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CASE REPORT

Adeloye – Odeku Disease in Turkish children: A Case Report and a Short Literature Review

ABSTRACT

Congenital dermoid inclusion cysts located in the anterior fontanelle were defined as Adeloye-Odeku Disease by Adeloye and Odeku in 1971. The epidemiological characteristics of this rare disease are still unclear. These cysts are subgaleal benign lesions, and the main treatment is surgical excision. Radiological imaging is important in diagnosis and surgical planning due to their location. Although adult cases have been reported in the literature, Adeloye-Odeku Disease is primarily a subject of pediatric neurosurgery. In this study, we present a 10-month-old case of an anterior fontanel dermoid cyst and conduct the first national review of the disease with 9 cases reported from our country.

Keywords: Anterior fontanelle, congenital, dermoid cyst, pediatric

Dermoid cysts arise from the cystic enlargement of ectodermal inclusions in the neural tube during the 3rd-5th weeks of fetal life (1). Craniospinal dermoid cysts were first described by Cruveilhier in 1892. Adeloye and Odeku introduced anterior fontanel dermoid cysts as Adeloye-Odeku Disease in 1971 by publishing a case series of 18 Nigerian patients (2). Although it was thought to be an African cyst for many years due to this case series, cases from different nations have been reported subsequently (3-5). Anterior fontanel dermoid inclusion cyst and bregmatic dermoid cyst are other names for this disease. Congenital anterior fontanel dermoid cysts, which constitute 0.1-0.5% of childhood brain and skull tumors, are very rare (5). Although it is a very rare disease, they are significant lesions from a surgical perspective because of features such as their midline location, being congenital cranial lesions, and their adjacency to the superior sagittal sinus and cerebrum (6,7). The curative treatment for Adeloye-Odeku Disease is always surgical excision. Adjuvant treatment is not required because they are benign lesions. No recurrence has been reported in the literature (8). In our literature review on Adeloye-Odeku Disease in Turkish children, we found that a total of 9 pediatric cases were reported in 5 different articles from our country (5,7,9-11). Since it is not possible to obtain large case series for a disease with such a low incidence, case reports are valuable for providing the necessary accumulation in the literature and understanding rare diseases. In this study, we aim to contribute to the literature by presenting a pediatric case of Adeloye-Odeku Disease that was successfully treated in our clinic. Additionally, this report is the first review of 9 case reports originating from Türkiye.

CASE

The parents brought a 10-month-old girl with no significant medical history to our clinic with a complaint of a palpable swelling in the frontal midline of the head. The patient's parents stated that they noticed the swelling when the patient was only 2 months old and that it increased in size over time. She had no neurological deficit, and growth and development were compatible with her age. On physical examination, we detected a mass of approximately 3x4x4 cm, located on the anterior fontanel, covered with normal skin and partially fluctuant (Figure 1a and 1b). The transillumination test was positive (Figure 2). Laboratory tests were normal. Computed tomography (CT) showed a cystic lesion on the anterior fontanel without bone erosion or depression (Figure 3a, 3b, and 3c). T1 and T2 weighted (T1W, T2W) magnetic resonance

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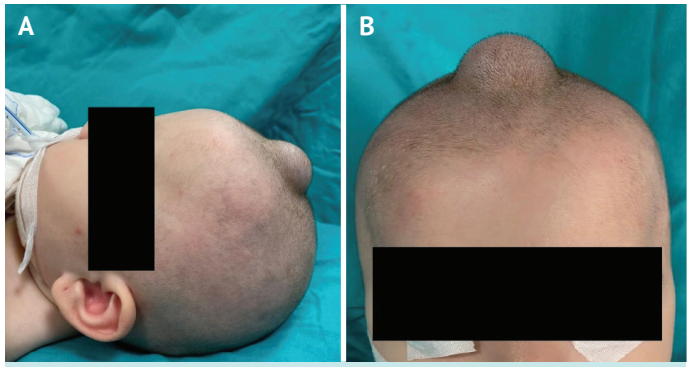

Figure 1a and 1b. Morphology of the cycst.

