ORIGINAL PAPER

Calvarial dermoids and epidermoids in infants and children: sonographic spectrum and follow-up

Thomas Riebel · Siegfried David · Ulrich W. Thomale

Abstract

Objective Dermoids and epidermoids are defined as ectodermal inclusion cysts. The aim of this study was to evaluate the spontaneous natural behavior and the ultrasonographic appearance of calvarial dermoids and epidermoids.

Materials and methods The ultrasonographic image datasets of 100 consecutive children up to 4 years of age (52 females, 48 males; age range at first examination 1 week to 40 months, mean age 8.3 ± 6.9 months) presenting with a firm palpable calvarial mass (103 lesions) were studied retrospectively. All ultrasound (US) examinations were performed using a 7- to 10-MHz linear transducer including B-mode and color Doppler sonography. US follow-up studies (up to 47 months) could be achieved in 30 patients with 33 lesions.

Results At first presentation, all 103 lesions demonstrated very similar US features: a round or oval configuration (diameter 3–18 mm), hypoechogenic, and homogeneous internal structures with a marked hyperechogenic superficial capsule, which were localized adjacent to or expanded into the osseous external calvarial table. No conspicuous flow signs on color Doppler were seen. In 33 lesions with US follow-up investigations, 49% showed variable signs of regression: reduction of size, increase of internal echogenicity, and decrease of demarcation. Eight lesions (24%) remained unchanged. A slight progression up to a maximum diameter of 17 mm but without any increase in osseous destruction was observed in the remainder (27%). There was no lesion with a complete destruction of the underlying bone and no intracranial extension in any of the cases.

Conclusions Calvarial dermoids and epidermoids in infants and children show a benign natural behavior with spontaneous regression in a large number of cases. On US, they demonstrate uniform pathognomonic features enabling the correct diagnosis in any of those lesions. Thus, additional, mainly radiation burdening and sometimes misleading imaging techniques should be restricted. Surgical treatment protocols should be handled conservatively and lesions should be primarily followed-up clinically and by US.

Keywords Dermoids/epidermoids ·
Ultrasound/ultrasonography/sonography · Calvarium ·
Infants/children

Thomas Riebel and Siegfried David contributed equally to this study.

T. Riebel (✉)
Department of Pediatric Radiology, Charité Centrum 6, CVK,
Charité-Universitätsmedizin Berlin,
Augustenburger Platz 1,
13353 Berlin, Germany
e-mail: thomas.riebel@charite.de

S. David
Department of Pediatric Surgery,
Charité-Universitätsmedizin Berlin,
Berlin, Germany

U. W. Thomale
Department of Pediatric Neurosurgery,
Charité-Universitätsmedizin Berlin,
Berlin, Germany

Introduction

Dermoids and epidermoids belong to the family of ectodermal inclusion cysts. They represent early embryonic displacements of epithelial nests in association with the closure of the neural tube and the clefts of the viscerocranium. Such epithelial cysts are found in different localizations of the head and neck [6, 8]. In the skull, they are primarily found in the diploe and in some cases with involvement of adjacent soft-
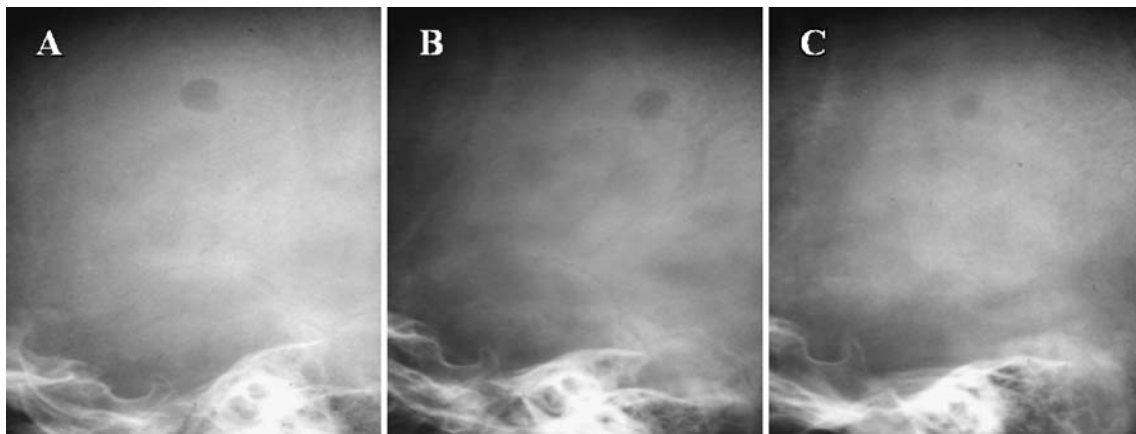
Fig. 1 Radiological follow-up of a suspected spontaneously regressing calvarial epidermoid in a young child. **a** Initial X-ray demonstrating a typical round lytic lesion with a sclerotic rim, **b** controls after 21 months, and **c** 43 months show the lesion with decreased size, loss of sharp demarcation, and increase of internal density

