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Adeloye-Odeku Disease: An African Disease in the Indian Child?

Adeloye-Odeku Hastalığı: Hintli Bir Çocukta Bir Afrikalı Hastalığı

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ABSTRACT

The "Adeloye-Odeku disease" or congenital dermoid cysts over the anterior fontanelle, is a rare congenital disorder of children initially described in Nigeria. It has been rarely reported in the Indian population (3 cases in 2 reports). These rare lesions are operated on by neurosurgeons in view of their location and differential diagnoses. We present two infants with the condition and a comprehensive review of the literature on pubmed using six common phrases used to describe this condition. The review was undertaken to analyze the reasons for the paucity of cases from the Indian sub-continent. The ethnicity, differential diagnosis, radiological features and management is discussed.

KEYWORDS: Adeloye-Odeku's disease, Dermoid cyst, Anterior fontanelle, Pediatric

ÖZ

"Adeloye-Odeku hastalığı" veya anterior fontanel üzerinde konjenital dermoid kistler ilk olarak Nijerya'da tanımlanmış nadir bir çocuk çağı konjenital hastalıktır. Hint popülasyonunda nadiren bildirilmiştir (2 sunumda 3 vaka). Bu nadir lezyonlar konumları ve ayırıcı tanıları nedeniyle beyin cerrahları tarafından ameliyat edilir. Bu durumun görüldüğü iki vakayı ve bu durumu tanımlamak üzere sık kullanılan altı sözcük kullanılarak pubmed literatürünün kapsamlı bir taramasının sonuçları sunuyoruz. Derleme hint alt kıtasında vakaların azlığının nedenlerini analiz etmek için yapıldı. Etnik köken, ayırıcı tanı, radyolojik özellikler ve takip anlatılmaktadır.

ANAHTAR SÖZCÜKLER: Adeloye-Odeku hastalığı, Dermoid kist, Anterior fontanel, Pediyatrik

INTRODUCTION

"Adeloye-Odeku disease" is the presence of a congenital inclusion dermoid cyst over the anterior fontanelle. It was first described by Adeloye and Odeku from Nigeria in 1971 (1) and was considered a disease of African children. Subsequent reports, in various races, have appeared from across the globe (2-20). There are only three documented cases in two reports of Indian origin in the English literature (18,14). We present two interesting cases of Indian origin and review the available literature from Pubmed. Most of these cases are operated by neurosurgeons and need to be differentiated from other scalp swellings specifically meningoencephalocoeles. The implications, diagnosis and management are discussed.

CASE REPORT

Case One:

An 8-month-old female child presented with history of swelling over the anterior fontanelle since birth. It was soft, non-tender and had increased in size since 3months. The MRI of the skull revealed a lesion hypointense on T1Wimages and hyperintense on T2Wimages. There was no post-contrast enhancement and no obvious intracranial extension of the lesion (Figure 1A-D).

Case Two:

This 6-month-old male child was brought to us with history of gradual swelling in the region of the anterior fontanelle since birth. The swelling had increased in size since 3 months and would appear tense while crying. On examination a 9x10 cm swelling over the anterior fontanelle was present (Figure 3A) which was soft in consistency, non tender and demonstrated trans-illumination (Figure 3B). Figure 2A-D demonstrates the radiological workup. In addition to the standard CT and MRI an MR venogram and a 3D CT were also performed to delineate the size of the defect and demonstrate the non-involvement of the superior sagittal sinus.

Both infants underwent surgery under general anesthesia. A bicoronal incision posterior to the hairline was used. The cyst was excised in- toto by blunt dissection and excess scalp in the second case was trimmed (Figure 3C, D). There was no adherence to the superior sagittal sinus and no intra-dural extension was noted.

The incision was sutured with absorbable vicryl and neither of the children had a recurrence.

The content in both cysts was clear and the cyst walls were smooth. The lining demonstrated keratinized squamous

epithelium and formation of hair follicles and eccrine glands (Figure 4A,B).