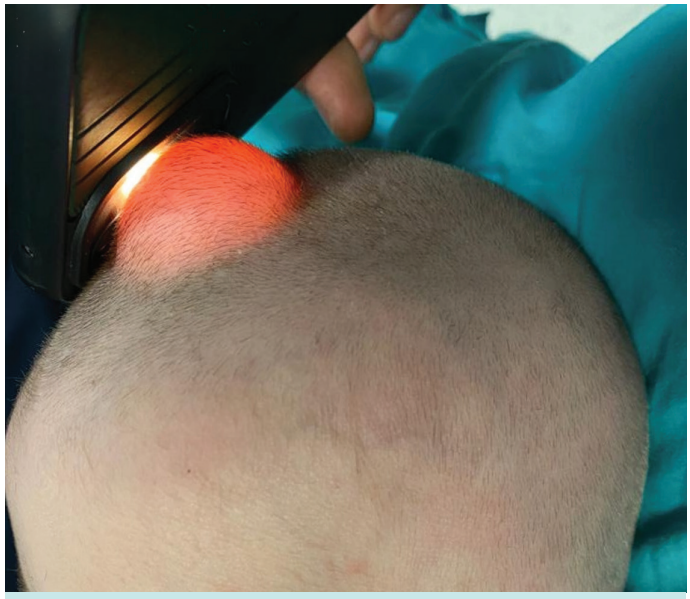

Figure 2. Transillumination test (positive).

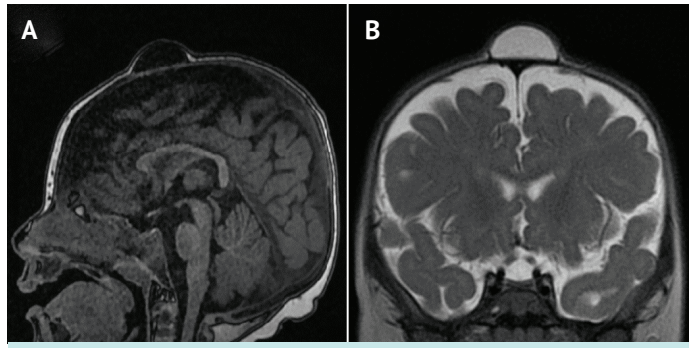

Figure 4. Magnetic resonance images of the cycst. (A) is sagittal view of T1W and (B) is coronal view of T2W images.

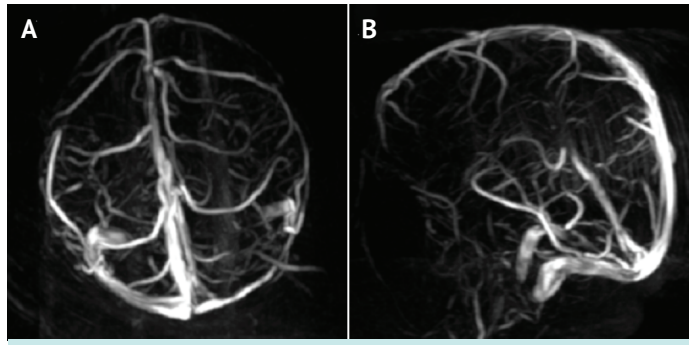

Figure 5. MR venography. Note that there is no irregularity or invasion of the superior sagittal sinus in neither axial (A) nor sagittal (B) view.

imaging (MRI) revealed a cystic lesion isointense with cerebrospinal fluid, located extracranially and without intracranial extension (Figure 4a and 4b). Magnetic resonance venography study was successful in demonstrating that the cyst was not associated with the superior sagittal sinus (Figure 5a and 5b).

Surgical technique

In the operating room, under general anesthesia, we made a coronal-oriented linear incision of approximately 5 cm, passing through

