Congenital Inclusion Cysts of the Anterior Fontanelle

Gervásio Teles Cardoso de Carvalho, M.D., Walter José Fagundes-Pereyra, M.D., João Antônio Pinheiro Marques, M.D., Fernando Luiz Rollenberg Dantas, M.D., and Atos Alves de Sousa, M.D., Ph.D.

Department of Neurosurgery, Santa Casa Hospital of Belo Horizonte, School of Medical Science of Minas Gerais-Belo Horizonte, Minas Gerais, Brazil


BACKGROUND
Congenital inclusion cysts of the anterior fontanelle are rare lesions. Both dermoid and epidermoid cysts are located in the midline of the scalp and occupy the subgaleal space.

METHODS
We report 7 cases, 4 boys (57.1%), and 3 girls (42.9%), with ages ranging from 3 months to 16 years (mean 40.85 ± 68.56 months; median 10 months). Four patients (57.1%) were white and 3 (42.9%) were Afro-Brazilian.

RESULTS
The cysts had manifested soon after birth in all patients. They gradually enlarged, with no intracranial extensions. Four patients had dermoid cysts and the others had epidermoid cysts. All of them underwent surgery with complete excision of the cyst and no capsular rupture. There were no complications and no deaths.

CONCLUSIONS
Congenital inclusion cysts of the anterior fontanelle are rare lesions that usually manifest at birth. The diagnosis is usually easy and surgery is mandatory, with a good prognosis. Recurrence is rare. © 2001 by Elsevier Science Inc.

KEY WORDS
Congenital inclusion cyst, anterior fontanelle, dermoid cyst, epidermoid cyst.

C ongenital inclusion cysts of the anterior fontanelle are rare lesions, accounting for 0.2% of all inclusion cysts [8,14,21,25]. The terms dermoid and epidermoid were suggested by Brostrøen [5,6] in 1897 to define the embryonic inclusions in the deep layers of the epithelial tissue. However, the first description of a dermoid cyst was that of Veratus, in 1745, and an epidermoid cyst was first described by Dumeril [cited by Carmel]. Cruveilheier (1829) introduced the expression “pearl-like tumour” (“tumeur perille”) to refer to them, as Bailey points out [3].

Because of the rarity of these lesions, we decided to write the present study, based on the analysis of 7 cases and on literature review, emphasizing the histological findings, radiological investigations, and surgical treatment.

Patients and Methods
Seven patients with inclusion cysts of the anterior fontanelle were studied. They were operated at the Department of Neurosurgery of Santa Casa Hospital in Belo Horizonte between 1992 and 1995.

Age ranged from 3 months to 16 years (mean 40.85, SD 68.56, median 10 months). Four patients (57.1%) were female and 3 (42.9%) male. Four patients were white and 3 were Afro-Brazilian.

In all cases, the lesion could be seen at birth as a soft tumor, neither painful nor adherent to the deep layer, showing progressive growth and no associated symptoms or signs (Figure 1). No previous occurrence of such lesions was found in the patients’ family histories.

They were all studied with standard skull X-rays and computed tomography (CT) scans, in both the preoperative and postoperative periods.

The patients were operated in the supine position, with the head straight. Six of them were under general anesthesia, and the other under local anesthesia and sedation. A coronal skin incision was made behind the hair line. Care was taken not to open the cyst capsule as the skin is being lifted (Figure 2A). Dissection was then performed around the lesion to completely excise the tumor without rupturing it (Figures 2B–D).
The presence of intracranial extension must be checked beforehand. If it is found, the dura mater must be carefully examined.

**Results**

The skull X-rays and the CT scans demonstrated bone erosion without intracranial communication in all patients (Figure 3).

We were able to completely excise the lesion in all 7 cases.

Histology showed that 4 of the lesions were limited by a squamous epithelium containing skin appendages, including hair follicles and glands, compatible with dermoid cysts (Figure 4). The other 3 lesions had a rather thin and pearly white wall, surrounded by a delicate layer of squamous cells, producers of keratin, containing cellular debris from the desquamating skin, compatible with epidermoid cysts (Figure 5).

The postoperative course was uneventful and the final outcome was good in all cases. There were no deaths in this series.

**Discussion**

Congenital inclusion cysts are usually located in the midline at the neural groove closure zone from the cranium to the sacrum [7,10,14,18,23,29,32]. They can be either dermoid or epidermoid cysts. The dermoid cysts are classified into three categories: 1) congenital dermoid cysts of teratoma type; 2) acquired implantation dermoid cysts, and 3) congenital inclusion dermoid cysts [29].

Dermoid cysts are distinguished from epidermoid by the presence of hair, sebaceous glands, and sweat glands [6]. Both present either desquamating keratin epithelium or clear fluid and cholesterol [9,22]. According to Naidich [17], 27% of those located in the neck and the cranium are in the anterior fontanelle, mainly at its anterior angle. However, New and Erich [19], who studied 495 cases of dermoid cysts at different locations, found 103 in the head (20.8%), only one of which was in the anterior fontanelle (0.2%). Fleming and Botterel [9] described four cases of dermoid cysts in the anterior fontanelle within a group that included patients with cysts in other locations. One of them was operated on at the age of 20, even though the lesion had been diagnosed at birth. Such lesions are rare in children and extremely rare in adults [1]. Hubault-Marcade et al [12] found only seven cases in adults in their review of the literature, and in all of them the lesion had been present since birth.

