Isolated Congenital Giant Fetal Upper Limb and Trunk Haemangioma Diagnosed Antenatally Using 4D Ultrasonography and Regressed by Age of Five Months. A Case Report

Amr Hassaan Farag (dr_amrhassaan@hotmail.com)
Ain Shams University https://orcid.org/0000-0002-7485-7571
Heba Sadek
Ain Shams University Faculty of Medicine
Mohamed Nasr El-Deen
Ain Shams University Faculty of Medicine

Research Article

Keywords: 4D, Doppler, fetal, haemangioma, limb

Posted Date: March 23rd, 2022

DOI: https://doi.org/10.21203/rs.3.rs-181240/v1

Abstract

Introduction: Congenital haemangiomas are benign vascular tumours that are the most common of all congenital anomalies.

Case presentation: A rare condition of isolated fetal upper limb and trunk haemangioma suspected with antenatal 4D ultrasound scan at 28 weeks gestation. The baby was delivered through an emergency caesarean section at 36 weeks gestation followed by postnatal diagnosis and medical treatment with complete regression at five months of age.

Conclusion: Expectant approach of congenital haemangiomas with propranolol can result in rapid regression as early as five months of age.

Introduction

Congenital haemangiomas (CHs) are benign vascular tumours that are the most common of all congenital anomalies with an incidence of 0.3% [1]. They are of known etiology and usually involve the skin and the subcutaneous tissue but may also involve skeletal muscles, liver, bone, or central nervous system [1]. Approximately 75% of all hemangiomas are present at birth, with 60% occurring in the head and neck region [2]. Almost all of these lesions have no clinical importance and resolve spontaneously during infancy without complications [2]; however, large hemangiomas have been associated with several life-threatening complications, including high-output cardiac failure [2] or platelet trapping with severe consumption coagulopathy (Kasabach-Merritt syndrome) [3], profound hypotension leading to brain death [4] and also non-immune hydrops [5] and ultimately intrauterine fetal death [6].

The prenatal diagnosis of hemangiomas can be challenging, and optimal diagnosis is essential to ensure optimal antepartum and intrapartum care. We present a rare case of pregnancy complicated with a giant fetal hemangioma covering the right arm and upper trunk that was diagnosed prenatally with 4-dimensional (4D) sonography and Doppler studies.

Case Presentation

A 33-year-old primigravid lady was referred to Fetal Medicine Unit, Ain Shams University Maternity Hospital, Egypt, at 27 weeks gestation for 2-Dimensional (2D) ultrasound scan evaluation after her mid-T scan had been reported as normal. She had no relevant medical or surgical history and her Rhesus blood group was positive. The lady denied any use of medications during her pregnancy. Her family history disclosed no neural tube defects, mental restriction or any other birth defects.

On admission, 2D and Doppler ultrasonography were performed and revealed a singleton pregnancy with breech presentation consistent with 27 weeks and 4 days and an estimated fetal weight of 884 g (68th percentile). On 2D sonography, a large well-circumscribed right upper limb mass starting from the axillary level to the elbow that measured 10.1 × 5.4 × 3.8 cm and was cystic and multi-septated and having areas

of echogenic components mixed with predominantly hypoechoic structures (Figure 1a). Blood flow signals were also visualized in the mass with power Doppler sonography initially accomplished with the volume contrast imaging technique and showing high vascularity (Figure 1b and 1c). The spinal cord appeared to be intact and without evidence of neural tube defect or any connection of the mass to the neural tube. The rest of the fetal anatomy was within normal limits. The amniotic fluid index showed polyhydramnios, hence fetal echo was performed and there was no evidence of fetal hydrops, tricuspid regurgitation, or heart failure.

The lady was referred for 4D Ultrasonography at 28 weeks that showed huge swelling of the fetal right arm (Fig. 2) and TUI showing multiple cut section in the arm (Fig. 3).