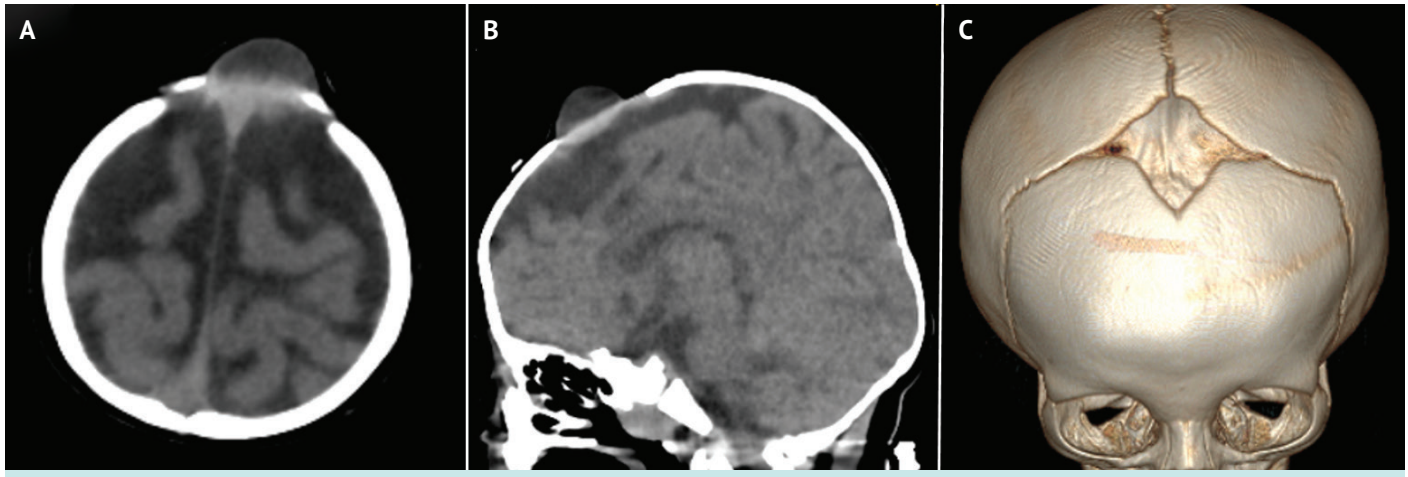

Figure 3. CT images of the anterior fontanelle dermoid cycsr. (A) is axial, (B) is coronal image. (C) ise 3D reconsruction of CT scan. Note that there is no bone flattening or erosion.