DISCUSSION

The Adeloye-Odeku disease (1) has been described by various terms (1-20). A pubmed literature review was carried out with the 6 commonest phrases outlined below and a total of 20 reports (of which 2 were epidermoids and were omitted from the review) were identified in literature: dermoid cyst over anterior fontanelle (1,3,10,11,12,16,17,20) (8 reports),

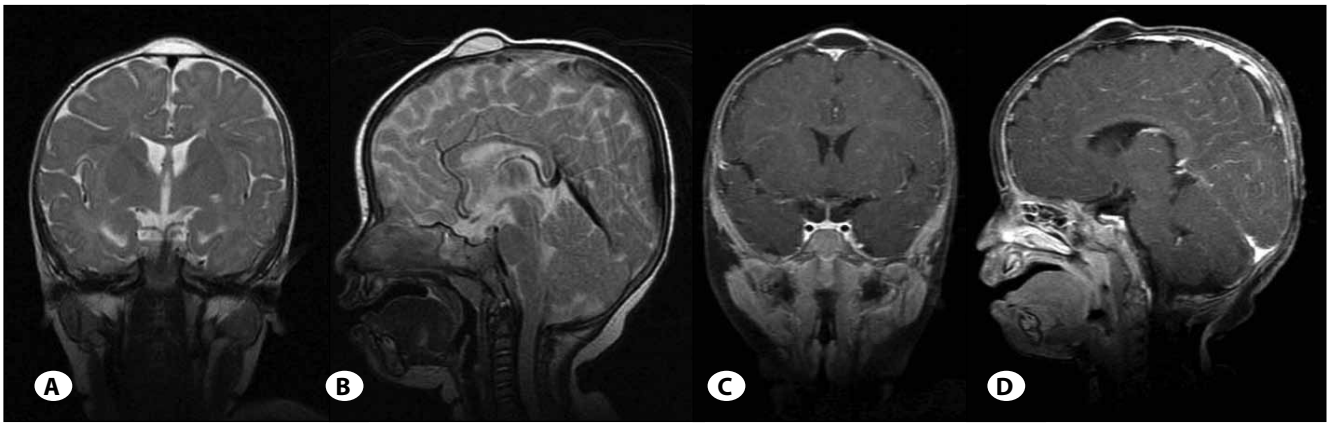

Figure 1: A, B) Reveal the coronal and sagittal T2 weighted images of case 1. **C, D)** A subgaleal cyst is visualized over the anterior fontanelle with no post contrast enhancement.