tissue layers of the cranial vault. Rarer are localizations in midline structures of the skull base [4, 5].

The histological differentiation of the cystic lesions is well-described. While epidermoids, according to its name, have an epidermal origin, dermoids may also include hair follicle and sebaceous glands. Thus, epidermoids are of pure ceratoid material, while dermoids also contain fatty material and hairs. Both lesions have, in common, the potential to grow as pseudotumors due to the progressive accumulation of debris.

Among the ectodermal cysts of the skull, these cysts of the calvarium represent a separate group with special characteristics and behavior [3, 4, 8]. They are typically seen in early infancy, while the major part identifies as epidermoids. A growth pattern is rare and, if observed, it happens slowly. In the latter group, a transformation in big intracranial expanding cysts is described incidentally. Any development toward malignancy has never been observed. The ectodermal cysts of the midline mostly represent dermoids and dermal sinuses of the glabella, the anterior fontanelle, and the occiput [1, 2, 9].

Calvarial dermoids and epidermoids are found soon after birth as well-circumscribed soft elevations from the convexity, which stick to the underlying calvarium during palpation. In X-ray-based imaging, a clearly demarked lesion with variable lucency and a small sclerotic bony rim (Fig. 1) can be visualized. As a treatment option, surgical removal is still frequently performed in those lesions [6, 8]. Thereby, the histological diagnosis is verified and the aim of treatment is to avoid potentially developing calvarial osteolysis, intracranial mass lesion, or secondary infections of these cysts [5, 6].

Instead of using radiation-exposing imaging techniques of these lesions, recently, high-resolution ultrasonography techniques are more frequently used. In the present study, we review our experience using ultrasound imaging in terms of diagnostic reliability for dermoids and epidermoids of the calvarium in infants. Because it has been reported incidentally that calvarial dermoids or epidermoids may regress spontaneously [3], this study also follows the natural course of these lesions in cases in which surgical treatment was avoided.

Materials and methods

In the total time frame of 9 years (January 1995 to October 2005), 100 consecutive infants and children up to 4 years of
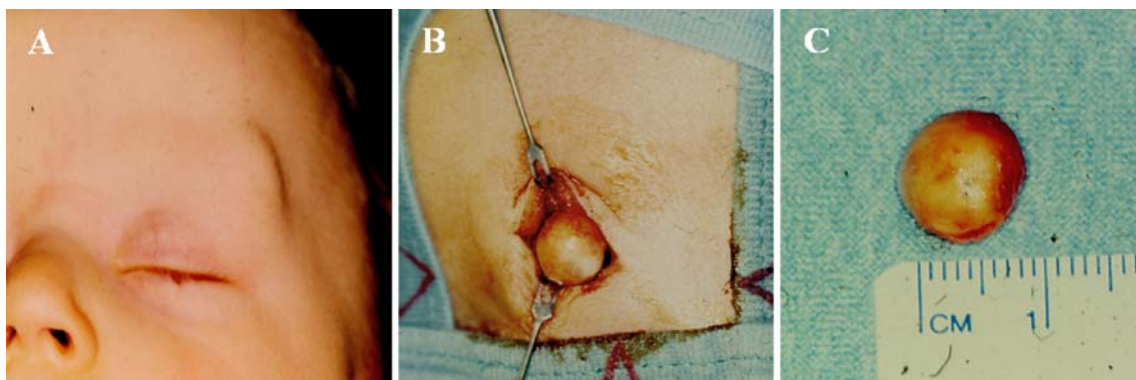

Fig. 2 Histologically proven supraorbital epidermoid in a young child. **a** External view, **b** in situ at surgery, and **c** surgical specimen

Table 1 Initial diameter of lesions and age of patients (months) in the respective group of patients

