

CONGENITAL DERMOID INCLUSION CYST OVER THE ANTERIOR FONTANEL

Report of three cases

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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 examined 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.

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Fig 1



Fig 2



Fig 3

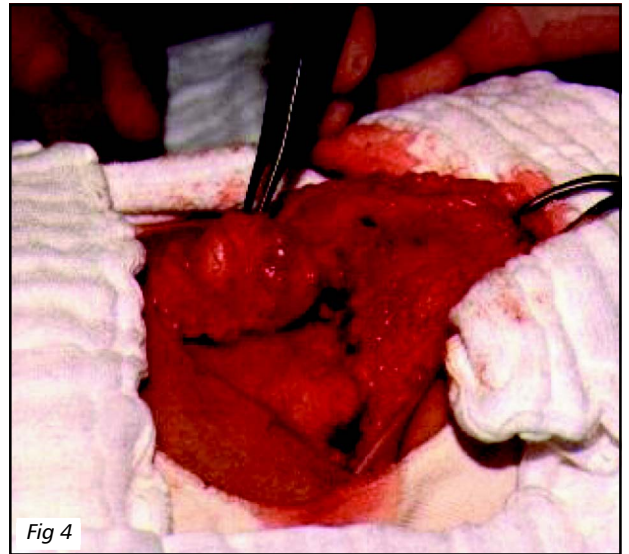


Fig 4

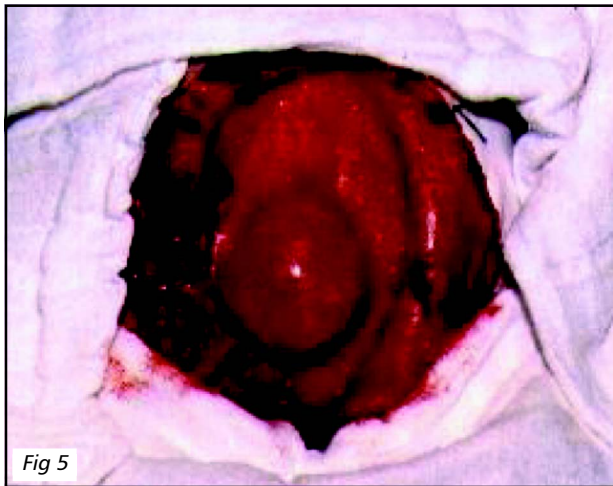


Fig 5

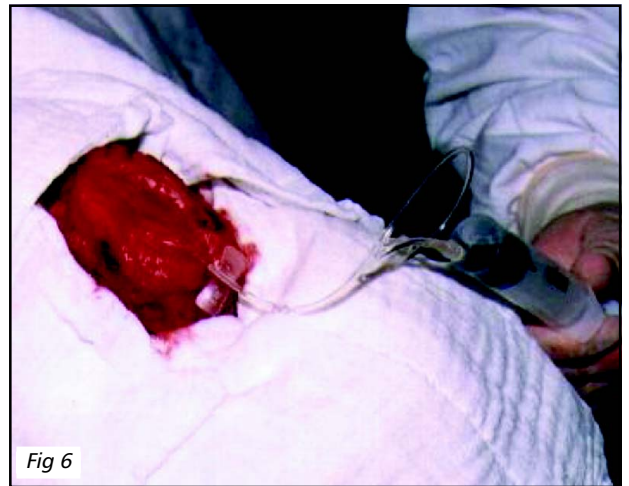


Fig 6

DISCUSSION

Congenital dermoid inclusion cyst over the anterior fontanel is reported as an uncommon cystic lesion, located over the anterior fontanel. Adeloje

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

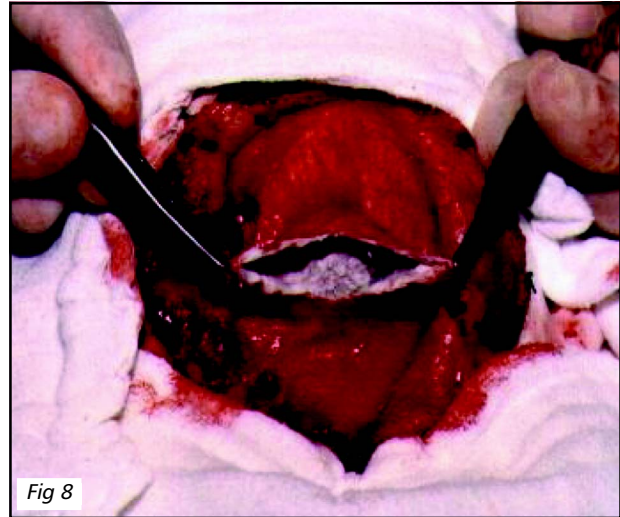


Fig 8



Fig 9

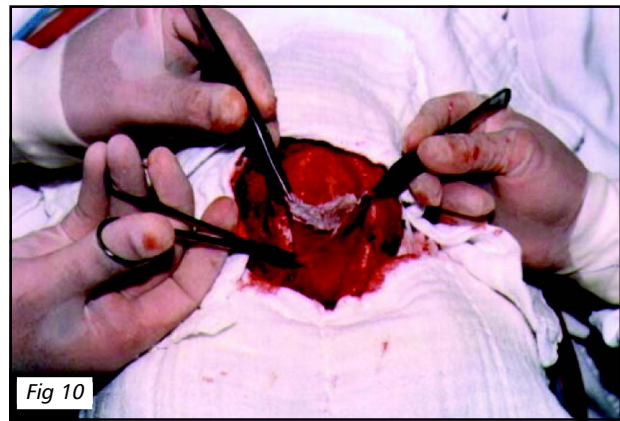


Fig 10



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 reported¹⁻⁴, 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

cases⁴⁻⁶. 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 literature 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 line⁷. Histologically, the cyst wall is lined by squamous epithelium and inside the cyst exists adnexial appendage structures including hair follicles, sebaceous and sweat glands⁸⁻¹³. 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 cavity¹⁸. Currently, CT and MRI are considered the best examination methods, to confirm its extracranial position¹⁹⁻²¹. Encephalocele, meningocele, hemangioma, lipoma, cephalohematoma, sebaceous cyst, pilonidal cyst and sinus pericranii are important parts of the differential diagnosis²²⁻³⁰. 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.

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