Arq Neuropsiquiatr 2003;61(2-B):448-452

CONGENITAL DERMOID INCLUSION CYST OVER THE ANTERIOR FONTANEL

Report of three cases

Humberto Belem de Aquino[1], Carla Ceres Villas de Miranda[1], Cyro Alves de Britto Filho[1], Edmur Franco Carelli[2], Guilherme Borges[2]

ABSTRACT - Congenital dermoid inclusion cyst over the anterior fontanel (CDIC) is an uncommon cystic lesion located over the anterior fontanel. It is a benign and curative lesion and most of the time, can be diagnosed at birth. From 1994 to 2001, three patients were operated with this kind of lesion and after reviewing the literature we found 229 cases and only 6 cases described in Brazil. Our objective in this study is to report three more cases.

KEY WORDS: congenital inclusion, dermoid cyst, epidermoid cyst, anterior fontanel.

Cisto dermóide de inclusão congênita sobre a fontanela anterior: relato de três casos

RESUMO - Cisto dermóide de inclusão congênita sobre a fontanela anterior (CDIC) é lesão rara localizada na região da fontanela anterior. Trata-se de lesão benigna e curável que, na maioria das vezes, é diagnosticada no nascimento. De 1994 a 2001, três pacientes foram operados com este tipo de lesão e, através dos dados disponíveis na literatura, verificamos somente 229 casos descritos, apenas 6 descritos no Brasil, o que nos motivou a registrar mais três casos.

PALAVRAS-CHAVE: inclusão congênita, cisto dermóide, cisto epidermóide, fontanela anterior.

Many different types of lesions over the children's skull exist and some are commonly diagnosed in daily practice. Congenital dermoid inclusion cyst over the anterior fontanel (CDIC) is a rare and benign lesion located over the anterior fontanel. Many children are examinated with lesions over the skull in our hospital. In a recent review of these children, our attention was drawn to three patients with a cystic rounded mass over the anterior fontanel. These three patients showed no neurological abnormality and the diagnosis of CDIC were confirmed by surgery and histological examination. Our literature search found 229 other cases (Table 1) of CDIC worldwide.

CASES

From 1994 to 2001, three patients were operated with a cystic mass over the anterior fontanel, being of different ages and without neurological abnormality at the time. The first patient a three-year-old-white boy. The second patient (Fig 1) a five-month-old-black girl and the third patient (Fig 2) a five-month-old-white boy. All patients were treated with surgery and using the same approach for each of them (Figs 3 to 12 - the second patient shows all the surgical steps, for better illustration). All three patients presented in this paper are similar in all aspects when compared to those in other papers. Two patients were reviewed recently. The first patient (Fig 13 - no photograph prior to surgery) has a learning disability but is not due to the cyst. He was noted to have a small lipoma over the callosal body. The recent photograph shows the cyst has not recurred. The second patient (Fig 14 - recent photograph) does not have any neurological abnormality and the cyst has not recurred. We were unable to contact our third patient but during his last examinations we did not find any problems with him. Photos published with written authorization given by the patient´s parents.

[1]Neurosurgery Service of Hospital Municipal Dr. José de Carvalho Florence, São José dos Campos, SP, Brazil[1]; [2]Discipline of Neurosurgery, Department of Neurology, Faculty of Medical Science, State University of Campinas UNICAMP, Campinas SP, Brazil

Received 12 April 2002, received in final form 28 January 2003. Accepted 30 January 2003.

Dr. Humberto Belem de Aquino - Rua Jaime Spinelli 5 - 12282-436 Caçapava SP – Brasil. E-mail: humebade@iconet.com.br


