CASE REPORT

Extensive Cerebriform Nevus Sebaceous In A Zambian Child: A Case Report

Muleya Inambao[1], Mwiche Nachona[2], Patrick Jila[3], Bwendo Nduna[4], Chishala Chabala[5]

[1] *Paediatrician, Arthur Davison Children's Hospital, Ndola, Zambia*
[2] *Paediatric Registrar, Arthur Davison Children's Hospital, Ndola, Zambia*
[3] *Paediatric surgery Registrar, Arthur Davison Children's Hospital, Ndola, Zambia*
[4] *Consultant Paediatrician, Arthur Davison Children's Hospital, Ndola, Zambia*
[5] *Consultant Paediatrician, University Teaching Hospital-Children's Hospital, Lusaka, Zambia*

ABSTRACT

Background

Cerebriform nevus is an extremely rare hamartoma of skin characterized by convoluted surface resembling cerebral gyri usually present at birth or in early childhood. It is a rare dermatological condition with extremely limited reports from Africa. Sebaceous nevus (SN) is a rare congenital hamartoma, most often located on the scalp, though it may also affect other areas of the head and neck. It is classified as an epidermal nevus characterized by the presence of sebaceous glands. Clinically, the lesion typically appears as a hairless plaque.

Main clinical findings

It typically presents as a hyperpigmented patch commonly over the scalp, trunk and lower limb. It acquires multiple convolutions over the surface giving rise to characteristic 'cerebriform' appearance and tends to increase in size during puberty under hormonal influence. The lesion though benign may pose a risk of malignant transformation to melanoma, particularly in giant or extensive lesions in adulthood. We report a case of extensive Cerebriform Nevus most likely sebaceous in a 5-year-old girl who presented with hyperpigmented indurations on the scalp, right side of the face which were non progressive in size.

Diagnosis and management

The definitive diagnosis of an extensive cerebriform nevus is made by histopathological examination of the skin biopsy which typically shows hyperplasia of adnexal structures (sebaceous glands, hair follicles may be reduced/absent) and thickened dermis with papillomatosis and cerebriform surface architecture

Conclusion

This is the first documented case from Zambia which highlights the rarity of the condition with limited knowledge, need for awareness,

Corresponding author:
Dr Muleya Inambao-
Arthur Davison children's Hospital.
P.O.BOX 240227, Ndola, Zambia
Email: muleyainambao@gmail.com

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psychosocial and cosmetic concerns ,diagnosis and treatment challenges in a resource limited setting.

INTRODUCTION

Nevus sebaceous (NS) is an uncommon epidermal nevus, predominantly composed of sebaceous glands, and usually presents at birth[1]. It typically manifests on the head or neck, most often the scalp, as a plaque with overlying alopecia[2]. Cerebriform NS is an atypical and rare morphological variant[3]. It was first described in a 20-year-old patient in 1998, with subsequent cases reported in an infant and in five neonates[4].

The exact cause of its cerebriform surface appearance remains unclear. NS generally progresses through three overlapping stages:

1. Infantile stage – characterized by a flat plaque with papillomatous hyperplasia and immature hair follicles.
2. Pubertal stage – marked by hormone-driven rapid growth, verrucous surface changes, development of sebaceous elements, and maturation of apocrine glands.
3. Adult stage – associated with the emergence of secondary neoplasms, both benign (reported in about 16% of cases) and malignant (approximately 8% of cases)[4].

There is very limited literature on cerebriform NS in African children. This could be due to its global rarity, a genuinely low prevalence in African populations, or underreporting. Underreporting may stem from limited access to dermatological expertise, misdiagnosis as other skin conditions, or restricted opportunities for publication within the region.This case report aims to contribute to the scarce global literature on cerebriform NS by documenting an extensive case in a Zambian child. It emphasizes the importance of awareness, accurate diagnosis through clinical examination and biopsy, and the need to expand reporting from African contexts.