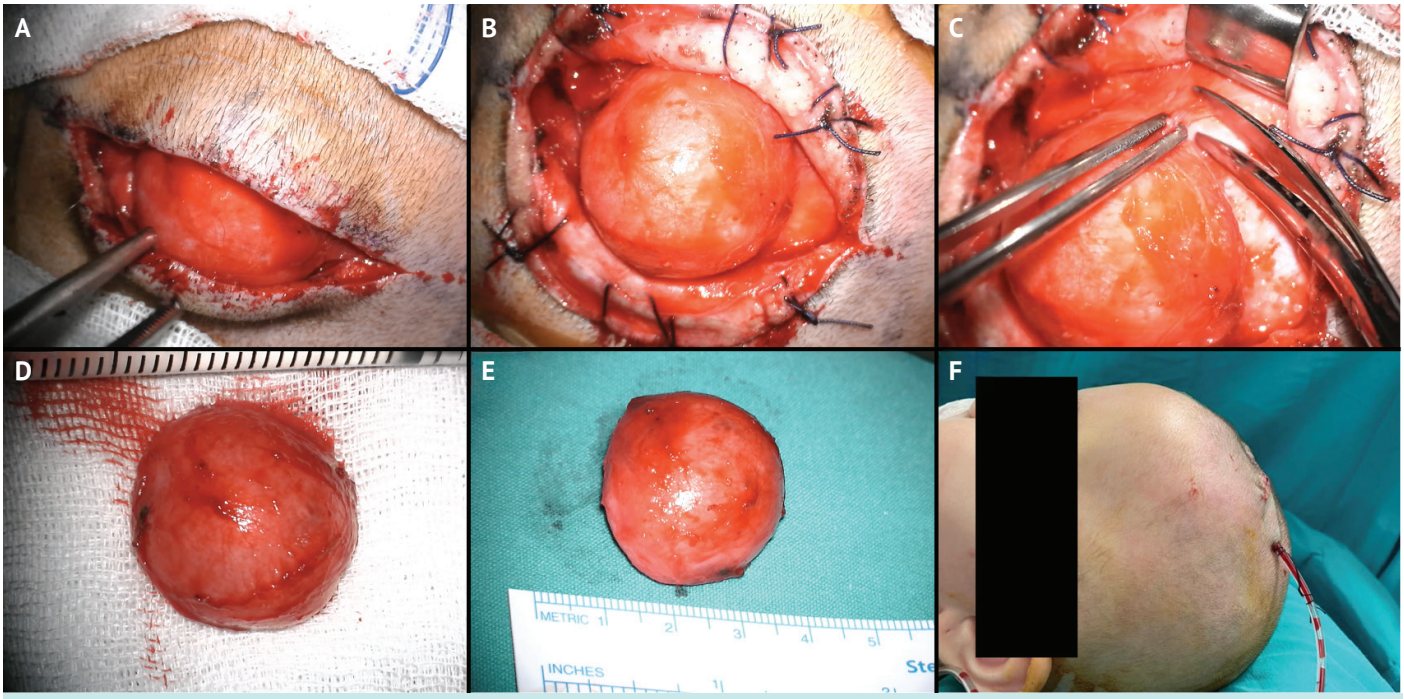

Figure 6. Intraoperative images. Note that the aponeurosis covering the cyst is thinned and adhered to the cyst wall in the Figure 6B. Figure 6C shows how we cut the dural aponeurosis from the neck of the cyst without dissecting it.

Table 1. This table summarizes the morphological and clinical features of cases reported from Turkey

Case	Age	Sex	Size	TI	Fibrous pedicle	Sinus invasion	Intracranial extension	Bone flattening	Bon erosion	Recurrence
İzgi et al.	16 years	Male	2x5x5 cm	-	-	-	-	-	+	-
Berkman et al. (1)	6 months	Male	2x2x2 cm	-	+	-	-	-	-	-
Berkman et al. (2)	8 months	Male	3x3x3 cm	-	+	-	-	-	-	-
Aslan et al. (1)	6 months	Male	3x3x3 cm	-	-	-	-	+	+	-
Aslan et al. (2)	5 years	Male	4x4x4 cm	-	-	-	-	+	+	-
Aslan et al. (3)	3 years	Male	3x3x3 cm	-	-	-	-	+	+	-
Aslan et al. (4)	5 years	Male	3x3x3 cm	-	-	-	-	+	+	-
Yılmaz et al.	11 years	Female	4x5x5 cm	-	+	-	-	+	+	-
Genç et al.	4 months	Female	4x5x5 cm	+	-	-	-	-	-	-
Emrahoğlu et al.	10 months	Female	3x3x3 cm	+	-	-	-	-	-	-

TI: Transillumition test

the apex of the cyst and including the skin and subcutaneous tissue, with the patient in a supine and head-neutral position (Figure 6a). The cyst was located subgaleally. The galeal aponeurosis covering the cyst was thinned and adhered to the cyst. To facilitate dissection and provide hemostasis, we preferred to traction the skin with silk sutures (Figure 6b). To avoid the risk of cyst rupture, we cut the aponeurosis overlying the cyst from the neck of the cyst without dissecting it using a surgical microscope (Figure 6c). Since the cyst was attached to the external dura layer on the ventral surface, sharp dissection was effective in preserving the superior sagittal sinus, and we were able to remove the cyst en bloc with this technique (Figure 6d and 6e). Finally, the excess skin was reconstructed and closed with 4.0 subdermal nylon (Figure 6f). The patient was taken to the ward in the early postoperative period. We did not detect any abnormalities in the physical examination, laboratory tests, and postoperative imaging. On the postoperative first day, 5 cc of serohemorrhagic fluid was present in the drain. We removed the drain and discharged the patient. The pathology examination results confirmed the dermoid cyst. In the postoperative 3-month follow-up of the patient, cosmetic improvement was excellent, and there was no recurrence.

DISCUSSION

Anterior fontanel dermoid cysts fall into the category of congenital inclusion cysts according to the new classification. They are theorized to form from the cystic dilatation of ectodermal remnants

in the neural tube and are situated under the galeal aponeurosis (12). This condition, first identified as Adeloye-Odeku Disease in the Nigerian population, seemed unusual in non-black races when considering early literature. However, subsequent publications have indicated that this rare disease is not exclusive to the black race (13). The literature includes 9 pediatric and 1 adult Turkish cases of Adeloye-Odeku Disease. İzgi et al. (10) reported the first of these in 1989. The characteristics of cases reported from our country are summarized in Table 1.