Follow up scans every 2 weeks revealed progressively enlarging mass. A local multidisciplinary team (MDT) consisting of a Consultant Obstetrician, Consultant Neonatologist, Consultant Radiologist and Consultant Vascular Surgeon were involved in planning the mother's and the baby's care. The baby was delivered at 36 weeks by planned caesearian section after administration of corticosteroids course to enhance lung maturation. The newborn was generally well and neonatal initial physical examination (NIPE) hasn't shown any other abnormalities apart from a large, soft and compressible swelling of the right arm involving the upper 2/3 of the right forearm and right side of the chest wall and showing bluish discolouration of the overlying skin which was intact with no ulceration or signs of infection (Fig. 4). Postnatal superficial ultrasound of the right arm was performed and revealed presence of soft tissue mass extending to the chest wall and lower 2/3 of the forearm.

A diagnosis of congenital high flow haemangioma of the right arm, forearm and upper trunk was made. The baby received oral Propranolol in a dose of 2 mg/kg/day to help reducing the mass size and was followed up till the mass completely regressed at 5 months of age (Fig. 5).

The lady and her husband were informed, counselled and educated about the care, prognosis, possible complications, treatment options and when to seek help. A conservative approach was adopted awaiting regression and the family were asked to monitor the size of the masses with the possibility of planning surgical treatment would be if the mass failed to regress with expectant management.

Discussion

Haemangiomas are vascular lesions that occur when endothelial vascular cells form abnormally and multiply more than they should [7]. They can be congenital (CHs) which are rare comprising 3% of cases and present at birth, or infantile haemangiomas (IHs) where they appear later in infancy and affect 4-5% of newborn infants [1]. Congenital haemangiomas may solely occur or can be a part of multiple malformations, the most common of which is Kasabach-Merritt syndrome with associated thrombocytopenia and coagulopathy [3] and PHACES syndrome [8]. They can be complicated with life-threatening bleeding and high-output heart failure [9].

According to the International Society of the Study of Vascular Anomalies (ISSVA) classification, vascular malformations are subdivided according to vessel type into high-flow lesions as arteriovenous malformations, low-flow lesions that can be venous, lymphatic, or mixed and capillary malformations as port-wine stains. Also congenital haemangiomas (CH) were classified into rapidly involuting congenital haemangiomas (RICHs), partially-involuting congenital haemangiomas (PICHs) and non-involuting congenital haemangiomas (NICHs) [6]. CHs most commonly affect the liver and cutaneous tissues where differential diagnoses include pyogenic granulomas, macrocystic lymphatic malformation and malignant tumors as sarcoma, cutaneous neuroblastoma and lymphoma [10].

Big size CHs are often diagnosed antenatally using ultrasonography where they usually appear as hypoechoic lesions compared to HIs that appear postnatally as isoechoic lesions [11]. Also on Doppler ultrasound, CH appear as a vascular mass, composed mostly of veins unlike IH [4]. The imaging findings of fetal upper limb hemangioma tumors may widely overlap, the early detection and prenatal follow up of these tumors are very important for fetal, maternal, and postnatal care.

The main differentials in our case were CH and arterio-venous malformations (AVMs) of the upper limb and since biopsy may result in massive haemorrhage, diagnosis is most often based on characteristic radiological findings of usually well-defined mixed, solid lesions with hypervascularisation and fine granular calcifications [7]. Although hemangiomas have been reported hyperechoic or isoechoic [9], our case showed a mutli-septated complex cystic structure with low resistance flow together with peripheral dilated vessels (Fig. 1b) which suggested of CH using 2D, 4D sonography and Doppler studies at 27 weeks of gestation rather than AVMs which characteristically appear as echogenic dilated vascular channels replacing cutaneous tissue parenchyma with high-flow Doppler characteristics that lack arterial pulsation.

Management of congenital hemangiomas is patient-specific and highly dependent on the size, location, ability to involute, and presence of complications. RICHs are self-resolving and usually expectantly managed, unless complications such as ulceration, infection or bleeding occur. Once involution is complete, the fibro-fatty residual tissue may be excised to enhance cosmesis [12].