Case Presentation

A 5-year-old girl from Mpongwe District, Zambia, presented with multiple raised, hyperpigmented, and indurated skin lesions that had been noted at birth. The lesions were non-progressive in size or extent, enlarging only in proportion to overall body growth. There was no history of seizures, visual or hearing impairment, or skeletal deformities. The lesions did not cause pain or functional limitation but had a significant negative psychosocial impact, as the child experienced social withdrawal due to teasing and stigma from peers. The mother expressed deep concern regarding the cosmetic disfigurement and its effects on her daughter's self-esteem and confidence.

She was born at term at a local clinic, weighing 3.5 kg, and was fully immunized for age. Soon after birth, she was referred to the Neonatal Intensive Care Unit (NICU) at Arthur Davison Children's Hospital for evaluation of extensive skin lesions. A biopsy for histopathological assessment was performed at that time, but the results were not communicated to the family as the mother relocated to another district, leading to loss to follow-up. There was no history of similar lesions in family members, and no known genetic disorders or syndromic associations reported. The child's developmental milestones were normal and appropriate for her age, and there were no comorbid medical conditions.

On examination, she appeared cheerful and developmentally normal. Multiple meandering, pedunculated, and non-pedunculated lesions were noted on the face and scalp, associated with areas of alopecia. The lesions were non-tender and exhibited a convoluted, cerebriform surface resembling sulci and gyri, particularly over the parietal and occipital regions. The ears and adjacent scalp were similarly affected. In contrast, the chest and back displayed flat, hyperpigmented plaques (Figures 1–6). A biopsy specimen taken from a convoluted scalp lesion (10 × 10 × 5 mm) revealed pseudocyst formation and papillomatosis consistent with *seborrheic keratosis.* No neuroimaging or systemic evaluations were performed, as there were no neurological or skeletal abnormalities noted.(Table 1)

Based on clinical morphology, especially the brain-like convolutions and characteristic histopathological findings, a diagnosis of **Extensive cerebriform nevus sebaceous** was established. The family received counselling regarding the benign but chronic nature of the condition, the importance of follow-up, and options for cosmetic management in the future. Skin care advice was provided, and regular reviews were scheduled in the surgical outpatient clinic to monitor any changes suggestive of secondary neoplasia or functional impairment.

Table 1: Timeline of the clinical events

Time	Event	Details / Findings
At birth (0 months)	Lesions first noted	Raised hyperpigmented indurated plaques on scalp and face noted at birth; non-tender; no systemic symptoms.
Neonatal period	Referred to NICU	Referred from local clinic to Neonatal Intensive Care Unit for assessment of extensive skin lesions.
Neonatal period (Day 3–7)	Biopsy performed	Skin biopsy taken for histopathology; results not communicated to family.
Infancy (0–12 months)	Clinical stability	Lesions remained stable in extent and size (enlarging only proportionally with growth); no seizures, vision, or skeletal abnormalities.
Early childhood (1–4 years)	Lost to follow-up	Family had relocated to another district, no dermatology or surgical follow-up and no interventions administered.
Age 5 years (current presentation)	Re-evaluation at surgical clinic	Mother sought review due to cosmetic concerns and social stigma. Lesions non-progressive; child otherwise healthy and developmentally normal.
At 5 years (clinical examination)	Findings	Cerebriform, meandering lesions on scalp and face with alopecia; flat hyperpigmented plaques on trunk. No tenderness or neurological deficits.
At 5 years (histopathology)	Diagnosis	Biopsy revealed pseudocyst formation and papillomatosis consistent with seborrheic keratosis, supporting diagnosis of cerebriform nevus sebaceous.
At 5 years (management)	Counselling and follow-up	Counselling provided to mother regarding benign nature, cosmetic impact, and need for long-term surveillance; follow-up scheduled at outpatient surgical clinic.

Table 2: Investigations done

Laboratory Test	Result
Full Blood Count	Haemoglobin: 11.7 g/dL WBC: 5.49 × 10^3/µL (Neutrophils 44%, Lymphocytes 46.4%, Monocytes 5.8%) Platelets: 231 × 10^3/µL RBC: 0.7 × 106 /µL HCT: 34.6% MCV: 71.6 fL MCH: 24.3 pg
HIV Test	Negative
Histopathology	**Microscopy:** Skin sections showed pseudocyst formation and papillomatosis lined by keratinising stratified squamous epithelium and a diagnosis of Seborrheic Keratosis was made

Figure 1: Occipital view showing multiple raised, hyperpigmented, pedunculated lesions on the face and scalp with areas of alopecia.

