanel is a very rare congenital lesion, less frequent than the dermoid cyst. We report a case of a four-month-old infant who consulted for a mass located at the level of the anterior fontanel. This lesion presented at birth as a small, soft mass covered with normal-looking scalp that gradually enlarged. Computed tomography showed the characteristics of a slightly heterogeneous cystic lesion limited by the superior sagittal sinus below, without intracranial extension. The surgical intervention allowed its ablation in one piece without adhesion. Histology confirmed the diagnosis. The evolution was satisfactory.

Keywords: Infant, Epidermoid Cyst, Anterior Fontanel, Imaging, Surgery, Histology.

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INTRODUCTION

Congenital epidermoid cysts are rare, slowgrowing benign tumors. They result from the aberrant inclusion of ectodermal elements during the closing of the neural tube, during the first weeks of embryonic development. They represent approximately 0.5% of all cystic inclusions. The first cases were described in black African children, but it is increasingly accepted that this expanding lesion is found in all races [1]. Its location on the anterior fontanel is very rare and unusual and even rarer than that of the dermoid cyst. It can be located anywhere on the body with preferential involvement, for the basilar sites cerebellopontine angle [1] . It can also sit preferentially at the level of the ovaries and testicles. Epidermoid and dermoid cysts are benign lesions of the malformative type, together they are referred to as pearlescent tumors. These anterior fontanelle cysts are rare, representing 23% of all scalp lesions in children. For the notion of frequency according to sex, there is a female predominance with two girls for one boy [2].

Clinically, anterior fontanelle epidermoid cyst is a soft, nonpulsatile, painless, poorly mobile or fixed

tumor. It can be observed at birth, covered with healthy scalp, with a tendency to gradually increase in size. It is usually isolated, with no other clinical and radiographic abnormalities [2].

The positive prenatal diagnosis of congenital epidermoid cysts can be made by ultrasound in the fetus as early as the 17th week of gestation [1] . On CT, postnatally, it appears as an iso- or hypo-dense mass with regular contours, without the slightest enhancement after administration of iodinated contrast [3] .

Magnetic resonance imaging shows with great precision the characteristics of the cyst, pushing back the cutaneous covering above, the superior sagittal sinus below, without intracranial extension. It is hypointense in T1, hyperintense in T2, not enhancing after injection of gadolinium. It also allows the analysis of the cyst content in protein, cholesterol and Calcium [3].

On the embryological level, these cysts are benign tumoral formations of slow growth, of congenital origin resulting from the aberrant inclusion of ectodermal element during the closing of the neural

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tube between the 3rd and 5th week of embryonic development. The anterior fontanel is the preferred site for embryonic fusion, which would explain this location. The possibility of intracranial extension has been reported. This possible extension would suggest that the lesion would occur at an earlier stage of intrauterine development [1].

The treatment of the epidermoid cyst is surgical, its purpose is diagnostic, therapeutic and aesthetic. Complete excision avoids the occurrence of recurrence. After incomplete excision, recurrence is inevitable after a very prolonged delay, evaluated at the development time of the initial tumor (YASARGIL, 1989) [3].

In the review of the literature and to our knowledge very few cases of epidermoid cyst of the anterior fontanel are reported, which is why we made this observation of a case of epidermoid cyst of the anterior fontanel in a four-month-old infant with aims to show the main lines of imaging in its diagnosis.

OBSERVATION

We report the case of a four-month-old female infant from a pregnancy carried to term without prenatal consultation from a village in the Kayes region of Mali. The delivery took place in a health center in the said area, the date of delivery is unknown by the parents, the birth weight was normal at birth according to the mother of the child.

She is the sixth child of a family of six living children with no particular pathological history who consulted at the age of 4 months for a mass of the cranial vault next to the anterior fontanel.

This lesion appeared at birth and gradually increased in size.

He was received in the radiology and medical imaging department at the CHU "Mère-enfant" Le Luxembourg in Bamako, on January 23, 2023 for a cranio-encephalic CT scan for a mass in the anterior fontanel.

On receipt, it was a four-month-old female infant with no medical and surgical history, the result of an unmonitored full-term pregnancy.

On clinical examination, we find a median frontal mass next to the anterior fontanelle that is soft, not very mobile, non-compressible, with no perception of intracranial pulsations and with regular contours.

Cardiovascular, pulmonary, neurological and musculoskeletal examinations were unremarkable.

Cranio -encephalic computed tomography was performed without and with injection of iodinated contrast product on January 23, 2023, which revealed a subcutaneous mass located next to the anterior fontanelle, of finely heterogeneous fluid density not enhanced after injection of contrast medium, measuring 27x 22 mm (**Figure 1: a, b, c and d**).

The result of the cranio-encephalic computed tomography the diagnosis of epidermoid cyst of the anterior fontanelle was evoked without any sign of intracranial involvement.

