Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Dermatology

Adams-Oliver Syndrome: About A Case

F. Sylla[4], Dr. Lassine Keita[4*], K. Ba[4], C. Keita[4], C. Traore[1], M. Kanté[5], N. Koné[6], Y. Fofana[3], A. G. Dicko[1], M. Diakité[1], N. Kontao[7], F. Dicko[2], M. Sylla[2]

[1]Bamako Dermatology Hospital, Mali
[2]Gabriel Toure Hospital, Mali
[3]Somino Dolo Hospital, Mali
[4]Commune II Reference Health Center
[5]Commune V Reference Health Center
[6]Kiffa Regional Hospital (Mauritania)
[7]Mohamed VI Perinatal Clinic, Mali

DOI: 10.36347/sjmcr.2023.v11i02.006 | **Received:** 19.12.2022 | **Accepted:** 25.01.2023 | **Published:** 05.02.2023

***Corresponding author:** Dr. Lassine Keita
Commune II Reference Health Center

Abstract **Case Report**

Adams-Oliver syndrome is a polymalformative disease rare whose incidence is estimated at 1/225,000. This is a female newborn, referred by the maternity ward on D0 of life, hospitalized for a polymalformative syndrome. We noted a shallow and non-traumatic parietal ulceration on a background of congenital aplasia of the scalp without bone defect, with peripheral venous ectasia, Cutis Marmorata, Congenital Telangiectatica. Moreover; second and third toe brachydactyly with nail agenesis. We managed the ulcerations with local care and offered regular follow-up to minimize the occurrence of serious complications. Unfortunately the newborn is lost to sight after a few days.

Keywords: Syndrome, Adams-Oliver, mottling, bradydactyly.

INTRODUCTION

Adams-Oliver syndrome (OSA) is a rare disease with an estimated incidence of 1/225,000 [1]. It combines congenital anomalies of the limbs (brachydactyly, agenesis of the nails, etc.) and of the scalp (aplasia of the scalp), often accompanied by defects in the ossification of the skull. The severity of the syndrome varies considerably from patient to patient [2].

We report a case of Adams-Oliver syndrome.

OBSERVATION

This is a female newborn, sent by the maternity ward on D0 of life, hospitalized for polymalformative syndrome. He is the result of a full-term pregnancy that was poorly monitored with a single obstetrical ultrasound performed in the third trimester without any particularities. The delivery was vaginal with an Apgar at 9/10 at the first minute. It is the fourth child of the siblings and has three older brothers who are all in good health. At birth, her weight was 2010g, her height 47cm, her head circumference 31.5cm and her general condition was good.

The physical examination found a shallow and non-traumatic parietal cephalic ulceration (2.2/2.4 cm) on a background of congenital cutaneous aplasia of the scalp without bone defect, with peripheral venous ectasia (Fig. 1). On the trunk there is a diffuse and wide marbling extending to the limbs with a collateral venous ectasia, peri-abdominal suggesting a Cutis Marmorata. Telangiectatica Congenita (Fig. 2). Moreover; brachydactyl and toenail agenesis (Fig. 3). The rest of the clinical examination was unremarkable. The paraclinical explorations in search of other abnormalities were also unremarkable ~~to~~ date. We managed the ulcerations with local care and offered regular follow-up to minimize the occurrence of serious complications. For this purpose, the child was seen three times in an interval of two months.

Unfortunately, he has been lost to sight ever since.