Fig 1


Fig 2


Fig 3

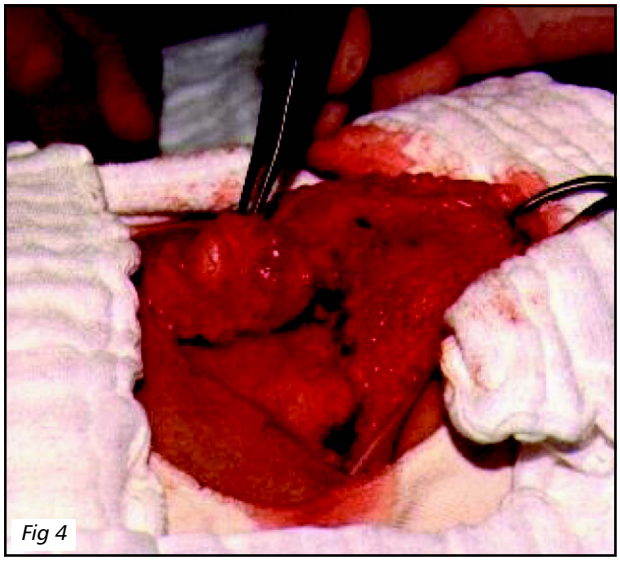
Fig 4

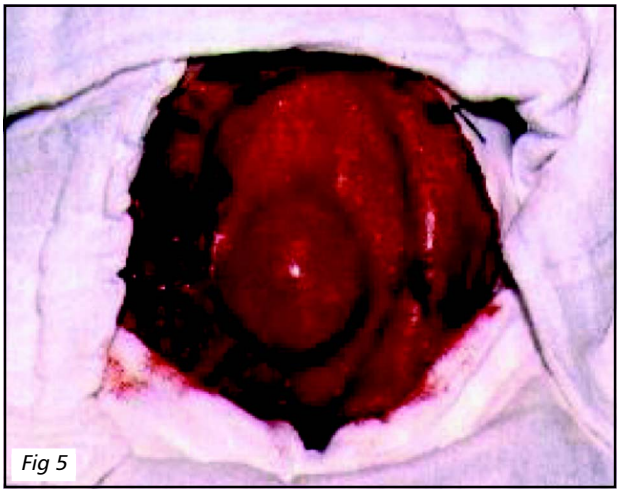
Fig 5

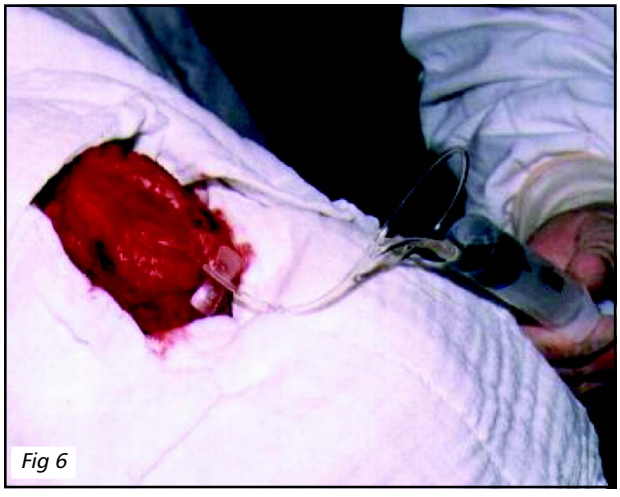
Fig 6

DISCUSSION

Congenital dermoid inclusion cyst over the anterior fontanel is reported as an uncommon cystic lesion, located over the anterior fontanel. Adeloye and Odeku (1971)[1] were the first to publish a clear and complete description about this lesion, having treated eighteen patients. CDIC is a cystic mass covered by normal skin. It is soft, mobile and it does