Following this result of the cranioencephalic computed tomography, a neurosurgery consultation was requested. A preoperative assessment was performed.

The preoperative biological assessment was without abnormality.

Surgical treatment was performed on February 9, 2023 in the operating theater of the mother-child university hospital "Luxembourg" in Bamako, which consisted of total removal of the tumor by a median transverse incision centered on the lesion under general anesthesia plus orotracheal intubation. Asepsis was performed plus a scalp incision perpendicular to the midline centered on the lesion (**Figure 2: a, b, c**).

Then we carried out a meticulous dissection of the mass in its entirety (in one piece) according to a cleavage plane without incident or accident, then to its complete excision. Finally, complete and satisfactory hemostasis was performed and then plan-by-plane closure plus dressing (**Figure 3**).

The postoperative course was simple (**Figure 4**). Macroscopic study showed a yellowish piece of firm consistency of pearly white coloration, containing hair with a fibrous shell (**Figure 5**).

On histological study, the cystic wall was lined with well-differentiated skin tissue, with keratin lamella; without cytonuclear atypia . The dermis was inflamed and made of lymphoplasmocytes . The epidermis was atrophied.

The anatomopathological study confirmed the diagnosis of epidermoid cyst without signs of malignancy.

Postoperative follow-up at 3 months with clinical examination, transfontannel ultrasound and cranioencephalic CT scan was unremarkable.

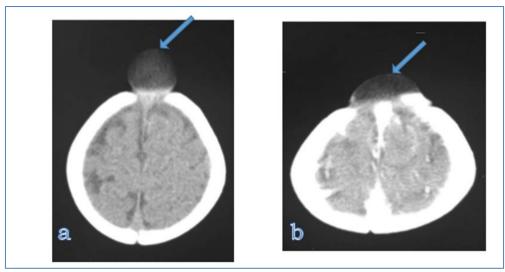


Fig.1 a et b: Cranio-encephalic CT in axial section without and with iv p PDC highlighting a cystic lesion

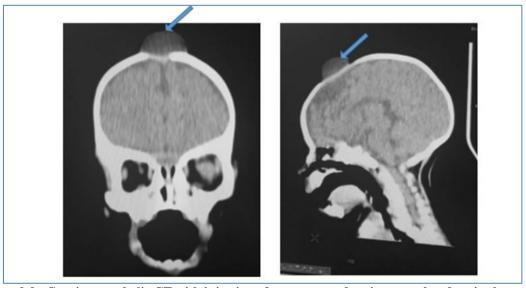


Figure 1 c and d.: Cranio-encephalic CT with injection of contrast product in coronal and sagittal reconstruction showing a cystic lesion of the anterior fontanel (blue arrows)

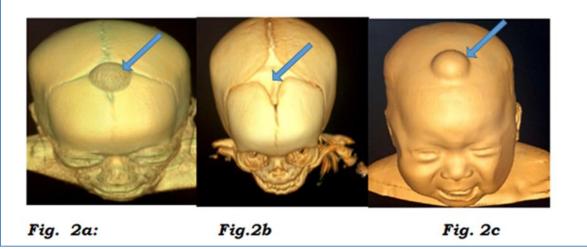


Figure 2: a, b, c: Cranial CT in coronal 3D reconstruction (anterior view) showing the anterior fontanelle, the mass and the sutures (sagittal, coronal, and metopic). See blue arrows.

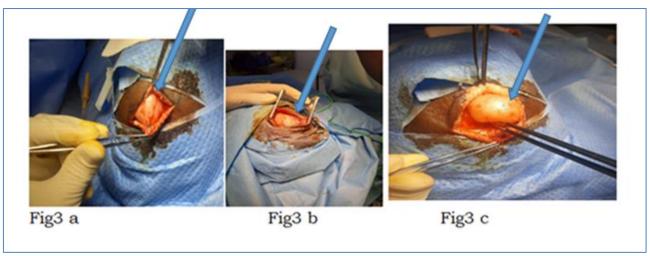


Figure 3: a,b,c: median transverse linear skin incision centered on the lesion (see arrows).

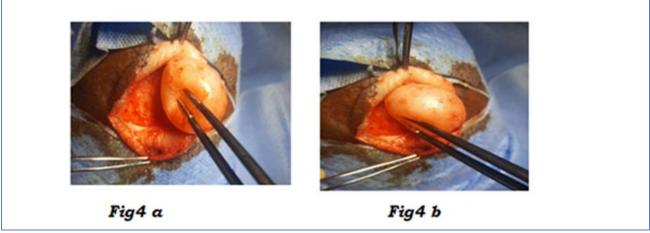


Figure 4: a and b: Dissection of well-encapsulated non-adherent soft consistency cyst

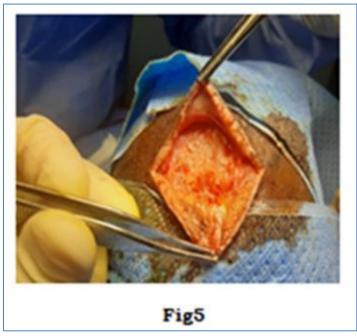


Figure 5: image showing the extra cranial lodge of the mass at the level of the anterior fontanel without intracranial extension.