Figure 3: Right parietal region showing extensive cerebriform plaques.

Figure 2: Close-up of convoluted scalp lesion demonstrating cerebriform surface with sulci and gyri-like pattern on right parietal, frontal, face regions and chest with hyperpigmentation.

Figure 4: Cerebriform plaques on left parieto- occipital, left post auricular, posterior surface of the left ear, face with hyperpigmented skin on the back and upper limbs

Figure 5: Flat hyperpigmented macular lesions over the upper back and chest.

Figure 6: Cerebriform lesions on the occipito-parietal, temporal, frontal regions and hyperpigmentation on the left shoulder and back

Differential Diagnosis

Cerebriform Nevus Sebaceous (CNS) is a rare variant of nevus sebaceous, characterized by a brain-like surface with multiple folds resembling sulci and gyri.[4] The condition is typically congenital and most commonly affects the scalp or face. Although benign, nevus sebaceous has the potential for secondary benign or malignant neoplastic transformation later in life, warranting long-term surveillance.[4] The main differential diagnoses for our patient included:

1. Epidermal Nevus – may present at birth with verrucous or hyperpigmented plaques; however, these lesions typically lack the cerebriform surface and sebaceous gland proliferation seen in CNS.[4]
2. Sebaceous Hyperplasia – usually occurs as small, yellowish papules in adults due to sebaceous gland overgrowth, rather than large congenital plaques.[4]
3. Nevus Sebaceous Syndrome (Schimmel penning-Feuerstein-Mims Syndrome**)** – characterized by nevus sebaceous associated with neurological, ocular, and skeletal abnormalities. In this case, the absence of seizures, developmental delay, or skeletal deformities made the syndromic form less likely.[5]

Diagnostic Work-up and Limitations

No neuroimaging or skeletal imaging studies were performed in this case. This was partly due to the absence of neurological or musculoskeletal abnormalities on clinical evaluation and also reflected limitations in diagnostic resources. MRI or CT scan were not readily available. Laboratory investigations and systemic screening were limited to basic clinical examination, and the histopathological diagnosis relied on a single biopsy specimen. The missed follow-up of the initial neonatal biopsy further delayed definitive diagnosis and highlights systemic barriers to continuity of care and record retrieval. Despite these limitations, the combination of characteristic clinical features (cerebriform surface with alopecia) and confirmatory histopathological findings were sufficient for diagnosis.

Diagnostic Challenges in a Resource-Limited Setting

The case underscores common challenges faced in low-resource environments: restricted access to specialized dermatology, imaging, inadequate patient tracking systems, and limited multidisciplinary support. Despite these constraints,

diagnosis was achieved through integration of clinical morphology and histopathological features.

Psychosocial Considerations

Beyond medical concerns, the psychosocial impact was profound. The child faced social stigma and self-esteem issues due to visible disfigurement, affecting school attendance and peer interactions. Early psychosocial support, counselling, and education for the family and community are essential components of care, especially in settings where cosmetic anomalies may lead to social exclusion. This case illustrates psychosocial challenges of managing a rare dermatological condition like cerebriform nevus sebaceous in a resource-limited setting. It underscores the need for improved access to multidisciplinary care including psychological support and patient education.

DISCUSSION

There is scarcity of epidemiological studies on cerebriform NS in African children making it a challenge to determine the incidence and prevalence. Many cases are not reported until adulthood, as the condition is often asymptomatic and patients seek medical attention for cosmetic reasons.

