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Adeloye-Odeku Disease in Irrua, South-South Nigeria. The experience so far in a rural neurosurgical setting

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ABSTRACT

First described in a publication by two Nigerian Neurosurgeons, Adeloye A and Odeku EL, in 1971, Adeloye-Odeku disease is a solitary congenital subgaleal inclusion dermoid cyst of the anterior fontanelle. This rare lesion, which makes up about 0.1-0.5% of all cranial tumours and 0.2% of all inclusion cysts, was initially thought to be found only in Africans. However, further reports have shown it to have a universal occurrence, as it has been reported in Caucasians, Chinese, Indians, and other part of the world. This lesion is also known as Congenital inclusion dermoid cyst (CIDS), is a benign slow-growing lesion, and if untreated, may persist to adult life.

This article gives a highlight of the disease and its management and goes further to report 3 cases of this rare benign lesion seen in Irrua, South-South Nigeria, a rural, low-resource tertiary health institution.

Incidentally and interestingly, all three cases presented within three consecutive months (January-March, 2019) at the neurosurgery outpatient clinic. Being uncomplicated cases, private and group counselling was done. The parents of the patients were much more reassured and relieved from their anxieties seeing others with similar problem. They were all worked up for surgery at different dates, had excision of the cysts with no complication and are currently being followed at the outpatient clinic.

INTRODUCTION

Adeloye-Odeku disease, also known as congenital inclusion dermoid cyst (CIDC), is a solitary congenital subgaleal inclusion dermoid cyst of the anterior fontanelle. It is named after Adeloye A and Odeku EL, who first reported it in 1971, presenting series of 18 patients seen at the University College Hospital, Ibadan. They also published a full description of the cyst and its management.¹

Keywords

Adeloye-Odeku,
 congenital,
 inclusion dermoid,
 Cyst,
 anterior fontanelle



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CIDC is an inclusion dermoid, a developmental anomaly in which displaced dermal elements are included in the neuroaxis along the embryonic fusion line. In this case, it occurs under the epicranial aponeurosis over the anterior fontanelle. Surgical excision, preferably enucleation, is curative for this disease, and no recurrence has been reported so far.

EPIDEMIOLOGY

Following its first report from patients in Nigeria in 1971, it was initially thought to be an African disease.² However, subsequent publications from various parts of the world have shown the cyst to be multiracial and to have a universal occurrence.^{3,4,5,6} The cyst accounts for 0.1 -0.5% of all cranial tumours⁶ and 0.2% of all inclusion cysts.⁷

Although present at birth, its initial small size draws little or no attention. Hence, most cases are seen after the age of 3 months. If left untreated, it may persist till adult life as already reported.^{8,9}

AETIOPATHOGENESIS

The disease is caused by a developmental anomaly in which displaced dermal elements are included in the neuroaxis along the embryonic fusion line around the epicranial aponeurosis over the anterior fontanelle. This forms a slow-growing inclusion dermoid with no intracranial connection.

The cyst has a fibrous tissue wall lined by stratified squamous epithelium and it contains mainly a clear and colourless fluid with striking resemblance with cerebrospinal fluid (CSF) but with much smaller quantity of sugar and protein. The fluid may be mixed with cholesterol crystal, sebaceous materials and desquamated epithelial cells. These may form clumps or floccules floating over the surface of the fluid or line the inner wall, giving it a rather rough and shaggy appearance.¹⁰

CLINICAL FEATURES

The history is usually that of a slow-growing swelling over the anterior fontanelle present since birth.

The swelling is oval, spherical, non-tender, fluctuant, with intact overlying skin. It transilluminates brilliantly but does not show transmitted cough impulse due to no intracranial communication.

The size of the cyst varies with the age of the patient at the time of the diagnosis.⁴ However, a range of 1-7cm have been recorded.⁶ The lesion is

loosely attached to the overlying aponeurosis and underlying pericranium and the absence of cough impulse differentiates it from a meningocele or encephalocele.

DIFFERENTIAL DIAGNOSES

The differential diagnoses of this condition include encephalocele, meningocele, sebaceous cyst, lipoma, haemangioma, cephalohaematoma. However, CIDC is differentiated from the above by its clinical features and this is corroborated by the findings of appropriate radiological and laboratory investigations employed.

MANAGEMENT

Usually, the patient is stable with an uncomplicated lesion, presenting at the clinic. A good history is taken and clinical examination done and all findings documented. Necessary radiological and laboratory investigations are carried out.