Figure 6: surgical piece containing hair with a fibrous shell

DISCUSSION

The congenital epidermoid cyst is a rare lesion, presenting a clear predilection for the basilar sites and the pontine cerebellar angle. Its location on the anterior fontanel is very rare and unusual and even rarer than that of the dermoid cyst [1] . The first cases were described in black African children, but it is increasingly accepted that this expanding lesion is found in all races [2] .

*For the Notion of Frequency according To Gender, in 2021, a study was conducted by Agaly *et al.*, on the congenital epidermoid cyst of the anterior fontanel about a case in a seven-month-old female infant with no medical history. -surgical which was published in the Journal of Medicine and Biomedical Science. These have not found a predominance of one sex over the other in the literature [23]. The concept of frequency according to sex is only mentioned by a few authors in the literature.

According to Adeloye, this lesion occurs most often in women with a 2/1 ratio.

Peter does not make the same observation on a series of 35 children, he has 20 boys and 5 girls, NDARAW NDOYE *et al.*, had a high proportion of female cases (4/1) in a study conducted on the dermoid cyst congenital extra cranial about 5 cases in 2002. AM YAHIOUI, *et al.*, have a similar study, there was a female predominance with two girls for one boy [2]. In our study we revealed a female case which confirms that of Peter; NDARAW NDOYE *et al.*, Adeloye and AM YAHIOUI, *et al.*,

*On the Embryological Level , these cysts are benign tumoral formations of slow growth, of congenital origin resulting from the aberrant inclusion of ectodermal element during the closure of the neural tube between the 3rd and 5th week of embryonic development [1]. The anterior fontanel is the preferred site for embryonic fusion, which would explain this location. The possibility of intracranial extension has been reported. This possible extension would suggest that the lesion would occur at an earlier stage of intrauterine development [1]. In our case the cyst was located at the level of the anterior fontanelle since birth and which gradually increased.

*Clinically, the epidermoid cyst of the anterior fontanel is a soft, non-pulsating, painless tumor that is not very mobile or fixed. It can be observed at birth, covered with healthy scalp, with a tendency to gradually increase in size. It is usually isolated, with no other clinical and radiographic abnormalities [1]. The positive prenatal diagnosis of congenital epidermoid cysts can be made by ultrasound in the fetus as early as the 17th week of gestation [2]. In our case, the prenatal consultations were not adequate.

*On CT scan, it appears as an iso or hypodense mass with irregular contours, without any enhancement after administration of iodinated contrast product [3].

*Magnetic resonance imaging shows the characteristics of the cyst with great precision, pushing the cutaneous covering upwards, the superior sagittal sinus downwards, without intracranial extension. It is hypointense in T1, hyperintense in T2, not enhancing after injection of gadolinium. It also allows the analysis

of the cyst content in protein, cholesterol and Calcium [3].

*Differential diagnosis includes cephalohematoma, lipoma, pericranial sinus, angiomatous malformation, bone cyst and abscess. These pathologies are easily detectable from the epidermoid cyst [3] .

The treatment of the epidermoid cyst is surgical, its purpose is diagnostic and aesthetic. Complete excision avoids the occurrence of recurrence [2]. Cyst puncture before surgery is not recommended because there is a risk of infection and reconstitution of the cyst [1].

*Technically, a linear incision centered on the tumor is performed. The detachment of the subcutaneous tissue from the tumor capsule is often easy. If there is a dural attachment, ligation is performed followed by coagulation. The only difficulty reported by some authors is adhesion to the dura mater covering the superior longitudinal sinus, given the risk of hemorrhage [23].

*Histologically , the epidermoid cyst consists of a stratified, keratinized squamous epithelium with a lumen filled with the breakdown products of desquamated epithelial cells in the form of an amorphous material [2] . The absence of dermal elements (hair follicles, sebaceous gland, sweat and sometimes even fat follicles) differentiates it from the dermoid cyst. The prognosis is good without recurrence if the cyst is resected en bloc. Its malignant transformation into squamous cell carcinoma has been described but not in this location [1] .

DECLARATION OF CONFLICT OF INTEREST: The authors declare that they have no conflict of interest.