Groups	All patients (*n*=103)	Without surgery (*n*=93)	With surgery (*n*=10)	Follow-up (*n*=33)
Age (months)	8.3±6.9	8.3±6.8	9.6±11.5	8.5±7.5
Diameter (mm)	8.8±3.3	8.5±3.1	11.9± 3.7*	8.5±3.2
<6 months (mm)	8.3±3.5	7.9±3.4	10.7±3.5	7.8±3.2
6–12 months (mm)	8.6±10.4	8.5±2.6	13±0	8.6±3.0
13–24 months (mm)	10.4±3.1	10.1±3.2	–	14±1.4
25–48 months (mm)	11.9±3.7	9±1	18	8.5±0.7

*p<0.01 vs. nonsurgery

age were reviewed in which a solid, not relocatable lesion under the scalp could be palpated and a dermoid or epidermoid was diagnosed due to ultrasonographic imaging. In 10 children from the early period, lesions were surgically resected (Fig. 2). All of these histopathological investigations verified ultrasonographic diagnostic specificity and was in line with the previous report by Rochels and colleagues [7]. In all other patients (*n*=90) and in the further time course, surgical treatment was avoided primarily. In 100 patients (52girls, 48 boys), a total of 103 calvarial lesions were observed. Three patients showed two lesions, respectively. The mean age at first diagnostic investigation was 8 months (range 1 week to 40 months).

All ultrasonographic investigations were performed by the same radiologist (TR) using a recently manufactured imaging tool (Acuson 128 XP/10, Mountain View, CA, USA) with a high-resolution linear transducer (7 to 10 MHz frequency) and an aquasonic pad. Image acquisition was warranted using B-mode and color coded Doppler technique. No additional imaging techniques were used in all 100 consecutive children.

Follow-up studies using comparable ultrasonography were achieved in 30 patients and 33 lesions, respectively.

The follow-up period was between 4 and 47 months after initial diagnosis. Due to the lack of parental compliance, a unified protocol for follow-up periods in all patients was not applicable. The mean follow-up period in all patients was 15±10.5 months. In four patients, a follow-up period for more than 2 years could be achieved (follow-up time 25–36 months, *n*=2; 36–47 months, *n*=2). All other cases had a lower follow-up period (<12 months, *n*=14; 13–24 months, *n*=12).

The retrospective analysis of hard copy recorded image data sets was based on the descriptive initial sonomorphology of the lesions and the dimensional quantification. Changes over time revealed morphological patterns and diametric quantification compared to initial investigations in the respective follow-up studies.

All results are given as the mean±standard deviation. Statistical analysis was performed to compare the respective groups. A subsequent unpaired or paired *t* test was used combined with the Bonferroni correction of factor 3. The level of significance was α=0.05.

Results

The total number of 103 lesions were located in the temporal (*n*=39), frontal (*n*=30), parietal (*n*=21), and occipital (*n*=13) regions. The initial documented mean diameter of all lesions was 8.8±3.3 mm (range 3 to 18 mm). The age analysis in all lesions revealed that the initial investigation was most often already performed in infants (51%). Dichotomized groups showed an increase of the mean diameter of the lesions parallel with age (<6 months, *n*=51, 8.3±3.5 mm; 6–12 months, *n*=32, 8.6±2.6 mm; 13–24 months, *n*=16, 10.4±3.1 mm; 25–48 months, *n*=4, 11.3±4.6 mm). The mean diameter of the lesions in surgically treated patients was significant larger than in the conservatively followed-up patients (11.9±3.7 vs. 8.5± 3.1 mm; *p*<0.01). If the patients with surgical treatment were excluded (remaining 93 lesions), the mean diameter in the highest age group was slightly smaller compared to the
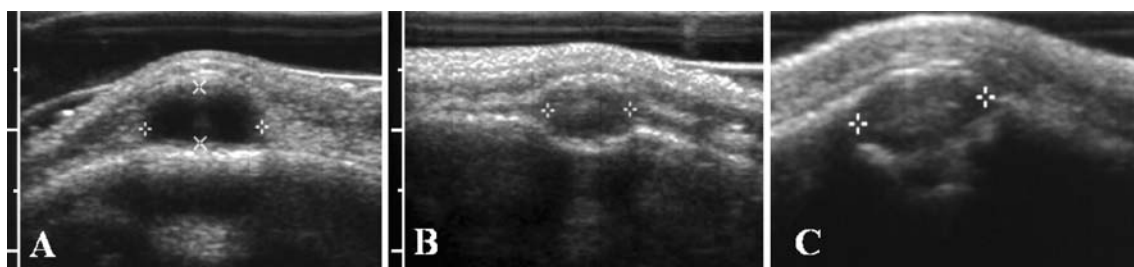