Cerebriform nevus sebaceus is very rare and has been reported by Ramesh et al in an adult with a large lesion on the right parieto-occipital region, diagnosed as NS.[4] Subsequently Bomztyk et al reported a case in a 6-month-old infant in the postauricular region [5].Correale et al reported five(5) cases of a rare form of NS describing it as large, papillomatous, and pendulous.[6] Patients presented at birth with large exophytic lesions including Seborrheic keratosis and epidermal nevus which were clinically difficult to differentiate.[7] NS can be extensive covering multiple body regions and raising concern for an associated syndrome.[8] Rarely, NS can present as a cerebriform, exophytic plaque or tumor.[9] During puberty Nevus Sebaceus undergoes hormone-driven changes, becoming thicker and more verrucous (wart-like) in texture, which can often lead to cosmetic concerns for patients and/or their parents.[10] Though in our patient, the verrucous (wart like) presented much earlier before puberty.

On histology, Nevus Sebaceus (NS) exhibits follicular, sebaceous, and occasionally apocrine components, with specific features varying based on the patient's pubertal status.[10] In prepubertal NS lesions, the superficial dermis contains numerous but underdeveloped and malformed folliculosebaceous units, characterized by abortive hair follicles and small sebaceous glands.[10] After puberty, NS lesions show a more acanthotic, papillomatous, and verrucous epidermis, accompanied by fibroplasia in the papillary dermis.[11]Whittington et al. described numerous immature, malformed sebaceous units located in the superficial dermis, along with tiny, misshapen hair follicles and an acanthotic, papillomatous epidermis on histology in a neonate.[9] Histopathology for our patient showed seborrheic keratosis indicating sebaceous glands involvement although confirmation was limited due to the single biopsy site.

The differential diagnosis include Sebaceous adenomas, epidermal nervus, and sebaceous hyperplasia.[12] Sebaceous adenomas have immature lobules while sebaceoma are characterized by basaloid cells and sebaceous ducts.[12] In contrast to NS, sebaceous hyperplasia shows mature sebaceous gland lobules and prominent sebaceous ducts.[13] Neurocutaneous syndrome may also occur, especially in extensive nevi, with epilepsy, mental retardation, or other neurological and skeletal abnormalities.[13]Our Patient had hyperpigmented skin lesions but no neurological deficit.

Management involves a stepwise early excision and is usually recommended to prevent potential malignant transformation in delayed excison.[14] Treatment favoured is wide excision followed by skin graft; when not feasible, close observation is recommended.[14] In our case, diagnostic

investigations were limited due to resource constraints. Counselling on skin care was done and subsequent follow up reviews planned to monitor for possible malignant transformation.[9]

The child's mother expressed deep concern over the visible skin lesions, describing them as a source of **social stigma and emotional distress** for both her and her daughter. She reported that other children often avoided her leading to **episodes of social isolation and school absenteeism**. She also shared frustration regarding the **fragmented nature of care** relocating to another district made continuity of care difficult. Nevertheless, she appreciated the recent counselling received at the surgical clinic and expressed hope that **further evaluation and treatment might improve her daughter's appearance and confidence,** allowing her to participate more freely in community activities.

The Follow-up management for our patient focused on the following:

- Clinic attendance: The child was reviewed at the surgical outpatient clinic at age 5 years for re-evaluation after relocation. This visit is the most recent documented follow-up.
- Growth and development: Growth parameters and neurodevelopment remain appropriate for age; the child continues to meet expected developmental milestones and is described as a "happy child." No new neurological, visual, or auditory symptoms have been reported since presentation.
- Local complications: At the time of the outpatient review there were no signs of active infection, bleeding, ulceration, rapid nodular change, or other acute complications in the lesions. The lesions remain non-tender and non-progressive in extent beyond proportional growth.
- Psychosocial impact: Ongoing psychosocial morbidity was documented: the mother reported social withdrawal due to teasing and stigma. Counselling was provided during the visit and a plan for psychological support was initiated.
- Definitive therapy and complications: No definitive surgical or ablative therapy has yet been performed. Therefore no post-operative complications, recurrence data or long-term local outcomes are available.