Counseling is done and this should cover the pathology, its cause and course, investigations required, treatment options and possible complications. Informed consent is obtained appropriately and the patient is worked up for excision thereafter. The cyst is excised completely and sent for histology so as to get a confirmatory tissue diagnosis. Patient is followed up appropriately.

INVESTIGATIONS

Radiological investigations

To identify the location, size, nature and possible contents of the cyst and find out if there is any intracranial communication. They also serve as a guide for the surgeon intra-operatively. They include:

Transfontanelle Ultrasound Scan /Ultrasound Scan of the mass

This is much used for this condition in low economic regions where there is no Computed Tomography (CT)/ Magnetic Resonance Imaging (MRI) scan or, even if present, patient cannot afford them as seen in Irua. The scan usually reveals a cystic mass with homogenous or heterogenous echogenicity, well circumscribed, and lying over the anterior fontanelle with no intracranial extension.

Computed Tomography (CT)/ Magnetic Resonance Imaging (MRI) scan

These outline the lesion and confirm the cyst. Usually, a well-defined extracranial, subcutaneous fluid-density cyst over the anterior fontanelle is seen. It also gives the opportunity to visualize intracranial contents and usually there is no intracranial extension.

Body fluid investigations

Necessary for preoperative work up to assess patient's fitness for surgery include: Full blood count, Serum electrolyte, urea, creatinine, and random blood glucose/ fasting blood glucose (especially for diabetics).

Histology and Cyst fluid analysis

This is done post excision of the cyst. It confirms the diagnosis and rules out malignancy. The usual finding is that of cyst wall connective tissue lined by stratified squamous epithelium while the cyst fluid is clear with varying amount of glucose, protein, potassium, sodium, urea and LDH. Some cysts will contain sebaceous glands, hair follicles, etc, in the adnexial layer.

TREATMENT

Surgery is the mainstay of treatment. This involve enucleation of the lesion through a transverse scalp incision, usually under general anaesthesia, is curative for the disease. Care is taken to avoid injury to the underlying dura if fontanelle is still patent, and the cyst is delivered without capsular rupture. Redundant scalp may be excised and the wound closed appropriately, bearing in mind the need to achieve satisfactory aesthetic outcome.

COMPLICATIONS

These are usually rare. However, some of the possible complications include:

Cyst rupture and secondary infection if left untreated. Dura breach with resultant CSF leak as a complication of surgery. There is however no report of such in the literatures reviewed. Surgical site infections may be seen.

THE IRRUA EXPERIENCE SO FAR (CASE SERIES)

CASE I

A 4-month old male infant who was brought to the outpatient clinic with a painless, progressive scalp

swelling over the anterior fontanelle which has been present since birth. No associated skin changes or discharge. No neurological symptoms or deficits. No swellings in other parts of the body. Developmental milestones were achieved at the appropriate time for age and patient was up to date with the immunization schedule. No family history of such swellings. Pregnancy, birth and neonatal history were uneventful and mother was delivered via a spontaneous vaginal delivery. Birth weight is 2.8kg. Examination revealed a 6cm diameter hemispherical mass over the anterior fontanelle which was cystic, non-tender, smooth, and non-pulsatile. There was no differential warmth and no cough impulse. It was loosely attached to the overlying skin and underlying pericranium and transilluminates well.

Other examinations revealed normal findings.

Transfontanelle ultrasound done revealed a well circumscribed cystic mass overlying the anterior fontanelle, containing dependent echogenic debris. It measures 8.76cm³. Underlying brain is normal and there is no intracranial extension of the mass.

A diagnosis of Adeloye-Odeku disease was made and the parents of the patient were counseled for surgery. Informed consent was obtained and preoperative work up done. Routine blood investigations were normal.

He had enucleation of the cyst via a transverse scalp incision under general anaesthesia. Intra-operative finding was a 6cm diameter oval cystic mass underneath the aponeurotic layer of the scalp over the anterior fontanelle. The tissue was sent for histology. Wound was closed with absorbable vicryl and patient did well postoperatively. He was discharged on the 2nd day post-operative and followed up in the outpatient clinic.