Fig. 3 Typical ultrasonographic pathognomonic features of calvarial dermoids/epidermoids (oval configuration, homogeneous, hypoechogenic internal structure, hyperechogenic surrounding wall, no obvious flow phenomena on color Doppler) with varying impression of the adjacent external skull table. **a** Distinct, **b** moderate, and **c** marked impression

Fig. 4 Complete regression of a suspected temporal dermoid/epidermoid. **a** Initial ultrasonogram of the 4-mm lesion at 6 months of age, **b** control 14 months later showing only minimal residual irregularities of the external skull table
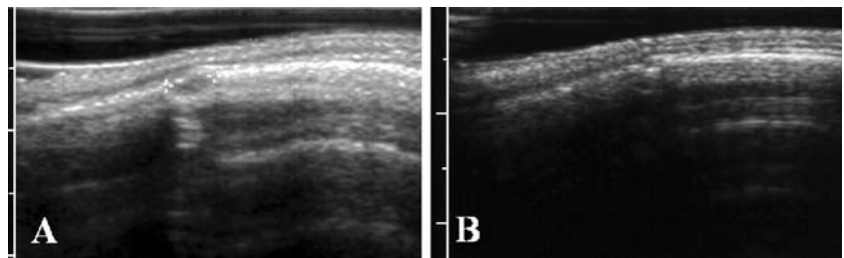

Fig. 5 Marked regression of a temporal dermoid/epidermoid during follow-up. **a** Typical lesion (maximum diameter of 5 mm) at 2 months of age with **b** intensive decrease in size and loss of demarcation 13 months later
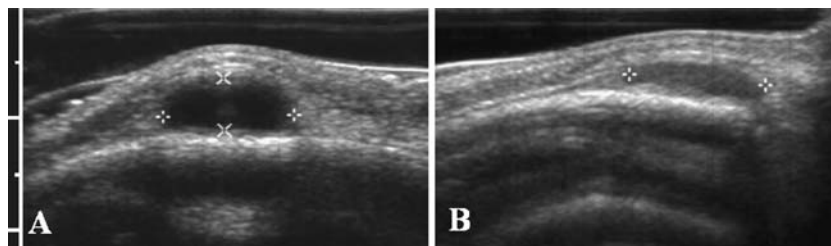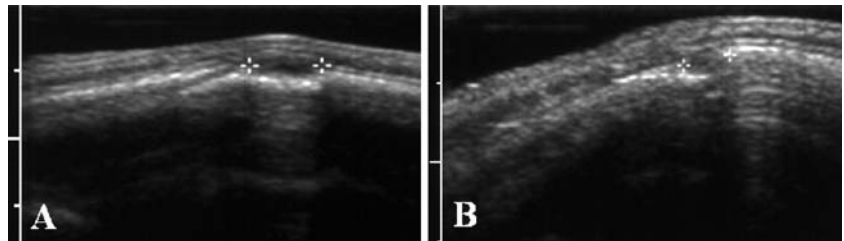

Fig. 6 Ultrasonographic follow-up of a frontal dermoid/epidermoid with moderate spontaneous regression during the time of observation. **a** Characteristic sonomorphology on the first examination at 3 months of age (diameter 4 mm), **b** control 13 months later showing the lesion with a same maximum diameter, but flatter and with increased internal hyperechogenic pattern
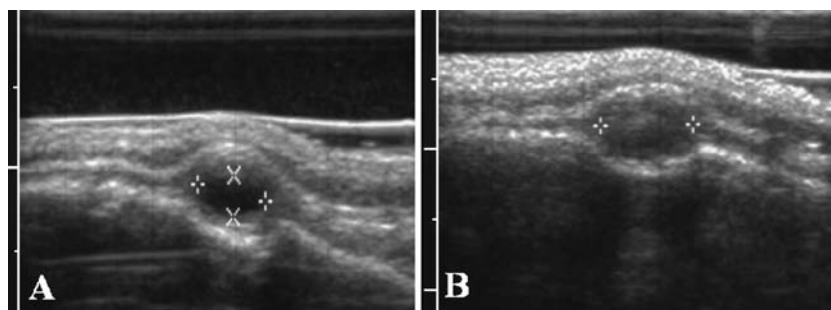