Fig 7

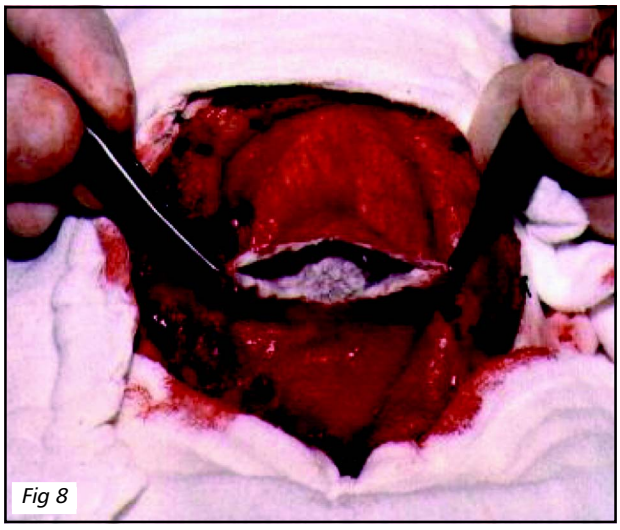
Fig 8

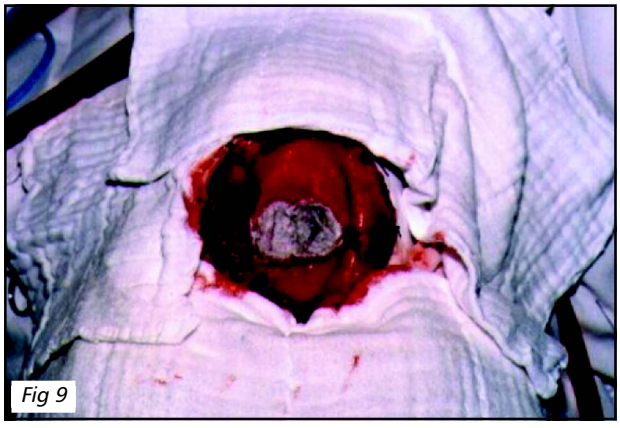
Fig 9

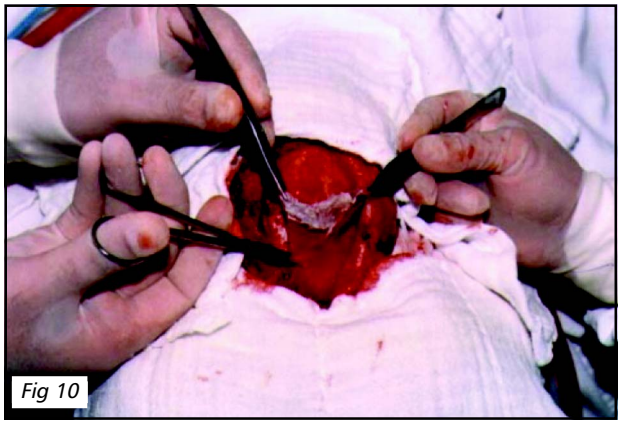
Fig 10

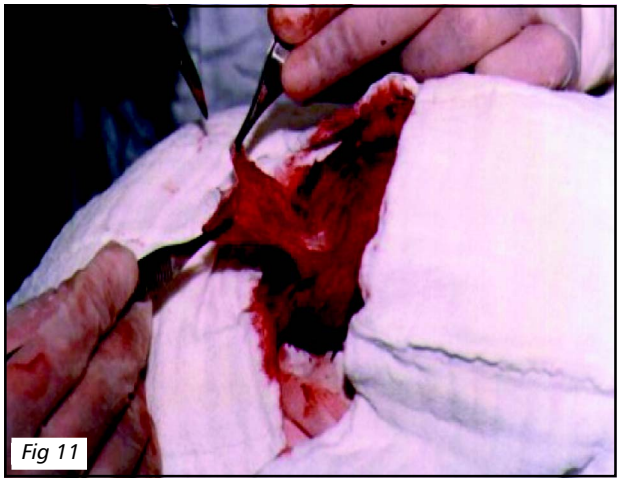
Fig 11

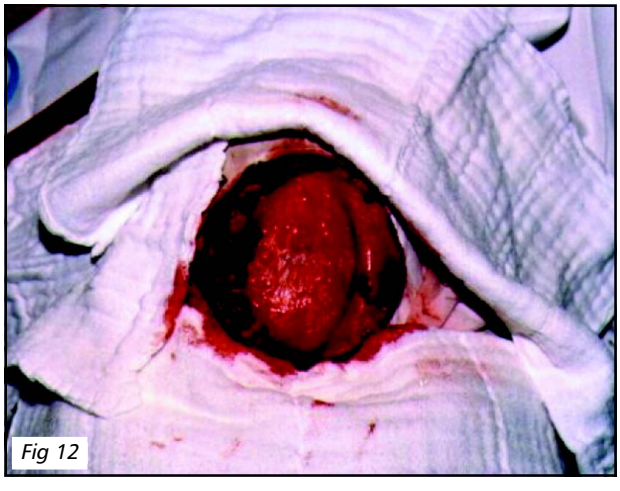
Fig 12

not cause discomfort, pain or throbbing. There is no communication between the cyst and intracranial cavity. Several cystic sizes have been reported[1-4], this depending on the age of patient at the time of diagnosis. Most of the time, the diagnosis can be made at birth, although some authors have reported adult cases[4-6]. CDIC is a developmental tumor due to inclusion of dermal elements within the neuroaxis between the third and fifth week of the embryogenesis when the ectoderm folds into the neural tube[6,7]. Based on its pathogenesis, they can be classified as: 1) congenital dermoid cyst of the teratoma type that


Fig 13


Fig 14

Table 1.CDIC registered cases.*

Hayat S et al.[4]	1989	1 case
Oliveira HA et al.[6]	1989	1 case
Macedo NTL et al.[7]	1985	1 case
Wong TT et al.[21]	1986	8 cases
Pereira CU et al.[13]	2000	2 cases
Tateshima S et al.[14]	2000	1 case
Parizek J et al.[11]	1989	13 cases
Hibaut-Macarde P et al.[16]	1991	1 case
Tan EC et al.[30]	1993	4 cases
Sinclair RD et.al[31]	1992	3 cases
Martinez LS et al.[32]	1992	3 cases
Mlay SM et al.[33]	1993	6 cases
Stannard MW et al.[34]	1990	6 cases
Saito M et al.[35]	1988	2 cases
Isozumi T et al.[36]	1995	1 case
Nicolau A et al.[37]	1986	6 cases
Peter JC et al.[38]	1992	35 cases
Total		94 cases

