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ADELOYE-ODEKU DISEASE IN ASSER REGION OF SAUDI ARABIA

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Abstract

Adeloye-Odeku disease, also known as congenital inclusion dermoid cyst (CIDC) of the anterior fontanelle was first described by Adeloye A and Odeku EL, as a solitary congenital subgaleal inclusion dermoid cyst of the anterior fontanelle. This rare lesion was initially thought to be confined to Africans . The universal distribution of this cyst has subsequently been shown in accumulated literature of other ethnic distribution ,Caucasian, Indians, Chinese, Japanese, Turkish and others. This is the first published report that we know of Adeloye-Odeku disease in Arab-Saudi children in Abha, Saudi Arabia. Four patients with Adeloye-Odeku disease, two boys and two girls, Saudi patients with ages ranging from 3months to 9 years are presented. Successful surgical excision of the cyst was achieved in each of the three patients who consented for surgery with no recurrence. The oldest patient did not go for surgery due to parental refusal but the lesion proved to be stable on follow up.

Key Words: Adeloye-Odeku, inclusion dermoid cyst, Saudi, Congenital.

Introduction

Congenital inclusion dermoid cyst (CIDC) of the anterior fontanelle is a rare lesion; 0.1-0.5% of all cranial tumors (Aslan, 2004) and 0.2% of all inclusion cysts (Carvalho et al; 2001). It is noticed soon after birth in all affected patients and is usually a solitary lesion.

Adeloye and Odeku in 1971, were the first authors who published a full description and management of congenital subgaleal cysts over the anterior fontanelle in Nigerians and named it Adeloye-Odeku disease. This condition was initially thought to be an African cyst (Peter et al, 1992). Worldwide occurrence of congenital dermoid cyst of the anterior fontanelle has since been shown in publication (Parizek et al; 1989, Aquino; 2003, Tateshima et al; 2000, Kanamaru et al; 1984, Agrawal et al; 2007 and Majed et al; 2008).

CIDC is a developmental anomaly in which displaced dermal elements are included in the Neuroaxis along the embryonic fusion line. There is no communication between the cysts and the intracranial contents. The cyst is a solitary slow growing, non tender fluctuant mass that transilluminate and is covered with intact skin over the anterior fontanelle. The size of the cyst varied with the age of the patient at the time of presentation (Aquino 2003) and a measurement range of 1-7cm have been recorded (Aslan 2004). All patients had no associated neurological findings and surgical excision is curative. Other important differential pathologies are encephalocele, meningocele, sebaceous cyst , lipoma , haemangioma and cephallhematoma. Some authors however have reported inclusion cysts diagnosed for the first time in adults (Ojikutu and Mordi, 1980 & Castro et al; 2007).

Patients and Method

Between 2004 and 2010,consecutive patients with Adeloye-Odeku disease were prospectively recruited into this study involving Arab-Saudi patients. Four patients with non-tender swellings over the anterior fontanelle were seen in the neurosurgical outpatients department of Asir Central Hospital, Abha, the regional referral center of the southwest of the kingdom of Saudi Arabia.

The patients presentation is as follow:

(1) A one and half year old female Saudi patient was admitted from the outpatients' clinic because the parents wanted cosmetic correction of a small painless scalp swelling in the middle of her head that had failed to disappear since her birth. There was no other complain in this thriving baby girl. Routine blood test were normal. CT & MRI brain confirmed the cyst, 15x20mm in diameter with no intracranial extension. The patient underwent surgery under general

anesthesia, a complete excision of the cyst was achieved through a coronal scalp incision over the mass with blunt dissection of the cyst from the underlying tissues. This enabled the cyst with clear content to be delivered without capsular rupture. Care was taken to avoid injury to the dura and cranium. There was no intracranial extension of the cyst. Haemostasis was achieve and a continuous suction drain inserted. Layered closure of the incision was achieved with vicryl and silk to skin. The drain was removed after 2 days. Microscopically, the cyst wall was made up of connective tissue lined by squamous epithelium. In the subepithelial layer were adnexial appendages like sebaceous glands, sweat glands and hair follicles.

Fluid from the cyst had protein: 1mg/dl; Glucose: 1mg/dl; potassium: 20.4mmol/l; Sodium: 10mmol/1; Urea: 162mg/dl; LDH: 109u/l; Amylase 1 u/l. Patient recovered without any complication and the patient's parents were satisfied with the aesthetic result. Follow up in the outpatients' clinic in a 2-year period showed no recurrence.

(2) A three-month-old male Saudi was admitted for the removal of a small lump occupying the anterior fontanelle noticed after birth. The swelling seemed to be growing slowly with the child who had no other neurological problem. The dome shaped mass was non-tender and transilluminates light. Pre-operative brain CT scan showed a well-defined extra cranial, subcutaneous fluid-density cyst over the anterior fontanelle. The cyst measured 20x10mm in diameter. CT brain showed normal ventricles with no midline shift or deformity; normal posterior fossa; no areas of recent blood density and no extra collections were seen. MRI of the brain showed a well-defined oval shaped cystic swelling over the anterior fontanelle. It measured 15x10mm in diameter. It had a thin wall, no interval septum or solid masses and had fluid intensity. The rest of the brain was normal. In the operating room under general anesthesia, a complete excision of the cyst was achieved without damage to the underlying midline structures through a skin crease incision and blunt dissection. Haemostasis was achieved and a continuous suction drain left in the sutured wound for 2 days after which the drain was removed.