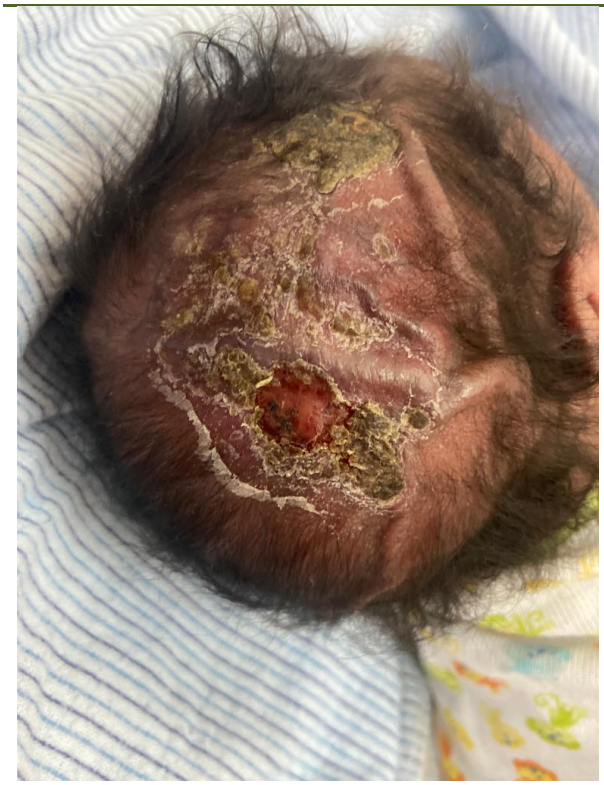

Figure 1

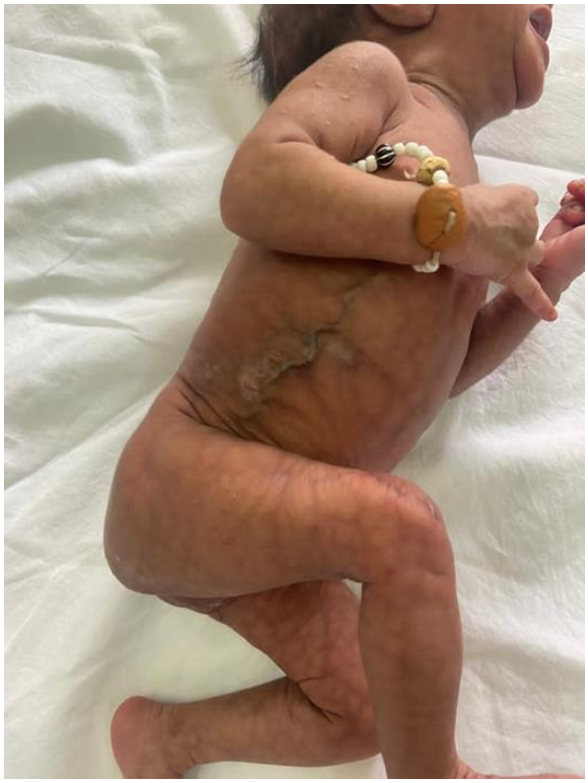

Figure 2

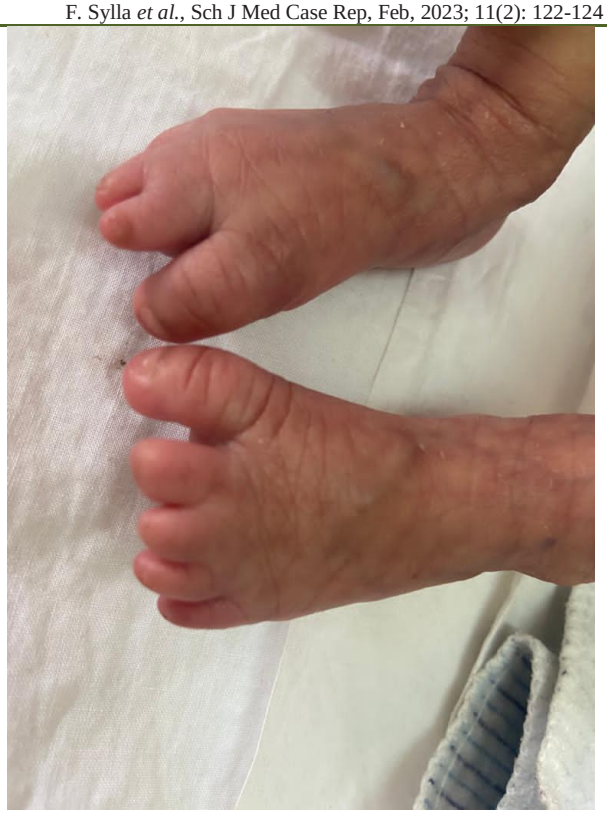

Figure 3

DISCUSSION

To our knowledge, this is the first case of Adams-Oliver syndrome described in Mali.

Adams-Oliver syndrome can be life-threatening when accompanied by cardiovascular, pulmonary or neurological manifestations. Our reported case has minor clinical presentations characterized by transverse abnormalities of the limbs (brachydactyly, nail agenesis) which constitute the first manifestation of the syndrome with a prevalence in the order of 84% [1], followed by aplasia congenital cutaneous skin of the scalp found in a frequency of 75% [3], the veno-capillary manifestations are constitutive of this syndrome of 15-25 [1] dominated by a Cutis Marmorata Telangiectatica Congénita (CMTC) corresponding to persistent reticular marbling after few days of life. It is a pathological state that is diffuse with dilation of the veins in the form of a transverse peri-abdominal and cephalic cord. However, there is the physiological CMTC in newborns which disappears on body warming a few hours after birth [4]. Pathological CMTC independently of Adams Oliver syndrome does not associate transverse anomalies of the limbs or congenital aplasia of the scalp, even less cardiac or neurological complications [3]. It was also noted a congenital cutaneous aplasia of the isolated scalp which does not integrate the poly malformation syndrome.

We could also mention other congenital diseases which have vascular malformations in common, in particular Blackenheimer syndrome or Klippel-Trenaunay syndrome; the latter has an easy diagnosis in front of the classic triad: associating an angioma in the limb, venous malformations and hypertrophy of the soft tissues and/or bone generally giving rise to asymmetry of the limb concerned [5]. The second is characterized by diffuse and large but painful venous ectasias frequently located at the level of a single limb, most often the upper limb [6].

It is probably sporadic because the interrogation did not reveal any family case and or a notion of marriage by consanguinity from which the newborn would have come.

CONCLUSION

This is a rare condition; follow-up must be multidisciplinary and regular in order to detect any late malformations that could be life-threatening.

BIBLIOGRAPHIC REFERENCES

1. Kojmane, W., Hmami, F., & Atmani, S. (2019). Adams-Oliver syndrome and cutis marmorata telangiectatica congenita. In: Annals of Dermatology and Venereology. Elsevier; p. 223-5.
2. Messerer, M., Diabira, S., Belliard, H., & Hamlat, A. (2010). Adams-Oliver syndrome: about a case of minimal expression. *Arch Pediatrics*, 17(10), 1460-4.
3. Yéo, S., Perrot, P., Bellier-Waast, F., David, A., & Duteille, F. (2010). Adams-Oliver syndrome. About a clinical case. *Chir Main*, 29(4), 274-6.
4. Baline, K., & Benchikhi, H. (2014). Cutis marmorata telangiectatica congenita revealing neonatal lupus. *Pan Afr Med J.*, 6, 339.
5. Samimi, M., & Lorette, G. (2010). Klippel-Trenaunay syndrome. *Medical Press*, 39(4), 487-94.
6. van Geest, A. J., Veraart, J. C. J. M,. de Haan, M., & Neumann, H. A. M. (1999). Bockenheimer's syndrome. *J Eur Acad Dermatol Venereol*, 12(2), 165-8.