*Review of the liteture by Macedo NTL et al. until 1984 – 135 cases reported

is derived from the embryogenic epithelium, confined to the ovaries and testis; 2) acquired implantation dermoid cyst formed by cells implanted traumatically into deeper structures; 3) congenital dermoid inclusion cyst resulting from the inclusion of displaced dermal cells along the embryonic fusion line[7]. Histologically, the cyst wall is lined by squamous epithelium and inside the cyst exists adnexial appendage structures including hair follicles, sebaceous and sweat glands[8-13]. There are factors that change other lesions over the anterior fontanel, such as epidermoid cyst. The fluid content can be clean or yellow, depending on the size and age of the lesion and exocrine sweat gland content and some authors discovered sodium, potassium, chloride and glucose concentrations within their cases, has been low incidence[14,15]. A skull X-ray will show the shadow of the swelling in the extracranial space and can reveal changes which include flattening of the outer table underneath the swelling or some depression[16,17]. In the past, some authors injected air or contrast medium (Pantopaque), others used ventriculography, in order to show interconnection between the cyst and intracranial cavity[18]. Currently, CT and MRI are considered the best examination methods, to confirm its extracranial position[19-21]. Encephalocele, meningocele, hemangioma, lipoma, cephalohematoma, sebaceous cyst, pilonidal cyst and sinus pericranii are important parts of the differential diagnosis[22-30]. The-

re are no reports about neurological abnormality or any recurrence of the lesion. The surgical indication is based on preventing subsequent infection, pathological diagnosis and an aesthetic aspect.

In conclusion this lesion is benign, simple and easy to treat by surgery and free of significant surgical complication. However, we believe there is a greater number of such cases, other than the 229 already reported. Perhaps other cases have not been reported because it is considered to be a simple lesion. In the cases reported in the literature, we did not find any description addressing the long-term outcomes of patients who were treated. Therefore, we are including our cases in order to provide a long-term view of this condition. We predict our results will prove that CDIC is a benign and curative lesion.

Acknowledgments - We thank Lynn Falkner from Salt Lake City (Utah – USA), Carolina Brom from Caçapava (São Paulo) and Andy Shepherd from England, for English support.