In international literature reviews, consensus on the disease's gender distribution is lacking, with inconsistent data (14). Nonetheless, a higher prevalence in females is often reported (8). Including the case we present, the female-to-male ratio among the 10 pediatric cases reported in our country is 3:7, contradicting the notion that the disease is more common in females.

Anterior fontanel dermoid cysts are present from birth, growing slowly due to content increase (13). They are typically noticed by parents as a swelling on the forehead in early infancy (5). These cysts are covered with normal skin and can be fluctuant or entirely solid, but not pulsatile. A negative transillumination test supports a dermoid cyst diagnosis (15). The cysts are painless, and manipulation does not trigger pain (4,7). In the case we presented, the patient's parents reported that the lesion enlarged over months. The swelling was fluctuant, immobile, non-pulsatile, and painless upon palpation. Other cases reported from our country similarly showed no pulsation or pain, with cyst fluctuation noted in 6 out of 9 cases. Contrary to general belief, our case had a positive transillumination test, aligning with a few rare instances in the literature (15). Among the 9 cases reported from Türkiye, only Genç et al. (9) noted a positive transillumination test in the 4-month-old patient they presented. The lesions in both our case (3x3x3 cm) and the other 9 cases reviewed (mean 3.1x3.7x3.7; min: 3 – max: 5 cm) vary in size from 1 to 7 cm, as documented in the literature (16). Literature suggests lesion size correlates with patient age, but our review's limited case number precludes testing this hypothesis (17).

Physical examination and radiological imaging are key diagnostic elements. X-ray imaging, ultrasonography (USG), CT, and MRI can aid in diagnosing Adeloye-Odeku Disease. Coronal and lateral radiographs can reveal whether the lesion is cystic and if any bone irregularities are present. USG can detail the cyst's morphology and, if the anterior fontanelle remains open, its intracranial extension and superior sagittal sinus invasion (18,19). CT scans effectively show the lesion's relationship with bone structures, with coronal and sagittal plane images almost perfectly displaying a hypodense extracranial cyst without calcifications. Some cases have reported external table erosion or flattening. Dermoid cysts typically appear hyperintense on T2W images and hypointense on T1W images in MRI scans. MRI excellently delineates the cyst's intracranial extension. Given the lesion's location, MR venography is invaluable for assessing its proximity to the superior sagittal sinus, a crucial surgical planning aspect (7,20,21). Differential diagnosis for Adeloye-Odeku Disease includes conditions like encephalocele, meningocele, sebaceous cyst, lipoma, cephalohematoma, hemangioma, lymphangioma, sinus pericranii, and meningioma, making radiological imaging techniques essential for accurate diagnosis (22,23). The case we present is our clinic's first operated case, utilizing both CT and MRI for diagnostic and surgical planning. The