Histology report revealed an ovoid cystic mass with shining grey-brown capsule measuring 3x2x2cm on macroscopy. Cut surface revealed a cystic cavity containing clear-white jelly substance. Microscopically, the sections showed a benign cystic lesion, containing keratinous debris and lined by keratinized stratified squamous epithelium with focal areas of attenuation. The wall is composed of fibrocollagenous stromal element with several skin adnexial structures. A histologic diagnosis of Scalp mass: Dermoid cyst, was made.

The patient is currently being followed up on outpatient basis and is doing fine with no complication or recurrence so far.

CASE II

A 22-month old female child with progressive painless scalp swelling over the anterior fontanelle present since birth. No associated neurological deficits. No skin changes or discharge for the swelling. No other swelling in any other part of the body. Pregnancy, birth and neonatal history were uneventful. Developmental milestone was attained at appropriate time and the child will be properly immunized for age.

Examination reveals a 4cm diameter spherical swelling over the anterior fontanelle which was non-tender, cystic, with no differential warmth. It was loosely attached to the underlying structure and overlying skin and transilluminates. Other examinations were normal. A clinical diagnosis of sub galeal dermoid cyst was made.

Transfontanelle USS done revealed a cystic sub periosteal lesion measuring 19.9 X 19.2 X 78mm located in the midline between the outer and inner table of the frontal bone. There is no demonstrable communication with the CSF or brain meninges. Both lateral, third and fourth ventricle are normal in size and outline. No sonographic evidence of intraventricular hemorrhage, mass or hydrocephalus. The brain parenchyma appears normal sonographically with normal grey and white matter differentiation. The parents were counseled for surgery and informed consent was obtained. Pre-operative investigation was normal. She subsequently had excision biopsy done under general anesthesia via a transverse scalp incision.

The intra-operative finding was that of 3cm diameter ovoid cyst in the subgaleal layer of the scalp. The sample sent for histology and the wound closed with absorbable vicryl, both deep and subcuticular layer. Post-operative condition was satisfactory and patient was discharged on the second day post-op.

Histology report revealed: macroscopically, an encapsulated cystic mass that is grayish-white, weighing less than 50g, measuring 1.9 X 1.5 X 1.0cm. Sections showed a cystic lesion lined by stratified squamous epithelium with lamellated keratin in the cavity. The lining epithelium is in connection with a pilosebaceous unit in some areas while in the other area by sebaceous gland. The wall is extensively fibrocollagenous with collagenization in some areas and fibrosis and hemorrhage in other areas.

Histological diagnosis of scalp mass-epithelial inclusion cyst was made. No evidence of neoplasm. The patient is currently being followed up on outpatient basis with no complication or recurrence so far.

CASE III

A 7-month old female infant with a small painless scalp swelling on the anterior fontanelle noticed since birth. The swelling was progressively increasing in the size, though slowly. No associated overlying ulcer or discharge. No swelling in other parts of the body and no family history of such swelling. No neurological deficit. The pregnancy, birth and neonatal history were normal. No delayed developmental milestones and she was up to date with the immunization schedule.

Examination revealed a localized spherical swelling on the anterior fontanelle of about 5cm diameter. It was non-tender, well defined, and cystic, with no differential warmth. Not attached to the overlying skin and transilluminates. Other findings were grossly normal. A clinical diagnosis of Adeloeye-Odeku disease was made.

Transfontanelle USS requested revealed a thick-walled echogenic collection with amorphous intralesional solid components that appear villiform, overlying the anterior fontanelle. The collection measures about 23.0cm³. There is no communication between the collection and the intracranial structures. Intracranial structures appear grossly normal.

The parents were counseled for surgery (excision biopsy) and with informed consent obtained, she was worked up for surgery. Preoperative investigations were normal. She had enucleation of the cyst via a transverse scalp incision under general anaesthesia and the intra-operative findings was a 4cm diameter cystic mass underlying the aponeurosis over the anterior fontanelle and overlying the dura. The tissue was sent for histology and wound closed with absorbable vicryl and dressed appropriately. The post-operative condition of the patient was satisfactory and she was discharged on the second day post op.

The histology report revealed: macroscopically, a spherical grayish-white cystic tissue measuring 3x2x2cm. Cut surface revealed a colourless fluid, a cystic wall coated with whitish friable material with interspersed shaft. Microscopically, histologic

sections showed a benign cystic lesion which contains keratin debris and surrounded by a wall made up of fibrocollagenous with collagenization in some areas and fibrosis and hemorrhage in other areas.

A histological diagnosis of anterior fontanelle swelling: Benign inclusion cystic was made.