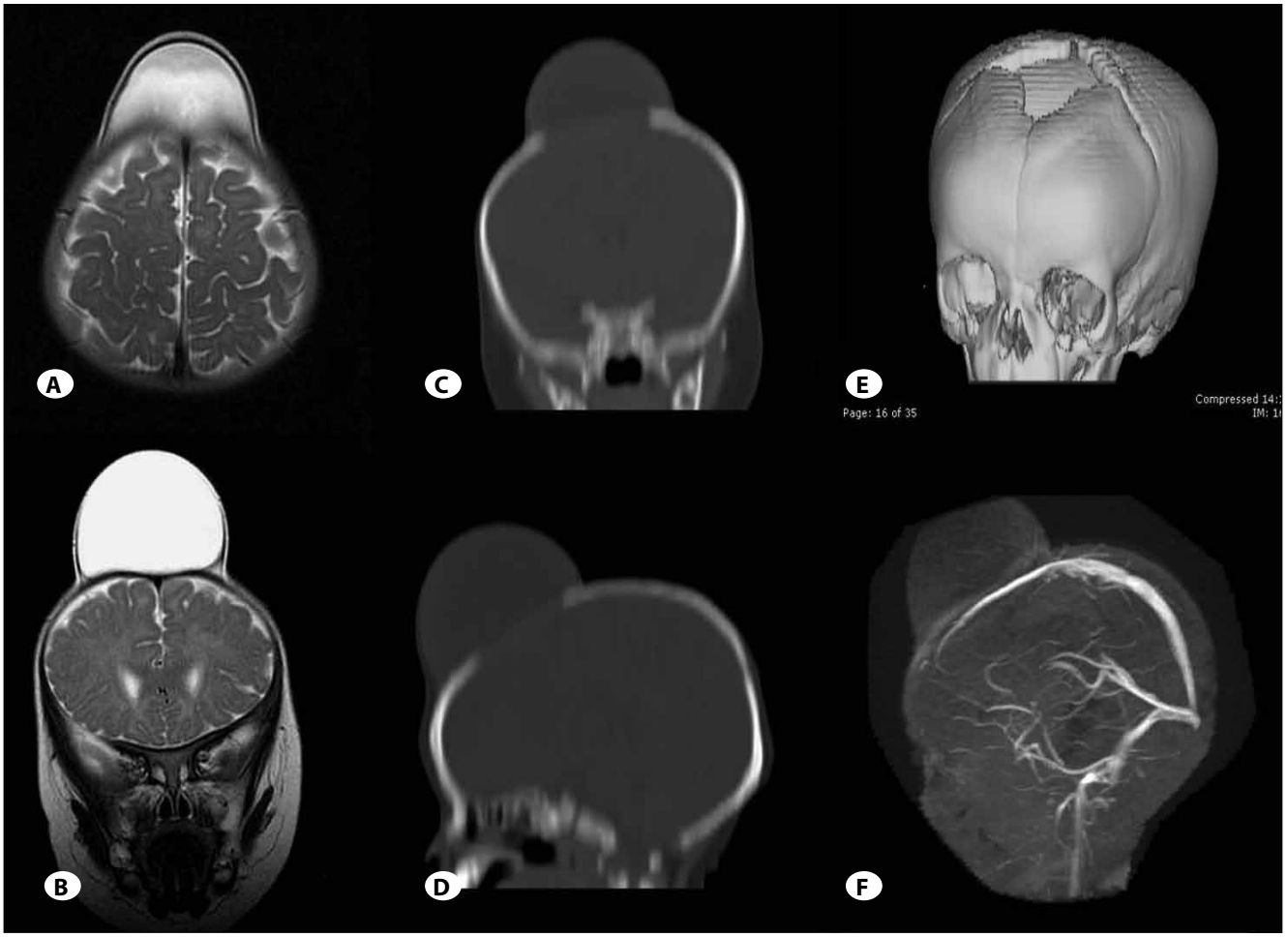

Figure 2: Depicts the radiology in case 2. **A,B)** reveal the axial and coronal characteristics of the cyst filled with csf intensity. **C, D)** depict the asymmetric enlargement of the anterior fontanelle more on the right side. **E)** depicts the 3D CT reconstruction of the same and in f the MR venogram is depicted with demonstration of negative involvement of the superior sagittal sinus.