BIBLIOGRAPHIC REFERENCES

- Agaly, H., Djerma, I., Tounkara, M., kanikomo, D., Sogoba, B., Sogoba, Y., et al. (2021). Congenital epidermoid cyst of the anterior fontanelle: apropos of a case. *Health Sci Dis.*22 (4), 3
- 2. Yahiaoui, A., Abzouzi, B., Bekkouche, M., Ioualalen, N., Ahmed, R. b. (2006). The Epidermoid Cyst of the Anterior Fontanelle. *J Neurochir*. 27(4), 3.
- Dermoid and epidermoid cysts Neurosurgery Campus https://neuro-dev.unilim.fr > IMG > article_a358 - Google search [Internet]. 2009 [cited 2023 Mar 6]. Available at: https://www.google.com/
- Anterior fontanel anatomy Google Search [Internet].
 [cited 2023 Mar 6]. Available at: https://www.google.com/
- Hayman, A. L., Shukla, V., Ly, C., & Taber, K. H. (2003). Clinical and imaging anatomy of the scalp. *Journal of computer assisted tomography*, 27(3), 454-459.

- 6. Scalp anatomy pdf Google Search [Internet]. [cited 2023 Apr 2]. Available at: https://www.google.com/
- 7. Sharman, A. M., Kirmi, O., & Anslow, P. (2009, December). Imaging of the skin, subcutis, and galea aponeurotica. In *Seminars in Ultrasound, CT and MRI* (Vol. 30, No. 6, pp. 452-464). WB Saunders.
- UFO themes. 4. Bones, muscles, joints | Medicine Key [Internet]. 2017 [cited 2023 Mar 27]. Available at: https://clemedicine.com/4-les-os-les-muscles-lesarticulations/
- 9. Savoldi, F., Tsoi, J. K., Paganelli, C., & Matinlinna, J. P. (2018). The biomechanical properties of human craniofacial sutures and relevant variables in sutural distraction osteogenesis: a critical review. *Tissue Engineering Part B: Reviews*, 24(1), 25-36.
- Palacino, P. V. D., Munoz, E. E. R., & Zamora, M. P. G. (2022). Fontanelles and Sutures: From Theory to Practice, A Literature Review. Arch Clin Trial & Case Rep, 1(2), 30-36.
- Lipsett, B. J., Reddy, V., & Steanson, K. (2019). Anatomy, head and neck, fontanelles.
- 12. Popich, G. A., & Smith, D. W. (1972). Fontanels: range of normal size. *The Journal of pediatrics*, 80(5), 749-752.
- D'Antoni, A. V., Donaldson, O. I., Schmidt, C., Macchi, V., De Caro, R., Oskouian, R. J., ... & Shane Tubbs, R. (2017). A comprehensive review of the anterior fontanelle: embryology, anatomy, and clinical considerations. *Child's Nervous System*, 33, 909-914.
- 14. Mackenzie, A., Barnes, G., & Shann, F. (1989). Clinical signs of dehydration in children. *The Lancet*, *334*(8663), 605-607.
- 15. Rothman, S. M., & Lee, B. C. (1998). What bulges under a bulging fontanel?. *Archives of pediatrics & adolescent medicine*, 152(1), 100-101.
- Bustamante, J. L., Miquelini, L. A., D'Agustini, M., & Fontana, A. M. (2010). Applied anatomy of the fontanelles. *Neurocirugia (Asturias, Spain)*, 21(3), 253-259.
- 17. Kiesler, J., & Ricer, R. (2003). The abnormal fontanel. *American family physician*, 67(12), 2547-2552.
- 18. Faix, R. G. (1982). Fontanelle size in black and white term newborn infants. *The Journal of Pediatrics*, 100(2), 304-306.
- AT DLGDES. Elementary basics of craniotomies Daniel Le Gars - Google search [Internet]. 2021. Available at: https://www.google.com/
- Cisse, I., Camara, M., Kone, C. A., Camara, T., Traore, D., Mariko, M., ... & Sidibe, S. (2022). Retroperitoneal Teratoma in Infants: About a Case in the Medical Imaging Department of the Mother and Child Hospital Center "Luxembourg". Open Journal of Medical Imaging, 12(02), 73-81.
- Adenigba, P. T., Lawal, T. A., Elemile, P. O., Onakpoma, F., & Adekanmi, A. J. (2019). The value of ultrasonography in the diagnosis of a rare congenital dermoid cyst of the anterior fontanelle in an infant. *Journal of the West African College of* Surgeons, 9(4), 21.
- ZULCHKL Brain tumors: their biology and pathology...
 Google Scholar [Internet]. [cited 2023 Mar 12]. Available at: https://scholar.google.com/
- Tan, E. C., & Takagi, T. (1993). Congenital inclusion cysts over the anterior fontanel in Japanese children: a study of five cases. *Child's Nervous System*, 9, 81-83.