Fig. 7 Occipital dermoid/epidermoid with intermittent slight progression and final complete involution (after 30 months). **a** Initial examination at 3 months of age: maximum diameter 6 mm, **b** 8 months later: identical morphology of the lesion, but some increase in size with up to 9 mm of diameter and slightly more pronounced impression of the external skull table

entire cohort (25–48 months, $n=3$, 9 ± 1 mm). All other groups showed similar results (<6 months, $n=44$, 7.9 ± 3.4 mm; 6–12 months, $n=32$, 8.5 ± 2.6 mm; 13–24 months, $n=14$, 10.1 ± 3.2 mm; Table 1).

The descriptive analysis of the initial ultrasonographic characteristics was defined as follows: a rounded or oval configuration of the lesion with homogenous hypoechogenity of internal structures, a marked capsulated hyperechogenic external circumference, and in color-coded Doppler, no vascular perfusion phenomena inside the lesion and in direct neighboring structures. Two types of infiltration patterns were observed. Either the lesions were only attached to the skull without structural affection or a variable grade of bony infiltration into the external calvarial table could be identified (Fig. 3). None of the lesions showed deeper affection of calvarial structures leading to intracranial expansion.

The follow-up investigations ($n=30$ patients, 33 lesions) were performed mainly in infants (52%). The initially measured mean diameter of the lesions in the respective age group showed a similar pattern compared to the entire patient cohort (<6 months, $n=17$, 7.8 ± 3.2 mm; 6–12 months, $n=12$, 8.6 ± 3 mm; 13–24 months, $n=2$, 14 ± 1.4 mm; 25–48 months, $n=2$, 9 ± 0.7 mm). The differences in diameter compared to the cohort were mainly due to lower patient numbers in both of the older age groups.

The spontaneous behavior in 49% of the lesions showed either complete disappearance (Fig. 4) or significant but incomplete regression (9 ± 3.9 mm vs. 4 ± 4.7 mm, $p<0.01$; Figs. 5 and 6). In the latter cases, we observed not only a diminished size diameter but also a decrease in tissue echogenicity and in border demarcation of the lesions. In one of these cases, a transient progression at the age of 11 months followed by complete involution after 30 months could be visualized (Fig. 7). Eight lesions (24%) were unchanged in all characteristics compared to the initial investigation. Also, in one case of this group, a slight and transient progression is documented. The remaining nine lesions (27%) showed only a progression in size, which never led to any osteolysis in all layers of the calvarium (7.1 ± 2.7 to 11.4 ± 3.5 mm; $p<0.05$). No infectious complications were seen in any of the cases (Table 2).

Looking at size and behavior of all of these lesions, the mean initial size was 9 ± 3.9 mm in the cases with regressive diameter size, while the lesions with progression showed a slightly smaller mean diameter of 7.1 ± 2.7 mm. Comparing the behavior with the follow-up period and the age at the time point of final sonographical imaging in the regressive lesions, the longest time frames were reached with 16.1 ± 7 months of follow-up and 10.2 ± 8.3 to 26.3 ± 9.1 months of age, respectively. In lesions with constant imaging characteristics, investigations were completed at an earlier time point (13.6 ± 13.3 months after first imaging, 5.3 ± 4.9 to $18.9\pm$ 14.4 months of age), while in cases of progressive behavior, the mean follow-up period was 14 ± 14.9 months, while the mean age was 3.4 ± 1.8 to 17.3 ± 13.9 months. The initial age in this group was statistically significantly different compared to the group with regressive diameters ($p<0.05$; Table 2). Considering one unusual case according to our series who was diagnosed at the age of 1 month with a lesion size of 10 mm and showed slow progression over 47 months to 17 mm, which might be excluded from the latter series, then the mean follow-up period is even lower with only 8.5 ± 3.6 months and the age of 3.7 ± 1.6 to 12.2 ± 3.1 months.

Discussion

In clinical diagnostics of local palpable tumors of the calvarium in neonates and infants, sonographical imaging serves as an excellent tool for differential diagnostic clarification by safely identifying benign lesions such as dermoids and epidermoids. Compared to previous published data, which reported experiences in small patient numbers of those lesions to characterize ultrasonographical patterns [7, 9], we are able to analyze these uniform characteristics in a total number of 100 children to actually define pathognomonic criteria. Thus, additional imaging techniques such as magnetic resonance imaging are dispensable and radiation burdening techniques as radiograms and computed tomography must be rated as obsolete. Moreover, the indication for surgical biopsy or even resection, as performed in previous studies [1, 6, 7, 8] and in ten earlier cases of this series, can be consequently avoided.