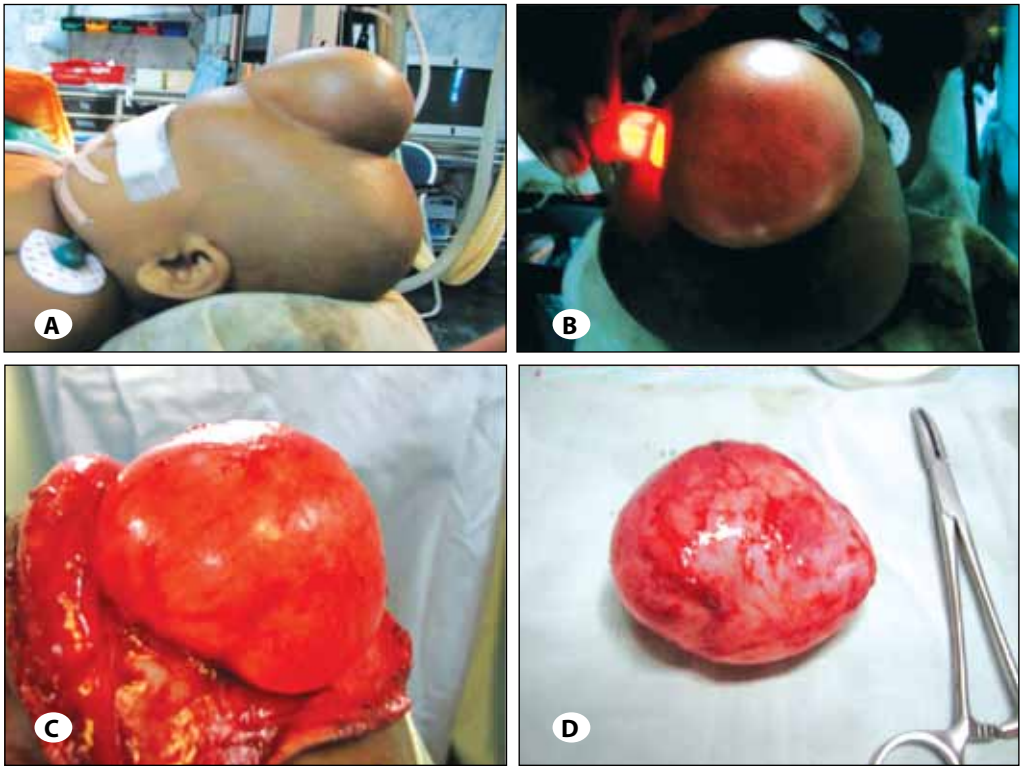

Figure 3: A) Shows the extent of the lesion clinically in case 2.
B) demonstrates the positive trans-illumination test.
C) reveals the intra-op dissection and
D) the removal of the cyst in-toto.

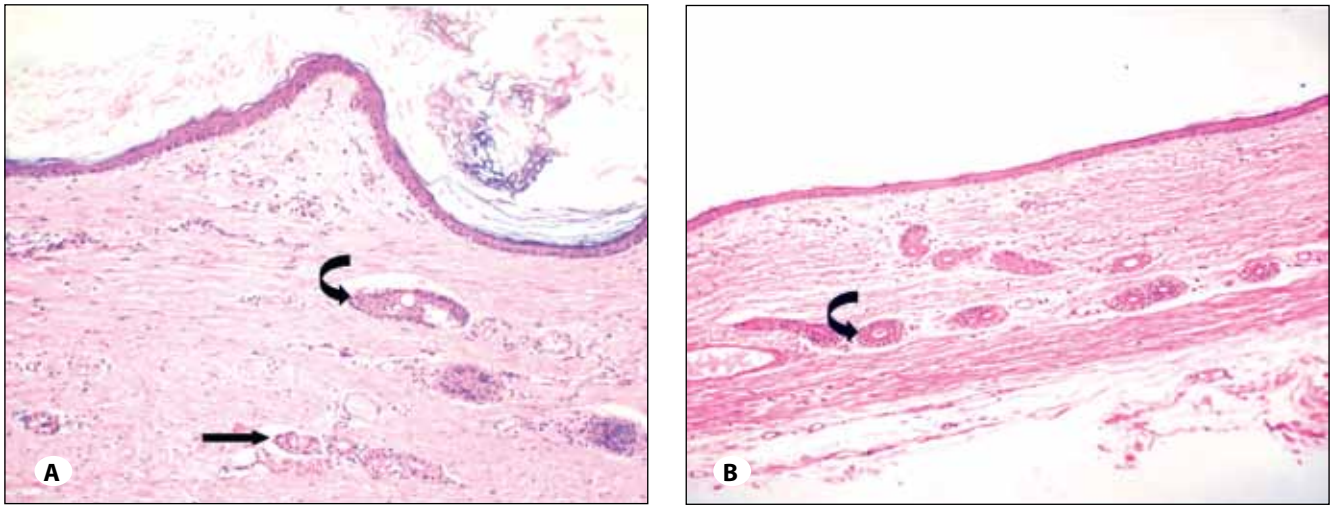

Figure 4: Paraffin section of **Case1 (A)** and **Case 2 (B)** showing cyst wall lined by keratinized squamous epithelium and formation of hair follicles (curved arrow) and eccrine glands (straight arrow). (Hematoxylin & Eosin x100).

Congenital inclusion cysts over the anterior fontanel (6, 7,14,15,19) (6 reports), Congenital inclusion cyst of the subgaleal space (2,4,5,7,9) (4 reports), congenital bregmatic cysts (bregmatic inclusion cysts / bregmatic dermoid cysts) (one report) (18). This data was scrutinized predominantly for ethnicity, country of origin and sex predeliction (Table I). The number of reports against each phrase has been mentioned in parenthesis. This different reports with demonstrates the need for unified nomenclature in medical literature for rare conditions to aid relevant research.