Treatment for patients with both high-flow and low-flow malformations is either expectant, medical, surgical, endovascular intervention or laser therapy. Only a low level of evidence supports the choice of treatment between these options, and the recurrence rates for large lesions are relatively high [13].

The majority of RICHs completely involute by 14 months however in our case the regression was faster and was complete by the age of 5 months with expectant management helped with oral propranolol [14].

Conclusion

Congenital upper limb haemangiomas, although rare, have to be diagnosed as early as possible in the antenatal period and to be differentiated from AVMs which have some similar sonographic criteria. Also,

associated life-threatening conditions can occur with CHs resulting in increased mortality and should always be considered.

Expectant approach with propranolol treatment has proved successful in this case with a good outcome showing rapid regression as early as five months of age rather than 12-14 months of age as described in previous reports. Further research using expectant management, would be warranted to support the findings of this case report.

Declarations

Funding

None

Conflicts of interest/Competing interests:

The authors declare that there is no conflict of interest regarding the publication of this article.

Ethics approval:

Ethical approval was waived at Ain Shams University as it is not an institutional requirement for publishing an anonymous case report.

Consent to participate

Not applicable.

Consent for publication:

The parents gave written informed consent for the case clinical details along with the images to be published.

Availability of data and material (data transparency):

Data material is available on request.

Code availability:

Not applicable.

Authors' contributions:

H Mamdouh: Data collection, performed imaging and reviewed images

MH Nasr El-deen: Has managed the case obstetrically and planned care and delivery

AH Farag: Manuscript editing and correspondence

References

1. Munden A, Butschek R, Tom W, et al. Prospective study of infantile hemangiomas: Incidence, clinical characteristics, and association with placental anomalies. Br J Dermatol. 2014;170(4):907-13.
2. Howell OM, Gumbiner CH, Martin GEO: Congestive heart failure due to giant cutaneous cavernous hemangioma. Clin Pediatr. 1984;23:504.
3. Currie BG, Schell D, Bowring AC: Giant hemangioma of the arm associated with cardiac failure and the Kasabach-Merritt syndrome in a neonate. J Pediatr Surg. 1991;26:734.
4. Silverman RA: Hemangiomas and vascular malformations. Pediatr Clin North Am.1991;38:811.
5. Stringel G, Mercer S. Giant hemangioma in the newborn and infant. Complications and management. Clin Pediatr. 1984;23:498.
6. Wildgruber M, Sadick M, Müller-Wille R, Wohlgemuth W. Vascular tumors in infants and adolescents. Insights Imaging. 2019;10:30.
7. Yu Y, Flint AF, Mulliken JB, Wu JK, Bischoff J. Endothelial progenitor cells in infantile hemangioma. Blood. 2004;103(4):1373-5.
8. Chamli A, Litaiem N. PHACE Syndrome. Stat Pearls. 2019.
9. Lewis D, Hachey K, Fitzgerald S, Vaidya R. Rapidly involuting congenital haemangioma of the liver. BMJ Case Rep. 2018:bcr224337.
10. Eberson S, Desai S, Metry D. A Basic Introduction to Pediatric Vascular Anomalies. Semin Intervent Radiol. 2019;36(2):149-60.
11. Gorincour G, Kokta V, Rypens F, Garel L, Powell J, Dubois J. Imaging characteristics of two subtypes of congenital hemangiomas: rapidly involuting congenital hemangiomas and non-involuting congenital hemangiomas. Pediatr Radiol. 2005;35:1178-85.
12. Delgadillo D, Carr L, Brgoch M, Ingraham J. Rapidly Involuting Congenital Hemangioma of the Hand. Eplasty. 2017;17:6.
13. Azma E, Razaghi M. Laser Treatment of Oral and Maxillofacial Hemangioma. J Lasers Med Sci. 2018;9(4):228-32.
14. Liang MG, Friedman IJ. Infantile and congenital hemangiomas. Semin Pediatr Surg. 2014;23:162-7.