About 174 cases of congenital inclusion cysts of the anterior fontanelle had been described in the world literature by 1989 [23]. Parizek et al [23] reviewed them and found that 30 patients were from Europe, 74 from America and 45 from Africa [1,2,15]. The first European case was described in 1984 by Stella et al [22]. In 1986, 8 cases were reported in China [23]. There is only one reference to a case in India [16]. Seven cases of such cysts were reported in Brazil [8,14,21,24,25]. Another 20 new cases have been described in the world literature since 1989 [23].

The largest series was by Pannell et al [22] with 25 congenital inclusion cysts of the anterior fontanelle among 94 cases of cysts located in the cranium (26.6%).

Several embryological theories have been proposed to distinguish dermoid cysts from epidermoid. The primitive epithelium has a high capacity to differentiate. Therefore, in the early stages of inclusion, they can give rise to both dermoid and
epidermoid cysts. However, if the inclusion happens later on, it will result in epidermoid cysts only [22].

Inclusion cysts are usually present at birth and can develop gradually as secretion and internal desquamation accumulate [1,7,9,10,13,17,18,20,22,25,27,29,32], although there have been reports of cysts that were found one year after birth [21]. They are single, soft, not painful, usually transparent, and can be pulsatile and increase in size with crying or physical effort [1,3,10,17,22,27,31].

At first, inclusion cysts were thought to be more frequent among Africans or Afro-descendants [1,10,20,27], but many authors have found this statement not to be true [12,13,14,17,18,22,25]. They occur predominantly in females (2:1) [10,17,31] except for 1 report in which there was a predominance in males (5:3) [34]. But no significant relationship to sex or race was observed in the present study. There have been no descriptions of associated malformations nor of relevant family history [1,10,17], despite the fact that some authors have reported the occurrence of dermoid cysts of the anterior fontanelle in parents and their children [19]. Multiple epidermoid cysts were found in 1 patient, including one in the anterior fontanelle [18].

These cysts are lesions that develop from congenital inclusions of epithelium or dermal tissues along the midline or lateral to it in the embryonic...
stage. They grow between the third and the fifth weeks of pregnancy as an ectodermal implant is held in the deepest layers of the skin [9,12,13,19,20, 24,32,33]. The lesion can be located in the diploe, in the pericranium, or in the scalp if the epithelial cells are involved in the membranous bone of the cranium during its growth [10,12,32]. Inclusion cysts can also develop as a result of the implantation of epithelial cells in the deepest layers by lumbar puncture, cranial trauma, or surgical procedures or experiments [9,12,17,18,19,32].

These cysts arising in the bones of the calvaria develop from inclusion material that proliferates within the bone, causes gradual absorption of the adjacent bone, and produces a well defined area of bone destruction [10].

Differentiation of dermoid and epidermoid cysts is not always easy [12]. The secretion of exocrine glands that are found in the cysts’ walls can produce a fluid chemically similar to sweat. Two cases had cells reactive to induced inflammation caused by the presence of cholesterol [22].

Although epidermoid cysts represent 0.5% to 1.8% of all embryonic malformations whereas dermoid cysts account for 0.1% to 0.7% [11], dermoid cysts are more common than epidermoid cysts in children [22]. According to Pannell et al in their literature review up to 1982, only 2 cases of epidermoid cysts of the anterior fontanelle had been described [22]. Epidermoid cysts are usually located at the base of the skull and at the cerebellopontine angle, whereas dermoid cysts are most commonly found in the midline. Dermoid cysts are encapsulated, subgaleal, and do not communicate with the skin nor with the brain [7,10,14,17,22,25,27,31]. They can sometimes erode and depress the skull [12,17,22,26]. Their size ranges from 1 to 7 cm [14,23,25].

Histological studies comparing craniopharyngio-
mas, middle ear cholesteatomas, and Rathke’s cleft cysts to inclusion cysts have demonstrated close similarities [18]. Inclusion cysts rarely become malignant [17,20,22]. Twelve cases in which squamous cells turned into carcinoma have been described, but they were not located at the anterior fontanelle [17,18,22,31,32].

Skull X-rays may aid in diagnosis as they reveal erosion and the flattening of the external skull bone structure [1,10,12,13,14,17,20,22]. Ultrasonography may be helpful for diagnosis in children with open fontanelles and it also shows possible communication between the cyst and the brain. The CT scan shows the lesion very clearly: isodense or hypodense, encapsulated—thus, separated from the dura mater and the skull—without contrast enhancement and sometimes with erosion of the skull bone [12,14,17,18,22,29,31]. The magnetic resonance image (MRI) is the most reliable test, demonstrating all the results of the other available tests with greater accuracy [31].

The differential diagnosis includes the anterior encephalocele, sebaceous cyst, lipoma, hemangioma, cephalhematoma, subgaleal hematoma, lymphangioma, sinus pericranii, and abscess [1,10,14,20,22,26,27,30,35].

Treatment is surgery, and the whole lesion should be excised including its capsule that can be totally separated from the galea and the skin by means of a linear sagittal incision across the middle of the lesion [1,4,7,10,12,13,17,22,25,27]. If a mistaken attempt is made to drain the cyst by puncture, an infection might occur, and the cyst will also fill up again [1,10]. Therefore, this procedure should be avoided.

Prognosis is usually excellent, with almost no recurrence being reported [1,10,12,17,22]. There has been only one case of progressive and slow recurrence 6 years after surgery [1].

CONCLUSION
Inclusion cysts of the anterior fontanelle are rare lesions accounting for around 0.2% of all inclusion cysts. They manifest at birth and diagnosis is usually easy. Surgery is the only advisable treatment and the prognosis is very good, recurrence being rare.

REFERENCES