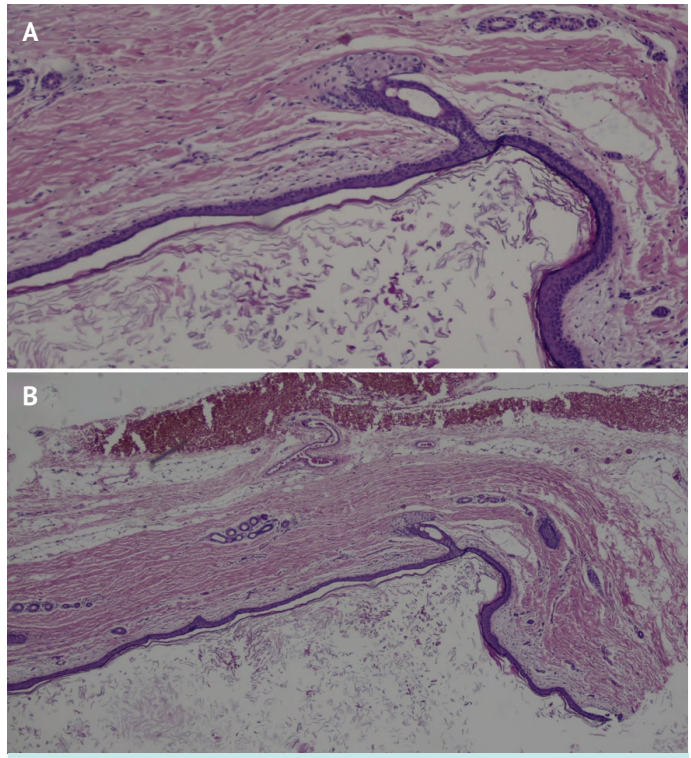

Figure 7. In the preparations stained with hematoxylin-eosin, a cystic lesion lined with stratified squamous epithelium and keratin lamellae are noted (7a x100, 7b x40).

lesion's radiological characteristics matched those in the literature, with no observed bone erosion or flattening. MR venography was particularly useful for surgical planning to assess the lesion's superior sagittal sinus invasion status. Bone structure erosion was noted in 6 of the 9 cases reported from our country, with 5 showing bone depression (5,7). None of the cases had superior sagittal sinus invasion. Despite CT's X-ray exposure drawback and MRI's limited resources, we recommend both for patients due to the surgical significance of the lesion's anatomical location and the prevalence of midline anomalies in the pediatric age group.

Anterior fontanel dermoid cysts are benign; the treatment is always surgical excision. Adjuvant therapy is not required after surgery (9). Surgical excision is performed for cosmetic reasons, to prevent infectious complications, and to confirm the diagnosis histopathologically. To avoid potential infectious spread and aseptic meningitis during surgery, it is crucial to maintain the cyst's integrity and protect the superior sagittal sinus (6). In the surgical management of the patient we presented, since the galeal aponeurosis covering the cyst was thinned and adhered to the cyst wall, we chose to remove the cyst along with the aponeurosis to prevent cyst rupture and shorten the procedure's duration. We believe this approach is safer and more straightforward when dissection of the aponeurosis and cyst is challenging. Additionally, while both elliptoid and linear coronal incisions have been suggested in the literature, we opted for a linear coronal incision. We reasoned that tissue loss from the first incision would be unsuitable for reconstructing the remaining skin after cyst removal. In reviewing cases from our country, we found that a linear incision was preferred in 7 of 9 cases and an elliptoid incision in 2, with no significant difference in outcomes.

Moreover, in 3 of the reviewed cases, the authors noted a fibrous pedicle connecting the cyst to the external dural layer (7,11). Our case did not have such a stalk. However, due to the adhesion between the external dural layer and the cyst, we deemed blunt dissection risky for superior sagittal sinus penetration and thus preferred sharp dissection.

The diagnosis of a dermoid cyst is confirmed by histopathological examination, identifying a cyst lined with stratified squamous epithelium and skin appendages such as keratin lamellae, sebaceous glands, and sweat glands (24). In our case's histopathology preparations, a cyst lined with stratified squamous epithelium and keratin lamellae was observed (Figure 7).

Adeloye-Odeku Disease is benign, with no additional treatment required post-surgery (1,4,5). No recurrence was reported in any case from Türkiye, including the case we presented, aligning with the global literature.

CONCLUSION

Adeloye-Odeku Disease denotes congenital dermoid inclusion cysts located in the anterior fontanelle. The condition is quite rare, leaving its epidemiological characteristics yet to be fully elucidated. In the case we presented, we highlighted the efficacy of a linear skin incision in surgical technique and the unnecessary dissection of the galeal aponeurosis and cyst due to the risk of rupture and procedure prolongation. Through this study, we aimed to add a case to the literature on Adeloye-Odeku Disease, a rare condition. Additionally, this study represents the first review discussing cases reported from Türkiye within the literature context.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.E.E., E.R.Y.; Design – H.D., M.E.E.; Supervision – E.R.Y.; Analysis and/or Interpretation – M.E.E., E.R.Y.; Literature Search – M.E.E., H.D.; Writing – M.E.E., E.R.Y.; Critical Review – E.R.Y., M.E.E., H.D.

Conflict of Interest: The authors declare that they have no conflict of interest.

Ethical Approval: This article does not contain any studies with human participants or animals performed by any of the authors.

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