The cyst fluid had clear contents and had Protein: 0.5mg/dl; Glucose: 1mg/dl; Potassium: 15mmol/l; sodium: 10mmol/l; Urea: 100mg/dl; LDH: 100u/l; Amylase 1u/l. Histopathology of the cyst wall showed connective tissue lined by stratified squamous epithelium. There were sebaceous glands and hair follicles in the adnexial layer. The patient had an uneventful postoperative period and there was no recurrence during the 2 years follow up period.

(3) A one-year-old Saudi girl presented with a swelling over the anterior fontanelle, which was present at birth and did not seem to cause the child any problem but had failed to go away. CT scan brain confirmed a dermoid cyst, 15x20mm in diameter. This cyst was removed under general anesthesia in the operating room through a skin crease incision and careful blunt dissection that avoided Injury to the underlying dura mater. A continuous suction drain was left in the sutured wound for 2 days after which the drain was removed. The cyst fluid had clear contents and had Protein: 1mg/dl; Glucose: 1mg/dl; potassium: 20.0mmol/l; sodium: 5mmol/l; Urea: 150mg/dl; LDH: 110ul/l; Amylase: 1.2ul/l. Histopathology of the cyst wall showed connective tissue lined by stratified squamous epithelium. No recurrence has been shown in the outpatients follow up of 2 years.

(4) A nine-year-old boy presented with a small lump in the middle of the head. This was only noticed a month before presentation because of headaches. He had no fits or vomiting. There was no other neurological deficit. CT & MRI brain showed an extra cranial small well-defined cyst 20x20mm in the middle of the frontal bone abutting the saggital sinus but separate from the intracranial and the CSF spaces. The brain and the ventricles were normal. This atretic anterior fontanelle

cyst was suspected to be a dermoid cyst. Parents of this child declined surgical intervention but agreed to follow up in the outpatients' clinic for this asymptomatic small suspected dermoid cyst that did not grow over two years of follow up.

Discussion

Adeloye-Odeku disease is a rare lesion that has been shown to be worldwide and not African as was initially though (Castro 2007). The total number of published cases of congenital dermoid inclusion cyst of the anterior fontanelle in children worldwide in 1989 was 174; Americans were 74, Africans 45, Europeans 30 and others 25 (Parizek 1989). Many other authors have since added to that number in their publications.

A congenital dermoid inclusion cyst (CIDC), of the anterior fontanelle accounts for 0. 1-0.5% of all cranial tumors (Carvalho 2001) and 0.2% of all inclusion cysts (Aslan 2004). The lesion usually manifests at birth.

CT scan and magnetic resonance imaging (MRI) of the skull in our 4 patients confirmed these modalities as the most reliable and accurate in delineating the lesion as other authors had found (Carvalho 2001 & Agrawal et al; 2007). The indication for these investigation is the other differential pathologies and the medline position of this lesions that could lead to intracranial extension. In our patients CT scan did not add more information over the MRI. Given the young age of the patients that pose need for sedation during imaging and the radiation risk with CT scan; MRI alone is probably an adequate study with no need for further imaging.

The size of the lesion in our study was between 10-20mm. within the range of 10-70mm reported in other studies (Carvalho et al; 2001).

The histology of the cyst in all three cases in our study was the same as findings in published studies and showed squamous epithelium lining and skin appendages like hair follicles, sebaceous and sweat glands in the adnexial layer (Casro et al; 2007, Kanamaru and Waga 1984, Agrawa et al; 2007).

The three operated patients with Adeloye-Odeku disease in our study had clear colourless contents with low proteins, sugar and sodium chloride consistent with published cases (Tateshima et al; 2000 & Majed et al; 2008). Variation in the colour of the cyst fluid was found in other studies depending on the size and age of the lesion as well as the contained dermal elements (Castro et al; 2007 & Tateshima et al; 2000). Some authors have published cases of anterior fontanelle cyst in adults (Castro et al; 2007 & Ojikutu and Mordi 1980).

Aesthetics appearance of the patient is the main reason for seeking surgical intervention by parents. Other reasons are lesion growth, parental anxiety, unawareness, and fear of cancerous changes.

CDIC is a benign lesion that can be easily excised and cured with no recurrence. Excision of the cyst prevents possible rupture and infection and provides tissue for histological diagnosis and to rule out malignancy.

The 4 cases presented in this study have added Saudi patients to the worldwide published cases. There was no gender difference in our study. Surgical excision achieved an aesthetical satisfactory appearance and no neurological complication. No recurrence has occurred in the follow up period of two years of the operated patients. Histology confirmed dermoid cyst and no malignancy. Follow up in the outpatients clinic for the patient who did not have surgery showed the lesion to be stable.

Summary

Adeloyed –odeku disease also known as CDIC manifested soon after birth in three Saudi patients while the fourth patient's lesion was an accidental presentation at the age of 9 years. The lesion in this latter patient was small, stable in size, was hidden away behind the hairline and did not pose a problem to the boy. There was no gender difference in our study. CT and MR imaging of the brain confirmed the extra cranial position of the cyst and no intracranial extension. MRI alone probably would have been adequate. The size of the lesion in our study was between 10-20mm. Complete surgical excision with no complication or recurrence was achieved in the three patients operated. The

asymptomatic nature of this benign small lesion and the no change in size made the parents chose for follow up in the nine-year-old boy instead of surgical excision. The lesion did not grow over the two years follow up.

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