According to our follow-up observations in conservatively treated patients, the therapeutic considerations for typical calvarial dermoids and epidermoids, in terms of their prognostic behavior, must be addressed in a conser-

Table 2 Age and diameter of follow-up lesion ($n=33$)

	Regression (n=16)	Unchanged (n=8)	Progression (n=9)
Initial age (months)	10.2 ± 8.3	5.2 ± 4.9	3.4 ± 1.8*
Follow-up age (months)	26.3 ± 9.1	18.9 ± 14.4	17.3 ± 13.9
Follow-up time (months)	16.1 ± 7	13.6 ± 13.4	14 ± 14.9
Initial diameter (mm)	9 ± 3.9***	9 ± 2	7.1 ± 2.7**
Follow-up diameter (mm)	4 ± 4.7	9.5 ± 2****	11.4 ± 3.5*****

*$p<0.05$ vs. regression; **$p<0.05$, ***$p<0.01$ vs. follow-up diameter; ****$p<0.05$, *****$p<0.01$ vs. group of regression

vative manner. The exception from this is as mentioned above in rarer ectodermal cysts with typical localization in midline structures as the glabella, the anterior fontanelle, and the occiput [1, 2, 9]. Surgical excision is still indicated in all of these lesions. In all other lesions, it seems to be favorable to primarily follow-up these patients by clinical and sonographical observation. The natural course can be easily evaluated and the odds are well enough for a good case scenario that the lesion will show a continuous regression with final disappearance. However, there are also some of lesions which might transiently increase in size followed by spontaneous regression. However, this course of behavior is independent of the initial size and the lesions' vicinity to the skull. However, a primary or secondary osteolysis in all layers of the skull was never observed in the current series. This complication, which is described in previous studies [5, 6], can be easily excluded using sonographic imaging by evaluating the missing translucency through the intact skull and thus avoiding intracranial brain structure visualization. Furthermore, a secondary infection could never be seen as a complicated course in any of our patients. Hence, our data is not verifying these previously reported aspects as a reason for further imaging studies or even surgical interventions of calvarial dermoids or epidermoids.

In terms of the presented data, we also need to discuss methodological drawbacks. Our study is missing a coherent temporal protocol for follow-up investigations in terms of time interval and total period of imaging. Not all of our patients could be investigated until a total disappearance of the lesions could be proven. Especially, the analysis of progressive increasing lesions implies the limitation that this patient group has the shortest follow-up period but the youngest initial age compared to the other cases where a regression could be observed and thus a final conclusion is prevented. Because all patients were seen on an out-patient basis and the study was of retrospective nature, the decision of follow-up was based exclusively on the parent's compliance. The matter of fact that the ultrasonographic diagnosis of a benign lesion without risk of malignant transformation, which was explained toward the parents, might contribute to the decision of the relatives to discontinue the follow-up procedure. Due to the absence for further investigation, it might also be concluded that no further progression of the lesion by parental palpation could be observed nor any further complication, e.g., of an infection, was developed in theses cases.

Differential imaging diagnostic criteria of dermoids/epidermoids needs to be discussed compared to eosinophilic granuloma. In X-ray, both of the lesions show lucency at their local site; however, dermoids/epidermoids primarily show a sclerotic rim due to reactive osteogenesis in a slower growing pattern, which is also shown in Fig. 1. Ultrasonographic features in eosinophilic granuloma include high perfusion pattern in Doppler sonography and transparency of the skull to visualize intracranial structures. Both of these criteria are in contrast to dermoids and epidermoids, which do not show any perfusion signs within the lesion. Moreover, intracranial structures cannot be visualized due to the remaining bony structures represented at least by an intact internal layer of the skull.

As described earlier by Holthusen and colleagues [3] that there is a tendency of calvarial dermoids and epidermoids to spontaneously form back within the first ages of life can be underlined by our observations with a significant number of patients. In their study, where the results are based only on clinical observation, a transient increase of lesion size was also described. However, we strongly recommend the ultrasonographic diagnosis and follow-up of these patients. In our opinion, no indication for additional imaging techniques neither for primary diagnosis nor for follow-up studies is given. The surgical excision of ectodermal cysts is only indicated if the localization is involving midline structures of the skull.

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