About 20% of all scalp dermoids have been found in this location in a large series and it is thus said to be the commonest site on the scalp for dermoids (16). The preponderance to this location has been related to a greater chance of epithelial displacement at suture sites especially since the anterior fontanelle is the largest fibrous membrane suture in the skull (20).

This condition was initially associated with the race of origin of the children. The earlier reports were mainly from Africa, especially Nigeria (1, 5, 9,14). Over 229 cases of this

condition have been reported in literature until 2003 (6). The approximate number of total reported cases from different countries and races are as follows: (Table I) Nigeria/Africa – almost a 100 cases (1, 2, 5, 9,14,18), Japan- 15 (19), China 13 (20), Czechoslovakia – 13 (17), Canada -23 (16), Brazil 6 (6), Turkey -4 (3) and India 3 (13,18). A series of mixed ethnicity of Mestizo and Mullato children has also been published (8).

A large series of 35 Red Cross War Memorial Children's Hospital and Groote Schuur Hospital between 1969 and 1990 in which they reported 2 children being of Indian origin (18). The only other reference to this condition in an Indian child is by Mohanty et al. in a letter to the editor mentioning the extremely rare occurrence of this condition in Indian children (13) giving a figure of 1.3% of all reported cases. We report 2 infants presenting with this condition in a span of 3 years in a busy neurosurgical setup. This would pre-suppose a much higher incidence of the condition than reported earlier. The possible reasons for not recognizing this condition may be the management by diverse specialties, e.g., Pediatric surgeons, general and plastic surgeons.

Most cases reported in literature have been operated by neurosurgeons. This is primarily due to referral system to tertiary centers in which most congenital calvarial midline swellings in children are assumed to be meningoencephalocoeles (1,16,18). In the original description by Adeloye and Odeku one child had an encephalocoele (1). Encephalocoeles over the anterior fontanelle are extremely rare (1). We have used the eponymous term of "Adeloye-Odeku" disease for this condition to commemorate the original description of this rare disease entity (1, 8).

There have reports with both male preponderance (3, 7, 17, 18,19) and female preponderance in large series (8,16). In the present report we had one male and one female child. The size of the lesion has been associated with the age of the patient as a general rule in some reports (18) but in others this

Table I: Pubmed Review of all Reported Cases Using the Phrases Outlined Below

s. no.	Reference	Year	No. of patients	Race/Country	M:F
1	Adeloye A, Odeku EL [1]	1971	18 [O]	African/ Nigeria [a]	1:2
2	F.E Glauser et al. [9]	1978	11	African / Rhodesia [b]	1:2
3	Ojikutu NA et al. [14]	1980	2 [#]	African /Nigeria [b]	1:1
4	Chaudhari AB et al. [5]	1982	21	African /Nigeria [c]	1:2
5	Pannell BW et al. [16]	1982	25	Canada 23 Caucasians [a] 2 Not mentioned	1:2
6	Kanamaru K et al.	1984	1	Japanese	1:0
7	Chaudhari AB[4]	1984	23 ^	Caucasian [c]	N.A
8	Wong TT et al. [20]	1986	8	Taiwan/Chinese [a]	3:5
9	Parízek J et al. [17]	1989	13	Czechoslovakia [a]	2:1
10	Oliveira HA et al. [15]	1989	1	Brazil [b]	NA
11	Hayath S et al. [10]	1989		[a]	
12	Peter JC et al. [18]	1992	35	Britain 31 African [d] 2 Caucasian 2 *Indian*	4:3
13	Martínez-Lage Sánchez JF et al. [12]	1992	5^	Spain [a]	N.A
14	Tan EC et al. [19]	1993	5 (4/1) ^	Japan [b]	3(1):1^
15	Isozumi T et al. [11]	1995	1	Japanese [a]	1:0
16	de Carvalho GT et al. [7]	2001	4/3 ^	Brazil 4 Caucasian 3 Afro Brazalian [c]	4:3 ^
17	de Aquino HB et al. [6]	2003	3	Brazil [b]	2:1
18	Aslan O et al. [3]	2004	4	Turkey [a]	4:0
19	Asani MO et al. [2]	2005	1	African/Nigeria [c]	1:0

