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Parasitic Craniopagus with Cervical Myelomeningocele and Hydrocephalus: Reporting the rarely reported

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Objective: parasitic craniopagus is an extremely rare neurosurgical condition globally, additional anomalies such as hydrocephalus and myelomeningocele make it even more uncommon and complex to manage. The objective of this report was to describe successful management of a child with triad of craniopagus parasiticus, congenital hydrocephalus and cervical myelomeningocele

Case presentation: we successfully managed a five-month-old female infant with parasitic craniopagus, cervical myelomeningocele and congenital hydrocephalus.

Conclusion: parasitic craniopagus remains an uncommon neurosurgical condition and can be associated with many congenital anomalies such as hydrocephalus and myelomeningocele, careful evaluation and management will confer favourable outcome.

Keywords: parasitic twin, craniopagus, hydrocephalus, myelomeningocele

INTRODUCTION

The term craniopagus is a Greek word with cranio meaning head while pagus means together, therefore craniopagus is defined as twins joined only at the head. Parasitic craniopagus otherwise called craniopagus parasiticus is an extremely rare congenital malformation of conjoined twins, with one poorly developed twin referred to as parasite united at the cranium with the other well-developed twin, termed autosite (1-5).

Parasitic craniopagus is an uncommon form of twining with incidence of about 4 to 6 per 10, 000,000 live births, with higher occurrence observed in Africa and South-west Asia (3,6,7)

Craniopagus accounts for 2 to 6% of conjoined twin.(2) Females are more affected with conjoined twin than male's counterpart with a male-to-female-ratio of 1:4. (6)

Craniopagus parasiticus dates back to antiquity with first case reported in 1790 and later on fewer cases surface. (1) Parasitic twin also called, heteropagus and asymmetric conjoined twins are rare forms of monochorionic monoamniotic twins. The term heteropagus was coined by Potter and Craig while parasitic twin was named by Willis. (8)

The pathogenesis of parasitic twin remains controversial, the hypotheses put forward to explain how this anomaly occurs include: fission theory, which suggests incomplete separation of the two foetuses from one zygote in second week of gestation. While fusion theory proposes union of two distinct foetus. Also, it was postulated that parasitic twin occurs due vascular compromise in utero leading to loss of blood supply from degeneration of umbilical cord in one of the twins thereby causing death and partial resorption of parts of its body 9-11)

The exact aetiology of parasitic twin is not clear, however, usage of contraceptives, abnormal calcium metabolism and severely underweight woman with ovulatory dysfunctions were implicated. (12) Spencer and colleagues classify conjoined twins based on the anatomical site of the union into: first; twins with ventral union cephalopagus (head), thoracopagus (Thorax), omphalopagus (Abdomen) and Ischiopagus (pelvis)], second: twins with a dorsal union; pyopagus (sacrum), rachipagus (spine, back) and craniopagus (cranium) and the last group, twins with a lateral union termed parapagus(4).

Depending on the location, size of the lesion and presence or otherwise of associated anomalies, cranio-cervical computerised tomography scan and or magnetic

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resonance imaging will show the exact location of the lesion, extents, contents and or consistency of the parasite, which will help in planning surgical excision and eventually favourable outcome.

The surgical excision of the parasitic twin is best and most acceptable treatment option. The goals should be to have healthy and neurologically intact autositic twin with also an acceptable cosmetic appearance. Timing of surgery was suggested to be between two-to-fifteen months in order to have good post-operative outcome.(2)

Coincidentally, the earliest report of conjoined twins (Omphalopagus) in Nigeria were born and surgically treated by a British missionary doctor in 1935 in Sokoto, city where the regional centre for neurosurgery that the index case was managed is located (13-14). To the best of our knowledge this is the first report of a parasitic craniopagus associated with congenital hydrocephalus and cervical myelomeningocele in Nigeria. As such, this paper aims to describe the successful management of this uncommon syndrome of craniopagus parasiticus, congenital hydrocephalus and cervical myelomeningocele and add to the volume of scientific literature on this interesting congenital condition.

