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Case Report

Epidermoid and Dermoid Cyst of the Anterior Fontanelle About 2 Cases

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Abstract: Introduction: Epidermoid cysts and dermoid cysts are uncommon benign tumors of congenital origin who result from the aberrant inclusion of ectodermal elements during closure of the neural tube between the 3rd and 5th week of embryonic development. The location of these cysts at the anterior fontanelle level is very rarely reported. These cysts evolve by gradually increasing in volume, causing aesthetic problems. Our aim is to report a case of epidermoid cyst (EC) and a case of dermoid cyst (DC) diagnosed and operated in our department. Observations: We report one case of epidermoid cyst and one case of dermoid cyst in female infants aged 10 and 4 months who presented with a subcutaneous medial fronto-parietal mass of the anterior fontanelle. These mobile, painless masses with a wide base of implantation measured 6 x 5.5 cm and 5 x 3.5 cm. After brain scan exploration, the total excision of the cysts was carried out. The postoperative progress was good without recurrence. The diagnoses of epidermoid and dermoid cysts were confirmed by histology. Conclusion: Anterior fontanelle epidermoid and dermoid cysts are congenital benign tumors who is rarely reported. They are often diagnosed in the first months of life. One piece excision gives excellent results without recurrence.

Keywords: Epidermoid Cyst, Dermoid Cyst, Anterior Fontanelle, Excision

1. Introduction

Epidermoid cysts (EC), also called beaded tumors, and dermoid cysts (DC), are uncommon benign tumors of congenital origin. Their anterior fontanelle (AF) location is rare. They result from the aberrant inclusion of ectodermal elements during closure of the neural tube between the 3rd and 5th week of embryonic development [1]. The spontaneous evolution of the AF cyst is towards a progressive increase in volume leading to an aesthetic problem motivating the consultation [2]. Our aim is to report a case of epidermoid cyst (EC) and a case of dermoid cyst (DC) diagnosed and operated in our department.

2. Observations

2.1. Case 01

A 10 month old female infant with no particular past medical history, who consulted for a congenital frontal swelling gradually increasing in volume. The physical examination found good general and neurological condition with no delay in psychomotor development. At the cephalic extremity, a subcutaneous medial fronto-parietal mass measuring 6 x 5.5 cm, mobile, painless, renitent, no pulsatile was found. It is developed on the anterior fontanelle with a wide implantation base and normal skin (Cf Figure 1). The remainder of the examination did not find any malformation

or other associated abnormalities. The Brain computed tomography (CT) showed a hypodense subcutaneous cystic lesion not taking up the contrast product developed on incompletely closed AF without endocranial extension. There was no malformation associated on CT. During surgical excision, we dissected in one piece an encapsulated, translucent, no hemorrhagic lesion that was easily detached from the cranial vault and the AF (Cf Figure 2). We sent the cystic lesion for anatomo-pathological study. The histological study concluded that there was KE, finding cystic wall fragments made of keratinized squamous tissue with keratin lamellae. The postoperative evolution was good without recurrence.

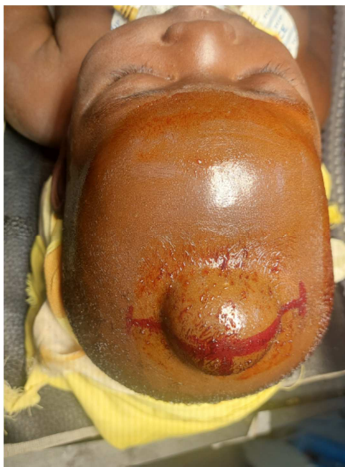

Figure 1. *Median frontal subcutaneous mass of AF.*

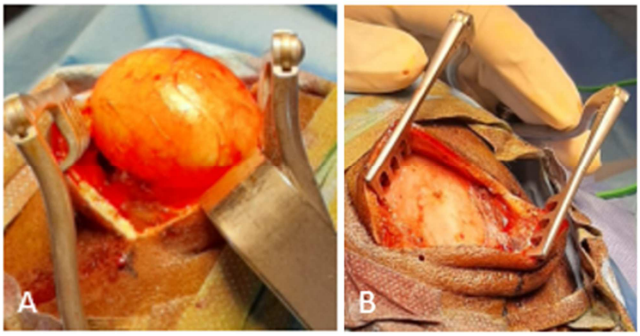

Figure 2. *Operative images, A: Dissection respecting the capsule of the lesion. B: One-piece excision.*

