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CASE REPORT Open Access

Prenatal detection of Kaposiform Hemangioendothelioma with Kasabach–Merritt phenomenon: a case report

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Abstract

Background Kaposiform Hemangioendothelioma is a rare vascular neoplasm that typically presents in neonates and infancy. Most of the cases of Kaposiform Hemangioendothelioma are reported in neonates (60%) and infancy (93%), and very few cases have been reported prenatally. We describe here a case which was diagnosed prenatally in the 3rd trimester with rapid growth of upper limb and appearance of non-immune hydrops.

Case presentation A 25-year-old primigravida, an antenatally booked case, presented for routine third trimester level III scan at 34-week gestation. Level I scan for nasal bone and nuchal translucency at 12 weeks and level II anomaly scan at 20 weeks were normal. Level III ultrasound scan revealed increased echogenicity and hypertrophy of soft tissues of right upper limb and trunk involving both the subcutaneous and muscular compartments. Fetal middle cerebral artery Doppler, echocardiography and magnetic resonance imaging revealed fetal anemia, cardiac failure, and non-immune hydrops. These findings with hemodynamic changes and rapid evolution were suggestive of Kaposiform Hemangioendothelioma with Kasabach–Merritt phenomenon.

Conclusion The diagnosis of Kaposiform Hemangioendothelioma should be suspected in prenatal period if there is unilateral hypertrophy of limb which has evolved rapidly over a short interval of time and shows findings suggestive of Kasabach–Merritt Phenomenon. Prenatal presentation is life threatening if it is associated with poor fetal hemodynamics. Prenatal counseling in such cases should include advice on termination of pregnancy with emphasis on neonatal management.

Keywords Kaposiform Hemangioendothelioma, Kasabach–Merritt phenomenon, Prenatal, Fetal, Case report

Background

Kaposiform Hemangioendothelioma (KHE) is said to be a locally invasive rare vascular neoplasm that typically presents in infancy with a prevalence and incidence of 0.91 and 0.071 per 100,000 children, respectively [1]. When large, it may present with Kasabach–Merritt phenomenon (KMP) which is consumptive coagulopathy characterized by thrombocytopenia and hemolytic anemia [1, 2]. Most of the cases of KHE are reported in neonates (60%) and infancy (93%) [1], and very few cases have been reported prenatally. In the prenatal period, KMP complicating KHE may be suspected by the sudden growth of a limb or body part with appearance of cardiac failure and non-immune fetal hydrops. We describe here a case which was diagnosed prenatally in the 3rd trimester with rapid growth of upper limb and appearance of non-immune hydrops.

Case presentation

A 25-year-old primigravida, an antenatally booked case, presented for routine third trimester level III scan at 34-week gestation. Level I scan for nasal bone and

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nuchal translucency at 12 weeks and level II anomaly scan at 20 weeks were normal. Patient had no significant past medical history and was not on any medication for any chronic illness. Her blood group was O positive. All hematological and serological investigations were normal. Thyroid profile and TORCH (Toxoplasmosis, Rubella, Cytomegalovirus and Herpes simplex virus) panel was normal. Dual and quadruple marker tests were normal.

Radiological findings

Level III ultrasound scan revealed increased echogenicity and hypertrophy of soft tissues of right upper limb and trunk involving both the subcutaneous and muscular compartments (Fig. 1a and b). There was no enlargement of the bones. There was subcutaneous edema involving the lower chest and abdominal wall suggestive of hydrops. The rest of the limbs were normal, and there was no ascites or pericardial effusion. There was hypervascularity in the affected limb with dilated veins and hypertrophied arteries on color Doppler (Fig. 2). Fetal echocardiography revealed cardiomegaly with mitral insufficiency suggestive of cardiac failure. Liquor was increased with amniotic fluid index of 24 cm. Fetal MRI was done for further evaluation which showed soft tissue hypertrophy of the right upper limb and trunk (Fig. 3a, b and c) with subcutaneous edema involving the abdominal wall (Fig. 3b). The muscles appeared bulky with heterogeneous signal intensity on T2WI. In view of non-immune hydrops and increased liquor, patient was subjected to fetal middle cerebral artery (MCA) Doppler. There was increased peak systolic velocity (PSV) of 84.59 cm/s (Fig. 4) which was 1.73 times the multiples of median (MoM) for the gestational age suggesting fetal anemia. The imaging findings, taken together with rapid evolution and hemodynamic changes of fetal anemia, cardiac failure, and non-immune hydrops, were suggestive of Kaposiform Hemangioendothelioma (KHE) complicated with Kasabach–Merritt phenomenon (KMP). Parents were counseled, and termination of pregnancy was proposed by multidisciplinary team in view of rapidly developing hemodynamic changes. The pregnancy was terminated, and a female baby was delivered by cesarean section.