[O]- Only 14 cysts had histological confirmation. One cyst was an encephalocoele
**- cyst contained clear fluid*
***Italics**- cases reported in children of Indian origin*
^ - Epidermoid cysts included in study.
[#] -Adult patients
[a] -dermoid cyst over anterior fontanelle
[b] -Congenital inclusion cyst of the subgaleal space
[c] -Congenital inclusion cyst of the subgaleal space
[d] -Congenital bregmatic cysts / Bregmatic inclusion cysts / Bregmatic dermoid cysts

association has not been found (16). The anterior angle of the anterior fontanelle has been described as the most common site of the lesion (16).

These cysts clinically are freely mobile (as in both our cases), usually trans- illuminate and may appear to increase on crying (1,13,16,18). Clinically, therefore, these are often confused with a dysraphic meningoencephalocoele (1,16,18,20). Unlike dermoid cysts in relation to other CNS location, those overlying the AF do not demonstrate any associated dermal sinus (3, 18). Both patients had clear cyst contents and trans-illumination was positive clinically (Figure 3B). The differential diagnosis should include; Hemangiomas, hamartomas, lipomas, encephalocoeles and epidermoids (1,18,20). There have been few reports of epidermoid cysts over the anterior fontanelle but are extremely rare (3) and said to be so in view of the embryogical differences with the dermoid cyst (3). Sebaceous cyst, lipoma, cephalhematoma, lymphangioma, hemangioma and sinus pericranii are said to be the other differential diagnosis (3).

Radiological diagnosis in earlier times ranged from puncture and aspiration, air cisternography and ventriculography to pneumoencephalograpy (1,3) to computerized tomography (C.T) and magnetic resonance imaging (MRI). Although benign these cysts are associated with bony erosion or flattening of the calvarium (3,20). This was also seen in our second case. Figure 2C, D and E demonstrates the asymmetrically widened anterior fontanelle. In the second patient, an MR venogram (MRV) delineated the non-involvement of the superior sagittal sinus (Figure 2F). It also delineates the anatomy to prevent necessitating burr holes, circumferential excision and exploration of the lesion up to the superior sagittal sinus, as has been described (16). Ruling out intracranial extension has been mentioned as an important consideration, although this may be seen in dermoids elsewhere on the scalp but none have been reported over the AF (16). Mid-sagittal or coronal MRI has been recommended to rule out intracranial extension of the lesion (11).

Although these lesions have no intracranial extension, they are usually treated by neurosurgeons because of the differential diagnoses in some of which an intracranial extension may be present and the proximity of the superior sagittal sinus (16, 17,20). The surgical excision is quite simple. The plane of these lesions is always sub-aponeurotic and approached with an elliptical, (1,16,18,20) or bicoronal incision (3,16). Blunt dissection is carried out and the lesions are excised in toto (1, 3,16). The redundant skin may be excised (18) as was done in both our cases. The only complications reported with surgery are dural tears and a post op leptomeningeal cyst formation (16). Neither of our cases had any complications.

Clear cyst fluid has been reported to be very rare in large cysts (5). Although in Adeloye et al's original report the smaller cysts had clear fluid (the exact number of patients is not mentioned) (1).

Histopathology always confirms the diagnosis (1-20). In both cases the diagnosis was clenched by histopathology. It has been stated that with increasing pressure the cyst lining elements may regress and may resemble an epidermoid cyst which is much rarer than dermoid cysts (17).

CONCLUSIONS

The Adeloye-Odeku disease, although well documented in the literature, is still rare in the Indian sub-continent. The review of literature revealed several interesting findings: The disease has been reported from virtually all races but continues to be most prevalent in the black African population, the earlier reports of female preponderance appear incorrect with many reports of male preponderance and the condition continues to be operated by neurosurgeons necessitating an increased familiarity in the neurosurgical community.

CONFLICT of INTEREST

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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