



Case Report

A surgical cure of a rare association parietal encephalocele with congenital aplasia cutis: A Case Report from Algeria

Cure chirurgicale d'une association rare d'une encéphalocèle pariétale avec une cutis aplasie congénitale :
un cas rapporté d'Algérie

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Abstract

Encephalocele is a congenital hernia of intracranial contents which protrude from a cranial defect. The intracranial contents which extrude to the exterior from the defect may include cerebrospinal fluid (CSF), meningeal structures, or brain tissue. Parietal encephalocele is rare. Congenital absence of skin or aplasia cutis is also a rare lesion that usually present in the midline over the vertex of the skull. The association of the two entities is very rare. The surgical treatment is challenging. we report a case of a fourteen months baby who presented with progressive swelling in parietal region and aplasia cutis congenital (ACC). The male baby was born at full term as the product of spontaneous delivery; the child was noted to have left parietal encephalocele of size 3 cm. Brain MRI revealed skull defect in the fronto-parietal region with aplasia cutis. The excision of encephalocele was carried out followed by a bone graft. After one year of follow-up, the boy is doing well. Parietal encephalocele are very rarely reported with only few published cases reported in the world literature and aplasia cutis with encephalocele in parietal

region has not been reported till date as we know. Excision and repair can give good results.

Keywords: Encephalocele, Aplasia cutis, Parietal, Algeria

Résumé

L'encéphalocèle se définit par une hernie du tissu cérébral et/ou des méninges à travers une déhiscence de la boîte crânienne. La localisation pariétale est rare. L'absence de peau ou cutis aplasia est aussi une lésion rare qui est habituellement sur la ligne médiane du vertex. L'association de ces deux entités est très rare et sa prise en charge chirurgicale est un challenge. Nous rapportons le cas d'un nourrisson de 14 mois né avec une voussure dans la région pariétale du crâne qui a augmenté de volume de façon progressive associée à une cutis aplasie congénitale. La TDM et l'IRM cérébrales ont objectivé un défaut osseux dans la région fronto pariétale avec une cutis aplasia. L'excision de l'encéphalocèle a été réalisée suivie d'une plastie osseuse.

L'enfant est en bon état général et la plaie bien cicatrisée après une année de suivi.

L'encéphalocèle pariétale est une lésion rarement rapportée dans la littérature mondiale, et l'association avec une cutis aplasia n'a jamais été publiée à notre connaissance. Une excision suivie d'une réparation chirurgicale peuvent donner de bons résultats.

Mots clés: encéphalocèle, aplasia cutis, parietale, Algérie

Introduction

An encephalocele is a congenital hernia of intracranial contents which protrude from a cranial defect. The intracranial contents which extrude to the exterior from the defect may include cerebrospinal fluid (CSF), meningeal structures, or brain tissue. Parietal encephalocele is a rare congenital anomaly of newborn with variable prognostic value. The congenital absence of skin or aplasia cutis is also a rare lesion that usually present in the midline over the vertex of the skull. The incidence is between 0,5 – 1/10000 of newborns [1]. The association of the two entities is very rare. The surgical treatment is challenging. here we report a case of a fourteen month baby who presented with progressive swelling in parietal region and aplasia cutis congenital (ACC).

Case Report

This is the case of fourteen months male baby brought to the CHU BEO Algiers with parietal swelling. The baby is full term normal delivered with 3.4 kg. There was no history of traumatisme, radiation exposure or any drug intake during antenatal period. The parents reported that the swelling was present at the time of birth and enlarged progressively but they did not take a

medical or surgical consultation for the baby during the initial 12 months. The baby is in good general health. On local examination, there was a swelling and a scalp ulcer of 3 cm size in the right parietal region involving only the skin (Fig.1) without cerebrospinal fluid (CSF) leak. Skull CT SCAN showed a focal defect in the parietal bone with protrusion of neural tissues suggestive of encephalocele (Fig.2). The brain MRI confirm the skull defect in the fronto-parietal region, the protrusion of neural tissues and the aplasia cutis (Fig.3). The patient underwent surgical intervention under general anesthesia. The excision of encephalocele was performed. After a dural plasty closed in a watertight manner, a bone graft was done over the defect. The edges of the wound were resected and a right inferior-lateral parietal scalp flap was rotated over the defect. After seven days a second operation was carried out because of the flap necrosis. We clean the wound by removing the tissue involved by the necrosis and put a skin graft (Fig.4). The post-operative course was uneventful after one year of follow-up (Fig.5).



Figure 1: Right parietal encephalocele



Figure 2: Skull CT scan showing a focal defect in the parietal bone



Figure 3: The brain MRI showing the skull defect in the fronto-parietal region, the protrusion of neural tissues



Figure 4: Image of the skin graft



Figure 5: One year post-surgery

Discussion

Encephalocele is a type of neural tube defect that occurs early in the fetal life resulting in neural tissue protruding through the defect in skull and/or dura. In developing countries, encephaloceles are not very uncommon. The most common site of encephalocele is occipital (75%) followed by fronto-ethmoidal (13-15%) [6]. Aplasia cutis with encephalocele in parietal region has not been reported till date. The index patient presented to us for the first time at an age of 14 months due to lack of awareness and probably slowly increasing size of swelling over the time. There are few more isolated cases reported suggesting parietal encephaloceles. Neuroimaging including computed tomography (CT) and magnetic resonance imaging (MRI) is the modality of choice for diagnosis. CT has the distinct advantage of showing bone defects while MRI has got greater value in delineating brain tissue anomalies [3].

Management of encephaloceles depends on type, size and hydrocephalous associated with encephalocele. Surgical treatment consisted of the excision of the sac and repair of dural defect after replacing the viable healthy brain tissue into cranial cavity [3].

Only aplasia cutis defects smaller than 1 cm may be expected to heal spontaneously [8]. Scalp defects should be treated surgically in patients where spontaneous epithelization from the edges of the wound cannot be expected to be complete within a few weeks.

Conservative management consists of regular wound cleansing and application of dressings along with the use of systemic antibiotics

Surgical management, on the contrary, includes various procedures. Standard surgical care includes primary wound closure, skin grafting (autologous or allografts), local scalp flaps with or without tissue expansion, free flaps, muscle flaps, full-thickness or split-thickness skin grafts, and cranial vault reconstruction using bone grafts. Specialized surgical techniques such as utilizing bipedicle opposing local flaps, rotational flaps, or L-shaped flaps have been used with satisfying results [8]. To prevent larger defects from drying out, it is necessary to cover the wound with the best suitable coverage available: either a flap or a split-thickness skin graft. Bony defects should always be covered by skin flaps or grafts. A skin graft will even take on dura and can give temporary coverage before a scalp flap is considered. A possible skull defect can be reconstructed with split-rib grafts or alloplastic materials, at a later stage. In larger defects, it may be necessary to cover the donor area with split skin grafts [2,7,9]. The end result of encephalocele surgery is usually not determined by the neurosurgical procedure per se, but by the amount of neural element herniating through the defect. The neural tissue is often dysplastic and gliotic and rarely functional [3,10,11]. Postoperative complications are usually hydrocephalous, infection and CSF leaking. Surgical treatment require most of the time a skin graft because of unhealthy skin over the encephalocele. This plasty could be done more than once.

Conclusion

Parietal encephalocele are rarely reported with only few published case reports in the world literature and cutis aplasia with encephalocele in parietal region has not been reported till date.

Surgical management remain a challenge because of the bad quality of the soft tissue.

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