Figures

Figure 1

2D sonography (a) and Doppler (b) images of fetal axilla and right arm at 27 weeks gestation showing cystic and multi-septated hypoechoic structures with areas of echogenic component (arrow) with high Doppler blood flow signals and high pulsed Doppler blood flow (c)

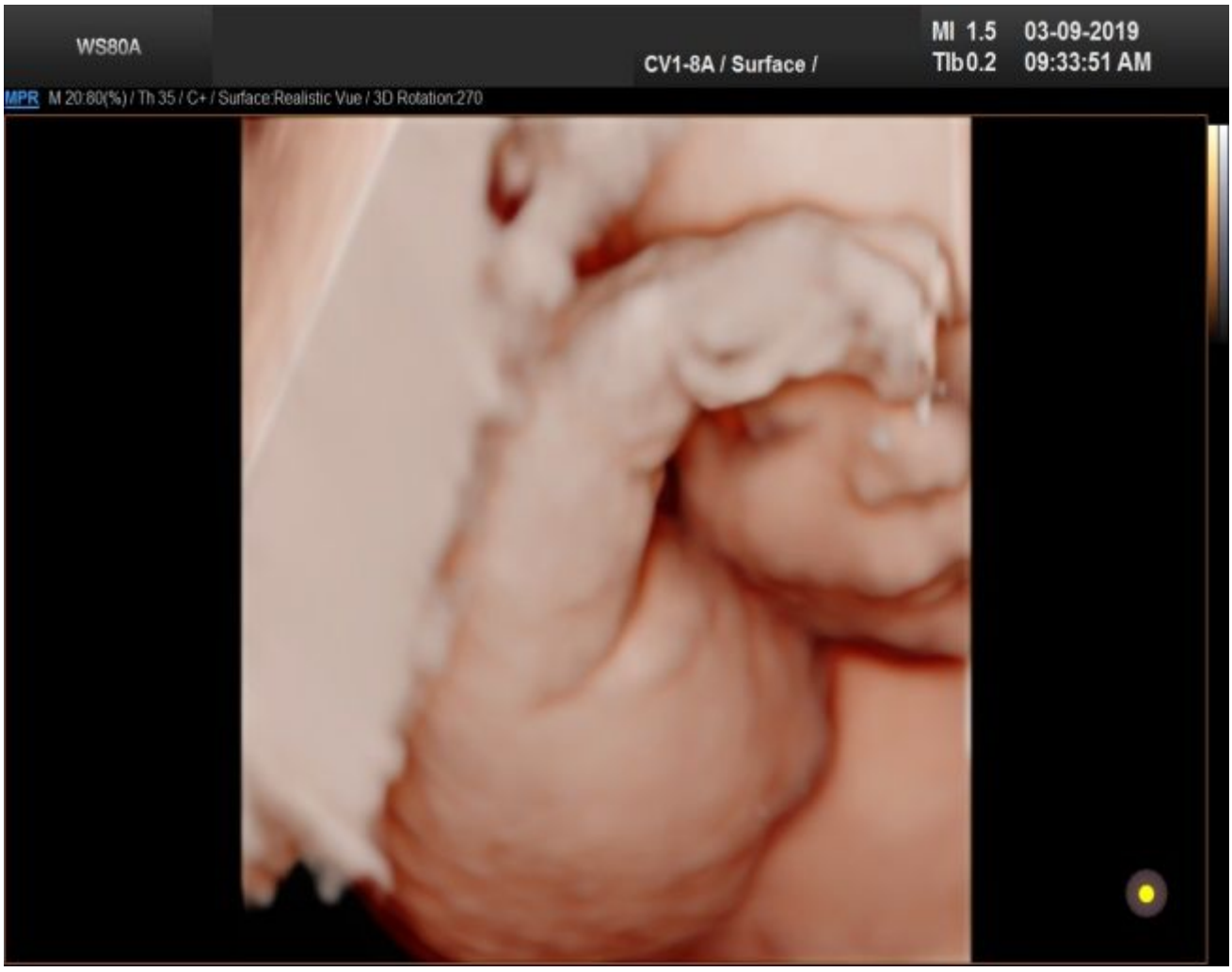

Figure 2

4D scan image showing hugely swollen right fetal arm

Figure 3

TUI showing multiple cut sections in the arm

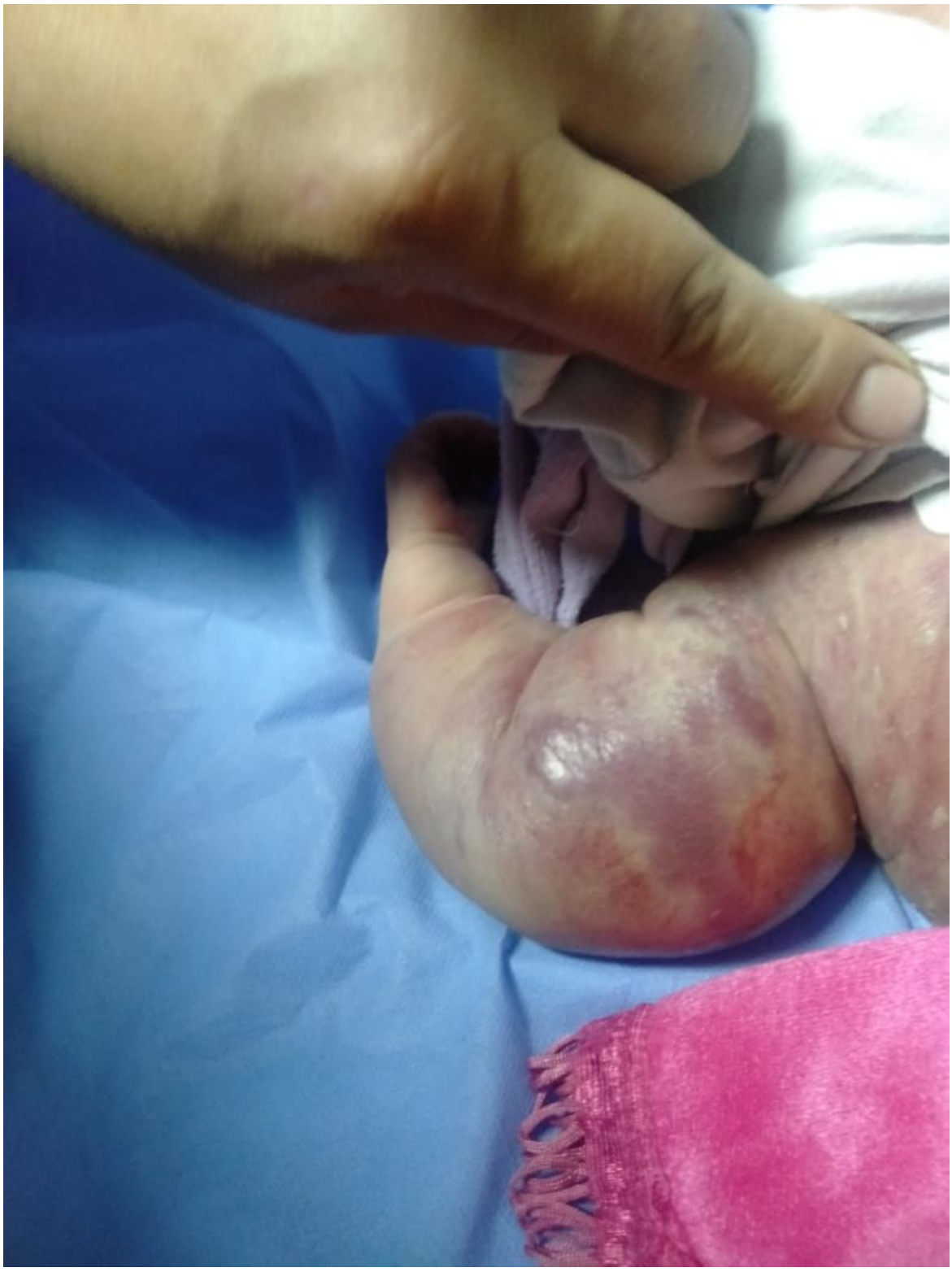

Figure 4

A medical photograph showing the right arm, forearm and trunk swelling shortly after delivery