The patient is currently being followed up on outpatient basis and is doing well with no complications or recurrence so far.



FIGURE 1. Preoperative photograph of Case II



FIGURE 2. Intraoperative, showing lesion being excised



FIGURE 3. Post excision



FIGURE 4. Just after wound closure



FIGURE 5. At first follow up visit post op

SUMMARY OF CASES

Three cases of this rare lesion were seen within three consecutive months and they involved two females and a male child, all within the age range of 4-22 months.

The scalp swellings, which had been present since birth in all cases, were of sizes ranging between 4-6 cm diameters, located over the anterior fontanelle, non-tender, cystic, smooth, transilluminant, loosely attached to the underlying structure and overlying skin, with no positive cough impulse. None of them was ulcerated and there was no discharge from any. Clinical diagnoses of Adeloje-Odeku disease was made in all three cases and they had radiological investigations done to properly further define the lesions, especially their location, size, content, and if they communicate with the intracranial structures.

With no intracranial communications confirmed, the parents were counseled and informed consent obtained. Thereafter, all three patients had enucleation of the lesions via a transverse scalp incision under general anaesthesia. There were no intra or post-operative complications.

Histology reports of these lesions gave confirmatory tissue diagnosis of dermoid cyst, epithelial inclusion cyst, and benign cystic teratoma respectively. All were in keeping with Adeloje-Odeku disease-congenital inclusion dermoid cyst over the anterior fontanelle. All three patients are currently being followed up with no complications or recurrence recorded so far.

DISCUSSION

Adeloje-Odeku disease still remains a rare lesion though reports have shown its universality in occurrence and its multiracial prevalence^{1,2,3,4,5,6,7,8,9,11,12,13,14} as against the initial thought of it being an African disease^{1,2,3,4,5,6,7,8,9}. So far, over 229 cases of this condition has been reported in literature until 2003.⁴

Nevertheless, a report by Dadlani *et al* suggest that its occurrence may be underreported and under-calculated due to observed non-uniformity in the nomenclature of the disease in various medical literatures and its management, in some cases, by other specialties (Paediatric surgeons, general and plastic surgeons).¹¹ Our report may give some credibility to the above as the 3 cases were seen within 3 months in a nascent neurological unit, just in less than 2 years of its existence.

Currently, the lesion accounts for 0.1-0.5% of all cranial tumours and 0.2% of all inclusion cysts.^{6,7}

Aesthetic appearance and anxiety over the cause of the swelling are the main reasons for seeking surgical intervention by parents.¹² This was clearly evident in the response given by the parents of the children in this index report during consultation. All three couples saw the lesion as a form of deformity and were worried about what could be therein.

All three lesions in our report were present since birth, slow growing and with sizes ranging from 4-6 cm, in keeping with the range reported in other studies.⁷

Our report involved 2 females and a male child, though no sex predilection has been established in this disease. While some reports have male preponderance^{2,3,6,7} others reveal the disease to be more in females.^{3,14}

The three cases in this report have the classical clinical features of the disease in keeping with other reported cases. There was, however, a major challenge in the radiological investigations to confirm the nature and extent of the lesion. In our report, transfontanelle ultrasound scan was used in place of CT and MRI scan due to financial constraints on the part of the parents of the patients who unfortunately have no health insurance. Nevertheless, we were able to establish the location, size and nature of the cyst and non-communication with intracranial structures via the USS done.

All three patients had excision of the cysts (enucleation) under general anaesthesia with no complications. The histology of the cysts in all 3 cases in our report showed benign cystic lesions lined by stratified squamous epithelium and containing skin appendages like hair follicles, sweat and sebaceous glands in the adnexial layers. This is in keeping with other reported cases.^{5,6,9}

Two of our reported cases (1st and 3rd) revealed the content of the cyst to be clear, colourless fluid consistent with published cases^{5,15,16,17}. However, further biochemical evaluation of its contents could not be carried out due to the technical limitations of our facility.

Nonetheless, the histology reports confirm all three cases to be dermoid cysts and ruled out any malignancy. All three patients are currently being followed up with no complications or recurrence so far and with satisfactory aesthetic outcome.

CONCLUSION

Adeloye-Odeku disease, first reported in Nigeria, and currently shown to be universal, remains a rare benign lesion of interest to the neurosurgeon. Total surgical excision is curative of the disease as shown in all reports. These case series have added credence to the above and has shown its successful management in a rural, low-resource setting.

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