Definitive management of nevus sebaceous involves **surgical excision**, either as a **single-stage wide local excision** for smaller lesions or **staged excision with/without tissue expansion** for larger scalp or facial lesions to preserve hair-bearing skin. **Reconstruction** may involve local flaps, skin grafts, or free/pedicled flaps, with scalp-specific techniques used to enhance cosmetic outcomes. In Zambia, advanced reconstructive surgery is available mainly at tertiary facility like the University Teaching Hospital with possibly limited capacity for complex or microsurgical procedures.

The Justification for Current Approach taken for our patient is the lesions are extensive but non-progressive, with no neurological or systemic involvement. The main impact is psychosocial. Given the family's previous displacement, missed follow-up after neonatal biopsy, and limited access to specialized care, a cautious, supportive, and coordinated approach with renewed psychosocial support is justified. Conservative management with counselling and follow-up was appropriate because the large lesion required complex, staged reconstruction only available at specialized Centres. The absence of neurological signs made imaging a lower priority, and limited access to advanced imaging further supported this approach. Priority was given to establishing a referral pathway and providing psychosocial support.The child remains stable under outpatient surveillance with ongoing psychosocial support and plans for future tertiary referral when feasible. The next steps for our patient are:

1. **Specialist Referral:** Refer to UTH plastic surgery or dermatology unit for multidisciplinary assessment.
2. **Definitive Surgery (if feasible):** for possible plan staged excision with tissue expansion or

local flaps to preserve hair; use skin grafts only when necessary.

3. **Psychosocial Support:** Provide counselling, involve the school and promote peer education to reduce stigma.
4. **Surveillance:** Conduct regular skin exams every 6–12 months, educate caregivers on warning signs, and maintenance of detailed records and long-term follow-up into adolescence.

CONCLUSION

Cerebriform NS is a rare epidermal nevus primarily comprised of sebaceous glands, causing significant cosmetic disfigurement like in our case. Giant cerebriform NS is extremely rare. Early diagnosis of Cerebriform NS is crucial for planning surgical excision especially for extensive lesions. Based on our patient's clinical and histopathological findings, Cerebriform NS is most likely the cause of the broad cerebriform appearance, which makes this case notable to be documented in Zambia. Long-term outcomes like malignant transformation, hair restoration, and psychosocial recovery remain unknown. Follow-up is ongoing, at 5 years the child remained developmentally normal with no local complications; however, significant psychosocial morbidity (school absenteeism and social withdrawal) persisted. No definitive surgical therapy has been undertaken to date, and a structured surveillance plan was established:

Strengths, Limitations, and Lessons Learned

This case contributes to the limited African literature on cerebriform nevus sebaceous, a rare congenital lesion infrequently reported in sub-Saharan settings. Its detailed clinical description, histopathologic correlation, and psychosocial perspective add regional relevance and highlight the need for greater awareness among clinicians. However, several limitations are acknowledged. No imaging studies were performed due to resource constraints and the absence of neurological symptoms; the diagnosis was based on a single biopsy site, which may not fully represent the lesion's histologic heterogeneity; and the long term follow-up for assessment of long-term outcomes such as recurrence or malignant transformation.Despite these limitations, the case underscores key lessons which are the need for early diagnosis, consistent follow-up, multidisciplinary care, and capacity-building in dermatologic and reconstructive surgery to enhance access and long-term outcomes in resource-limited African settings.

Acknowledgement

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Declaration of patient consent

The parent gave consent for the images and other clinical information of her child to be reported in the journal. The parent understand that the name and initials of their child will not be published, and due efforts will be made to conceal their identity.

Conflict of Interest

The authors declare that they have no conflict of interest

Consent to Publish declaration

The consent to publishing the case report was given by the Head of Institution of Arthur Davison Children's Hospital

Ethical Consideration

The case is original and has not been published anywhere. Written informed consent for publication of this case and accompanying clinical photographs was obtained from the child's mother after thorough explanation of the purpose, content, and potential educational value of the report and the patient's anonymity has been protected . We have followed ZMA's guidelines on confidentiality and anonymity. The authors emphasize the importance of **ethical sensitivity** in cases involving paediatric patients

with visible congenital anomalies, ensuring compassionate communication, psychosocial support, and family participation in decision-making throughout care and publication processes.

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