CASE REPORT

We managed a five months old female infant who presented with occipito-cervical and separate cervical swellings noticed immediately after birth. The masses were gradually increasing in size with no associated head enlargement. The child cried immediately after birth but there was no neck control or social smile at five-month of age. There was no history of maternal febrile illness or use of unprescribed medications. Mother is a 25-year-old para 3 and had no history of twining or any other congenital anomaly in the family. Child was delivered at term via spontaneous vaginal delivery at home under no supervision in neighbouring village in Kebbi state.

Examination revealed calm infant, not in any form of distress, not pale, afebrile with Occipito-frontal circumference of 44cm. He had two separate masses: irregular shaped Occipito-cervical mass measuring about 20 by 26 by 18cm, non-tender with no differential warm, mixed consistency with areas of hard and soft consistencies. With head and face-like structure consisting of forehead, hairy skin, poorly developed eye depressions and proboscis-like nose, mouth depression and jaw-like structure. While the inferior mass is cystic measuring about 30 by 34 by 24 cm, non-tender, fluctuant and transilluminate (Figure 1). Cranio-cervical magnetic resonance imaging shows marked ventricular dilatation, poorly developed cerebellum herniating into foramen magnum, Occipito-cervical mass containing soft and hard tissues of variable intensities and cervico-thoracic mass contains cerebrospinal fluid and soft tissues (Figure 2).

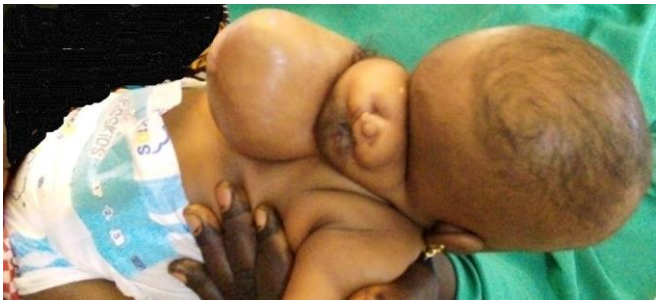

Figure 1-Clinical photograph showing craniopagus and cervical myelomeningoecele

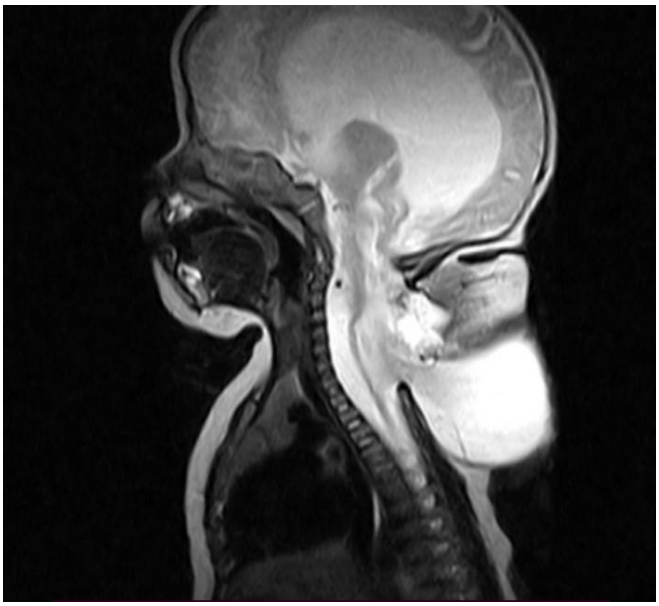

Figure 2-T-2 weighted Sagital view cranio-cervical MRI showing hydrocephalus, and contents of craniopagus and cervical myelomeningoecele

Haemoglobin level was 15g/dl, serum electrolyte, urea and creatinine were all normal. Echocardiography revealed normal heart. A diagnosis of parasitic craniopagus with congenital hydrocephalus and cervico-thoracic myelomeningocele was made. Parents were counselled and child subsequently had ventriculoperitoneal shunt, excision of parasitic craniopagus and excision and repair of myelomeningocele at the same sitting under general anaesthesia (Figure 3). Child recovered fully from anaesthesia and discharged home ten days after surgery, to see at neurosurgical out-patient clinic for follow-up. The child has remained stable and parent reported social smile and neck control one month after surgery (Figure 4 and 5). The resected tissues were sent for histology and histopathological analysis, which revealed dysplastic neural tissues, bone, and meninges. These findings are inconsistent with teratoma.