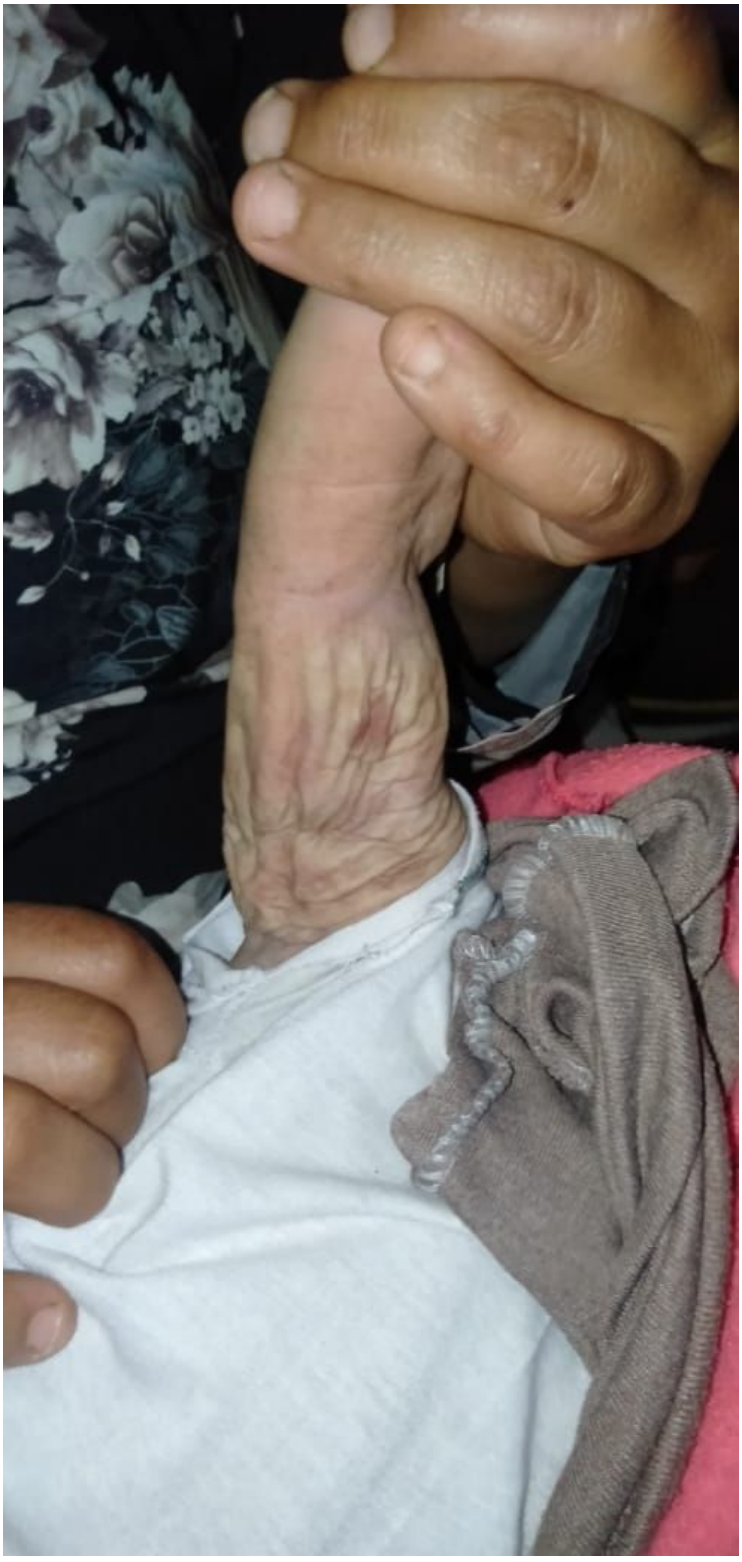

Figure 5

A medical photograph showing complete regression of the tumour at the age of 5 months

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- CAREchecklist.jpg