2.2. CASE 02

A 4 month old female infant with no particular past medical history, who consulted for frontal swelling evolving since birth. The physical examination found good general and neurological condition with no delay in psychomotor development. At the cephalic extremity, a subcutaneous medial fronto-parietal mass measuring 5 x 3, 5 cm, mobile, painless, no pulsatile was found. It is developed on the anterior fontanelle with a wide implantation base and normal skin (Cf Figure 3). The remainder of the examination did not find any malformation or other associated abnormalities. The Brain computed tomography (CT) showed a hypodense subcutaneous cystic lesion not taking up the contrast product developed on open AF without endocranial extension (Cf Figure 4). There was no malformation associated on CT. We performed surgical excision in one piece. We dissected an encapsulated, no hemorrhagic lesion that was easily detached from the cranial vault and the AF. After excision, we incised the lesion which had a fairly thick capsule containing whitish deposits and hairs (Cf Figure 5). We sent the lesion for anatomo-pathological study. The histological study concluded that it was DC, finding keratinized squamous tissue with keratin lamellae, hair follicles and hairs. The postoperative evolution was good without recurrence.

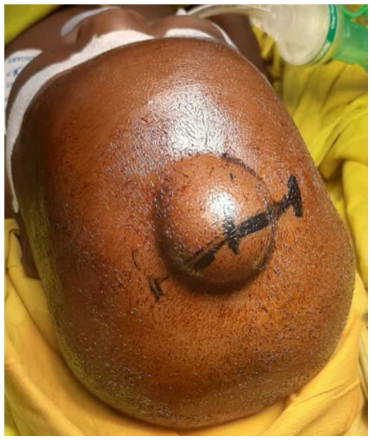

Figure 3. *Medial subcutaneous fronto-parietal mass of AF suggestive of an AF cyst.*

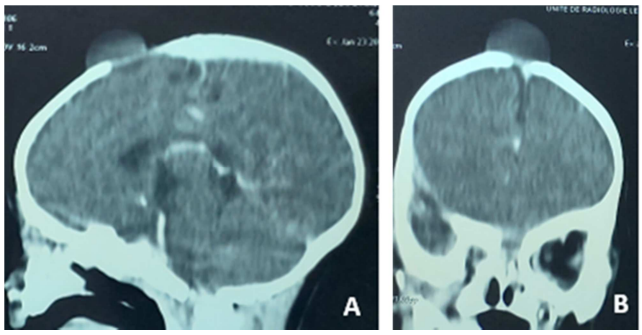

Figure 4. *Cranio-cerebral CT in sagittal (A) and coronal (B) reconstruction showing an isodense cystic subcutaneous lesion of the Anterior Fontanelle without endocranial extension.*

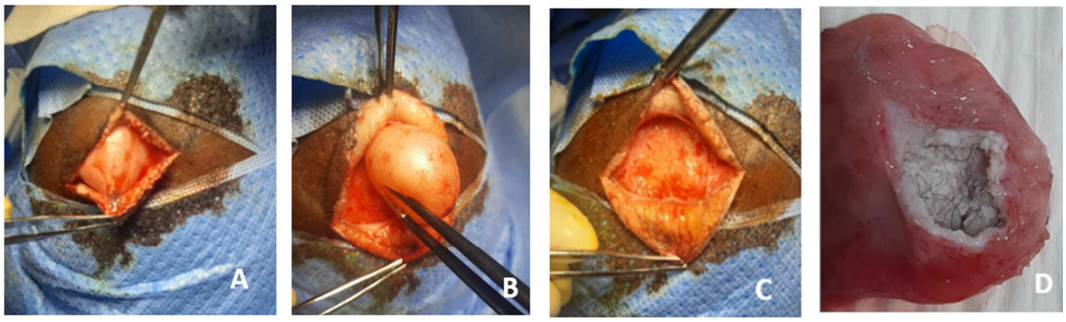

Figure 5. *Operative images, A and B: Incision and dissection respecting the capsule of the lesion. C: One-piece excision, D: Opening of the hull exposing viscous, whitish pasty contents and hair.*

