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**Aplasia Cutis Congenital of Scalp With Right Parietal Encephalocele- A Rare
Association With Surgical Apropos**

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CLINICAL HISTORY:

A 3.5-month-old male baby was brought with complaints of swelling on the right side of the head which was noticed by the mother at birth. It was associated with the watery discharge with a gaping scalp wound.

PAST HISTORY

The patient was a 3.5-month-old, male child delivered at 39 weeks of gestation by LSCS.

The child was previously operated on postnatal day 15 at a private hospital for complaints of well-demarcated right parietal scalp defect measuring 3x1cm with watery discharge and with surrounding hypoplastic scalp rim with the absence of hair. There was no fever or neck rigidity.

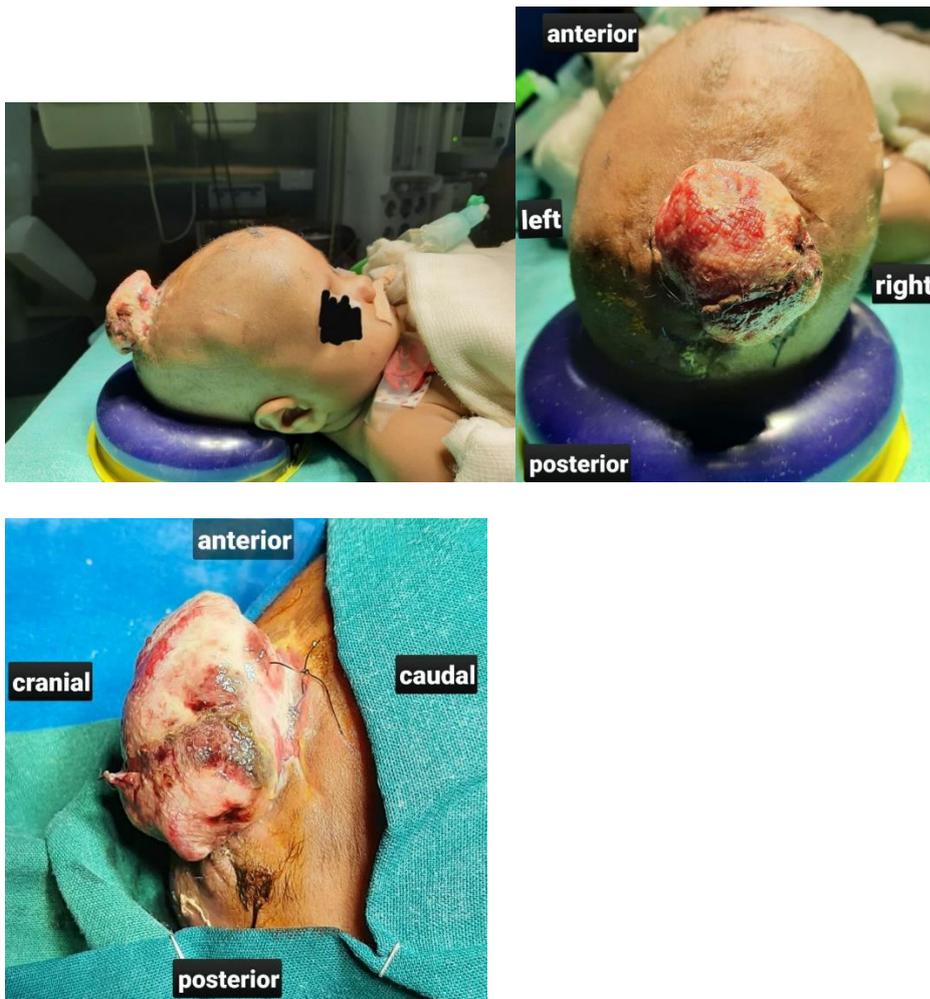
It was diagnosed as APLASIA CUTIS CONGENITAL of the scalp and was managed with the closure of the defect with sutures.

However, at 3.5 month of age, the child was taken to a private hospital for infected scalp post aplasia cutis, CSF leak & left upper limb focal seizures. There the baby underwent debridement with suturing for the same. The gaping wound was poorly managed by simple sutures of the scalp & the child was referred to a higher centre by the surgeon. Later the baby

was admitted to our hospital on 07/10/2020 due to increasing size of swelling with watery discharge and gaping wound margins.

The mother's obstetrical records revealed no exposure to teratogens or febrile episodes.

Except for the anomaly scan there were no other tests done for chromosomal anomalies.



EXAMINATION AND INVESTIGATIONS:

The baby was conscious, active, alert & was feeding well. Moved all 4 limbs.

LOCAL EXAMINATION

On local examination, there was an infected swelling in the right parietal scalp region just adjacent to the posterior fontanelle measuring 4 x 3 cm, translucent with overlying

granulation tissue. There were loose sutures from previous surgery in the gaping wound edges with watery discharge from the swelling. The swelling was almost reaching the midline and was surrounded by palpable bone anterolaterally and posterior fontanelle medially with an underlying stellate-shaped defect of the skull.