REFERENCES

1. Adeloye A, Odeku EL. Congenital subgaleal cysts over the anterior fontanelle in Nigerians. Arch Dis Child 1971;46:95-96.
2. Glasauser FE, Levy LF, Auchterlonie WC. Congenital inclusion dermoid cyst of the anterior fontanel. J Neurosurg 1978;48:274-278.
3. Sonntag VKH, Waggener JD. Congenital dermoid cyst of the anterior fontanel in a mexican-american. Surg Neurol 1980;13:371-373.
4. Hayath S, Seetharam W, Kumari G, Dinakar I, Nightingale F. Congenital dermoid cyst over the anterior fontanelle. Br J Clin Pract 1989;43:119- 120.
5. Ojikutu NA, Mordi VPN. Congenital inclusion dermoid cyst located over the region of the anterior fontanel in adult Nigerians. J Neurosurg 1980; 52:724-727.
6. Oliveira HA. Cisto dermóide de inclusão localizado na região da fontanela anterior no adulto. Arq Neuropsiquiatr 1989;47:375-377.
7. Macedo NTL, Ramos VP, Lins C. Cisto dermóide de inclusão da fontanela anterior. Arq Neuropsiquiatr 1985;43:407-412.
8. Zülch KJ. Brain tumor The biology and pathology Epidermoid and dermoid cyst. 3.Ed. New York: Springer Verlag, 1986:433-437.
9. Brownstein MH, Helwig EB. Subcutaneous dermoid cysts. Arch Dermatol 1973;107:237-239.
10. Naidich TP. Dermoids of the anterior fontanelle. Neuro Image Quiz, Answers 1988:278-279.
11. Parízek J, Nìmecek S, Nemecková J, Cernoch Z, Šercl M. Congenital dermoid cyst over the anterior fontanel: report on 13 cases in Czechoslovak children. Child's Nerv Syst 1989;5:234-237.
12. Chaudhari AB, Ladapo F, Mordi VPN, Choudhury KJ, Naseem A, Obe JA.Congenital inclusion cyst of the subgaleal space. J Neurosurg 1982;56:540-544.
13. Pereira CU, Santos DPS, Machado JC, Machado SC, Araújo ES, Costa MD. Cisto dermóide de inclusão congênita localizada na fontanela anterior. Arq Bras Neurocir 2000;19:32-35.
14. Tateshima S, Numoto RT, Abe S, Yasue M, Abe T. Rapidly enlarging dermoid cyst over the anterior fontanel: a case report and review of the literature. Child's Nerv Syst 2000;16:875-878.
15. Kanamaru K, Waga S. Congenital dermoid cyst of the anterior fontanel in a Japanese infant. Surg Neurol 1984;21:287-290.
16. Hubault-Marcade P, Hepner-Lavergne D, Pannier M. Kiste dermoïde du cuir chevelu: à propos d'un cas. Ann Chir Plast Esthét 1991;36;452-456.
17. Ogle RF, Jauniaux E. Fetal scalp cysts: dilemmas in diagnosis. Prenat Diagn 1999;19:1157-1159.
18. Shahabi S, Busine A. Prenatal diagnosis of an epidermal scalp cyst simulating an encephalocele. Prenat Diagn 1998;18:373-377.
19. Stokes RB, Saunders CJ, Thaller SR. Bregmatic epidermoid inclusion cyst eroding both calvarial tables. J Craniof Surg 1996;7:148-150.
20. Kriss TC, Kriss VM, Warf BC. Recurrent meningitidis: the search for the dermoid or epidermoid tumor. Pediatr Infect Dis 1995;14:697-700.
21. Wong TT, Wann SL, Lee LS. Congenital dermoid cyst of the anterior fontanelle in Chinese children. Child's Nerv Syst 1986;2:175-178.
22. Mohanty S, Clezy JKA, Adeloye A. Dermoid cyst of the anterior fontanel. J Neurosurg 1978;48:627-628.
23. Pereira WC, Andrade AF, Lopes PG. Cisto dermóide na região do bregma: relato de dois casos. Arq Neuropsiquiatr 1969;27:349-352.
24. Fujimaki T, Miyazaki S, Fukushima T, Sato Y, Fujimaki W, FujitaY. Dermoid cyst of the frontal bone away the anterior fontanel. Child's Nerv Syst 1995;11:424-427.
25. Yuasa H, Tokito S, Izumi K, Oyama M. Congenital inclusion dermoid cyst of the anterior fontanel in a Japanese infant: case report. Neurosurgery 1981;9:67-69.
26. Chaudhari AB, Rosenthal AD. Congenital inclusion cysts of the subgaleal space. Surg Neurol 1984;21:61-66.
27. Stella L, Spaziante R, Maiuri F, Gangemi M, Divitiis E. Congenital dermoid cysts at the anterior fontanelle. Neurochirurgia 1984;27:186-189.
28. Borges A, Carelli EF, Maciel Jr. JA, Alvarenga M, Castro R, Bonilha L. Pilonidal cyst on the vault:case report. Arq Neuropsiquiatr 1999;20:273-276.
29. Mehta MH, Patel RV. Congenital midline subgaleal cyst. Indian Pediatrcs 1990;27:403-404.
30. Tan EC, Takagi T. Congenital inclusion cyst over the anterior fontanel in Japanese children: a study of five cases. Child's Nerv Syst 1993;9:81-83.
31. Sinclair RD, Darley C, Dawber RP. Congenital inclusion dermoid cyst of the scalp. Australas J Dermatol 1992;33:135-140.
32. Martinez-Lage Sanches JF, Almargro Navarro MJ, Poza Poza M, Puche Mira A, Sola Perez J. Dermoide cyst of the anterior fontanelle in children: clinical significance and differentiation from encephalocele. An Esp Pediatr 1992;36:355-358.
33. Mlay SM, Sayi EN. Anterior fontanelle scalp cysts in infancy. East Afr Med J 1993;70:578-579.
34. Stannard MW, Currarino G. Subgaleal dermoid cyst of the anterior fontanelle: diagnosis with sonography. AJNR Am J Neuroradiol 1990;11:349-352.
35. Saito M, Takagi T, Ishikawa T. Dermoid cyst of the anterior fontanel: advantage of MRI for the diagnosis. Brain Dev 1988;10:252-255.
36. Isozumi T, Tsuji A, Nakasu M, Handa J. Congenital dermoid cyst over the anterior fontanelle: case report. No Shinkei Geka 1995;23:423-427.
37. Nicolau A, Daney I, Diard F, Risch M, Kind M. Midline subepicranial (subgaleal) dermoid cysts in children: report of 6 cases and review of the literature. Ann Radiol (Paris) 1986;29:511-518.
38. Peter JC, Sinclair-Smith C, De Villiers JC. The congenital bregmatic dermoid: an African cyst? Br J Neurosurg 1992;6:107-114.