3. Discussion

EC and DC are benign tumors of malformative congenital origin resulting from the aberrant inclusion of ectodermal elements during closure of the neural tube between the 3rd and 5th week of embryonic development [1]. EC and DC remain rare lesions at the anterior fontanelle. The EC also called primary cholesteatoma or cruveilhier's beaded tumor is more often found at the cerebello-pontine angle and the sellar region. It represents less than 2% of all primary intracranial tumors. The dermoid cyst represents 10% of all extracranial mass [2]. This extracranial location is uncommon. Indeed, EC and DC of the anterior fontanelle are rare and represent between 0.1 and 0.2% of all skull tumors [3, 4]. In the extracranial location, the location of these cysts at the AF level is very rarely reported. A review of the literature on AF cysts reveals that almost all authors report only one case [1, 4-13] or a limited series of cases [2, 3, 14]. Ndoye and al [2] in his study on 5 cases reported the location of the AF in 3 cases and the location front of the fontanelle on the midline of the frontal bone in 2 cases. All of our two cases was exclusively at the FA level. The female predominance in our series is confirmed by other authors [2, 3, 14, 15] who reached the same observation in slightly more extensive series. AF cysts are most often discovered at birth but diagnosed in the months following birth when the mass gradually increases in size. However, prenatal diagnosis is possible at obstetric ultrasound from the 17th week of gestation [1]. Our 2 cases were observed after birth between 4 and 10 months, like several authors [1, 2, 5-7]. Howewer Mentri and al [14], Agrawal and al [4] and Adelola and al [3] reported cases diagnosed in patients aged 4 years, 14 years and 18 years respectively.

Clinically, AF cysts appear in the form of a frontal or medial fronto-parietal subcutaneous mass, mobile with normal skin and a wide base implantation at the AF. The pulsatile character disappears with digital compression mentioned by some authors [2, 3, 7] was not observed in our 2 cases. The negativity of the translumination test is not specific to dermoid cyst [10]. The large diameters of the lesions in our 2 cases (5 and 6 cm) are greater than those of several authors [2, 6, 8] which are in proportions varying between 2 and 3.5 cm. However, the most largest lesion (15 x 10 x 6 cm) was observed by Adelola and al [3] in an 18-year-old patient. These AF cysts are usually isolated, with no other clinical abnormalities or malformations observed. The clinical appearance of AF cysts may suggest other mass such as: cephalhematoma, meningoencephalocele, lipoma, sebaceous cyst, bone cyst, cutaneous angioma, vascular malformation, externalized intracranial tumor. If the clinical examination can differentiate between the AF cyst and some of these lesions mentioned, exploration by brain CT or magnetic resonance imaging (MRI) is necessary to clarify limits, contents, possible intracranial extension of the lesion, and make the differential diagnosis. According to Yahiaoui and al [1] there is no specific image of EC and DC on CT and MRI. Some MRI sequences (FLAIR, C. ISS, DWI sequences and diffusion weighted imagery) allow us to differentiate the epidermoid cyst from other cystic lesions [1]. Our 2 cases were explored only with CT as in Agrawal and al [4], Ponce – Ayala and al [8] and Agaly and al [5]. We believe that CT is sufficient for diagnosis. Other authors [1, 7] had to perform angio-MRI to assess the relationship between cyst and superior sagittal sinus. We found no reported cases of anterior fontanelle cyst with intracranial extension.

Once the diagnosis of an AF cyst has been established, surgical treatment is unanimous. Total excision of the cyst in one piece is the rule in order to correct the aesthetic damage and avoid recurrence. It is easily done almost without bleeding by a dissection respecting the planes and the capsule of the cyst; from the periphery to the midline, carefully peeling off the middle portion of the FA last.

Only histological examination allows the diagnosis of EC or DC by observing heterogeneous cystic lesions that may contain keratin, cholesterol, fat, hair, sebaceous and sweat secretions [2]. According to Yahiaoui and al [1] the presence of dermal elements (hair follicles, sebaceous and sweat glands) specific to DC makes it possible to differentiate it from EC. Indeed in our case of DC, the histological study found a keratinized squamous epithelium with keratin lamellae, hair follicles and hairs.

The postoperative evolution of EC and DC is good without recurrence when the excision is total in one piece.

4. Conclusion

AF cysts (EC and DC) are congenital benign tumors whose localization in the AF is rare. They are often diagnosed in the

first months of life by the clinic which is supported by CT. Prenatal diagnosis is possible with obstetric ultrasound and allows for early treatment before the lesion is large with a more significant aesthetic problem. The treatment is surgical and is based on careful dissection. One piece excision gives excellent results without recurrence.

Conflicts of Interest

The authors declare no conflicts of interest**.**

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Biography

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