OTHER SYSTEMS

Respiratory System-B/L NVBS, No added sounds

CVS-S1, S2+, No murmurs

CNS-No focal neurological deficits.

Per abdomen-Soft, Non-tender

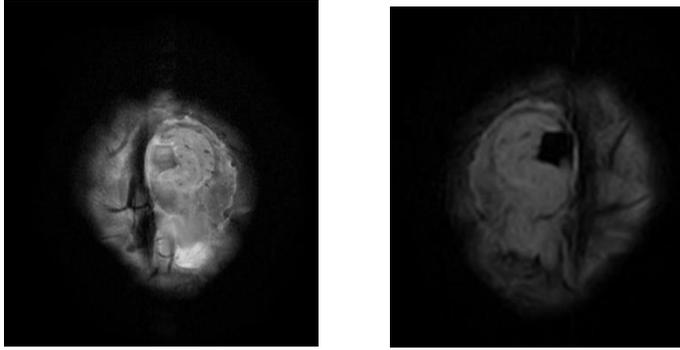
CT(Plain)-Gross widening of the sagittal suture, bregma, and adjacent coronal suture with herniation of right parietal lobe in extra-axial space.

Herniated brain shows gliotic changes.

Imp: Right parietal encephalocele.

MRI-Gross diastasis of the sagittal suture with right high parietal encephalocele.

Numerous small gyri along the inner cortex of the right frontal lobe with dilatation of extra-axial spaces adjacent to it with Polymicrogyria. The rest of the right holo-hemisphere & lateral ventricle is mildly displaced superiorly. Myelination is normal for age.



BLOOD INVESTIGATIONS:-

RBC-3.39mn/cumm

MCV-84.7FL

MCH-31.7gm/dl

TLC-16460 cells/cumm

PLATELET COUNT-6.36Lakh/cumm

FINAL DIAGNOSIS: APLASIA CUTIS CONGENITA OF SCALP WITH RIGHT

PARIETAL ENCEPHALOCOELE

DIFFERENTIAL DIAGNOSIS: -

-Right parietal encephalocele

-Cystic hygroma

-Epidermal scalp cyst

DISCUSSION:

Aplasia cutis congenital (ACC) is an uncommon condition with an estimated incidence of 3 in 10000 births with only about 600 reported cases to date (1). ACC is characterized by the congenital absence of variant layers of scalp- epidermis, and dermis and in some cases bone & dura involving the vertex. It is known to be associated with limb and cardiac anomalies and rarely with meningoencephalocele. Common aetiological factors are epidermolysis bullosa in the mother, chromosomal abnormalities, teratogens & intrauterine infections(2). Here we describe a case of ACC with right parietal encephalocele managed surgically with autologous fascia lata augmented duraplasty with temporo-occipital rotation scalp flap.ACC is a rare heterogeneous disorder with unknown pathogenesis. It is hypothesized to be due to developmental embryonic errors with incomplete closure of the outermost ectodermal portion of the neural tube(3). Other aetiological associations are epidermolysis bullosa, specific teratogens (methimazole and carbimazole), intrauterine infections, placental infarcts, chromosomal abnormalities & ectodermal dysplasias. This defect is usually covered by an ulcer or a thin membrane(4).

The child was evaluated clinically and radiologically at our center. The other authors have recommended CT head with 3D reconstruction, MRI head, MR venography and angiography, complete sonographic evaluation (ocular, cerebral, cardiac, abdomino genital), and X-ray of limbs (4).

Our aim of treatment was to achieve complete closure of the dural defect and hence avoid meningitis, bleeding, secondary local infection, and progressive brain damage. Earlier several authors have advocated conservative management to avoid operation and associated risks (2).

The size of the lesion and underlying skull defect are the criteria universally accepted

favoring the surgical approach (2,5). Lesions around critical areas such as sagittal sinus have increased chances of bleeding and infection which also favor surgical intervention. Other known complications are seizures due to electrolyte imbalance, nutritional deficiency from chronic blood loss. There are various surgical procedures which include excision and wound closure, skin grafting, local scalp flaps with or without tissue expansion, free flaps, musculocutaneous flaps, full-thickness or split-thickness skin graft, and cranial vault reconstruction. In our case, the dural defect was almost reaching the midline with only a 2mm rim of the dura adjacent to the right lateral border of the superior sagittal sinus. The autologous fascia lata graft was meticulously sutured to cover the dural defect. We did not close the associated skull defect as the child was only 4 months old and according to literature, a bone defect can regenerate or can be repaired as delayed cranioplasty(1). By surgical intervention we effectively avoided the major complications resulting from CSF leak with an exposed brain, bleeding from superior sagittal sinus, and lessened chances of infection by providing adequate scalp cover to underlying vital structures. We advocate surgical management of ACC with encephalocele.(6)

We would suggest complete syndromic evaluation of ACC and believe in an early surgical approach that minimizes complications and yet takes the advantage of innate regeneration ability of the newborn brain and the skull vault(7). This also highlights the need for referral of such children to specialized centers with sufficient experience and expertise(8).

ACKNOWLEDGEMENTS: None

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