Figure 3-Intra-operative clinical photograph showing contents of the lesion

Figure 4- Immediate post-operative image

Figure 5- One month follow-up

DISCUSSION

The development of parasitic twin is attributed mainly to abnormal twinning which was believed to be monozygotic but further scientific evidences have shown the probability of dizygosity.(4) Several possible explanations trying to unravel how parasitic twin develop were elucidated in literature: Faulty secondary neurulation, in which excess production of neural tube fluid leaking into the subcutaneous space differentiate to various types of tissues and body parts.(15)

Furthermore, some researchers have attributed the development of parasitic twin to changes in sonic hedgehog protein (SHH), a segment polarity protein responsible for growth, structure and patterning process of embryo. High levels of SHH have been implicated in the development of parasitic twins. Parasitic craniopagus could be seen in all genders but as shown in the index case slight female preponderance was reported in literature(6).

Our case in addition to parasitic craniopagus also had congenital hydrocephalus and cervical myelomeningocele, making this rare condition even more uncommon. There have been reports of parasitic twins coexisting with other anomalies but exact combination of the aforementioned conditions is extremely rare in published literature.(6) Several factors were attributed to development of conjoined and parasitic twins including: use of contraceptives, underweight mothers, abnormal calcium metabolism, mothers with ovulatory dysfunctions and consanguinity (1,4,12). No known risk factor was identified in the present case.

Neuroimaging such as brain computerised tomography scan or magnetic resonance is very important in evaluation and surgical planning of this conditions. Imaging helps to reveal neurovascular tissue present which guide decisions to operate or not in cases not expected to survive after surgery or refer to centres with all the required facilities required to manage conjoined and parasitic twins.

Rarity of conjoined and parasitic twins has made standardised treatment algorism unavailable, as such treatment depends on site of fusion, contents of the parasite, survivability of the autosite and the availability of required skills and facilities. The present case had three separate conditions all needing surgical operations. After careful evaluation and discussion with parents, the child was offered ventriculoperitoneal shunt, excision and repair of cervical myelomeningocele and complete excision of parasitic twin at same sitting. He had remarkable recovery and was discharge home and advised to be regular on follow up. Child's parents were happy with the cosmetic appearance and reported improvement in development milestones, social smile a month after surgery and attained sitting posture three months following surgery (at 8 months of birth). The improvement in development milestone could be attributed to relieve of intracranial pressure following ventriculoperitoneal shunt surgery.

CONCLUSION

Parasitic craniopagus is an extremely rare congenital anomaly globally, coexistence of other anomalies makes it more complex but careful selection, evaluation and management may bring an acceptable outcome to both parent and the surgeon.

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DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was not requested by the local Ethics Committee: "THE PATIENT WAS MANAGED WITHOUT ANY INFRINGEMENT IN THE RIGHT OF PATIENT OR INTERVENTION THAT WARRANTS ETHICAL APPROVAL."

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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CONTRIBUTIONS

-**ALIYU MUHAMMAD KOKO**: Conceptualization, Data curation, Formal Analysis, Methodology, Writing – original draft, Writing – review & editing
-**ALI LASSEINI**: Conceptualization, Methodology, Supervision, Writing – review & editing
-**NASIRU JINJIRI ISMAIL**: Formal Analysis, Methodology, Supervision, Writing – review & editing
-BELLO SHEHU: Supervision, Validation, Visualization, Writing – review & editing

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