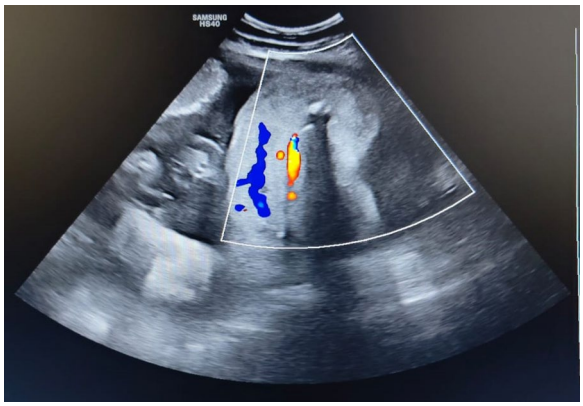

Fig. 2 Color Doppler image showing dilated vein and hypertrophied artery in right arm

Postnatal presentation

Postnatal examination of the neonate revealed marked hypertrophy of right upper limb and trunk with shiny purple discoloration of skin (Fig. 5). There was thrombocytopenia with platelet count of 15,000 per mcL and anemia with hemoglobin of 12 g/dL. Activated partial thromboplastic time was 42 s. There was no improvement in platelet count with repeated single-donor platelet (SDP) transfusion and sirolimus therapy. The neonate developed nasal and peroral bleed with intracranial hemorrhage on day 2, and it succumbed to the illness on day 7.

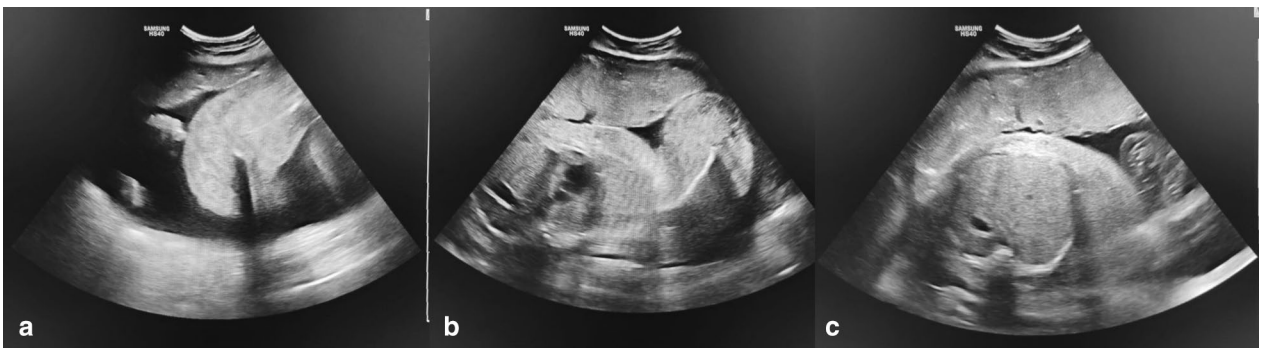

Fig. 1 **a**, **b** and **c** Unilateral hypertrophy of right upper limb. Ultrasound image showing increased echogenicity and soft tissue thickness of right forearm (**a**), arm (**b**), and upper trunk (**c**)

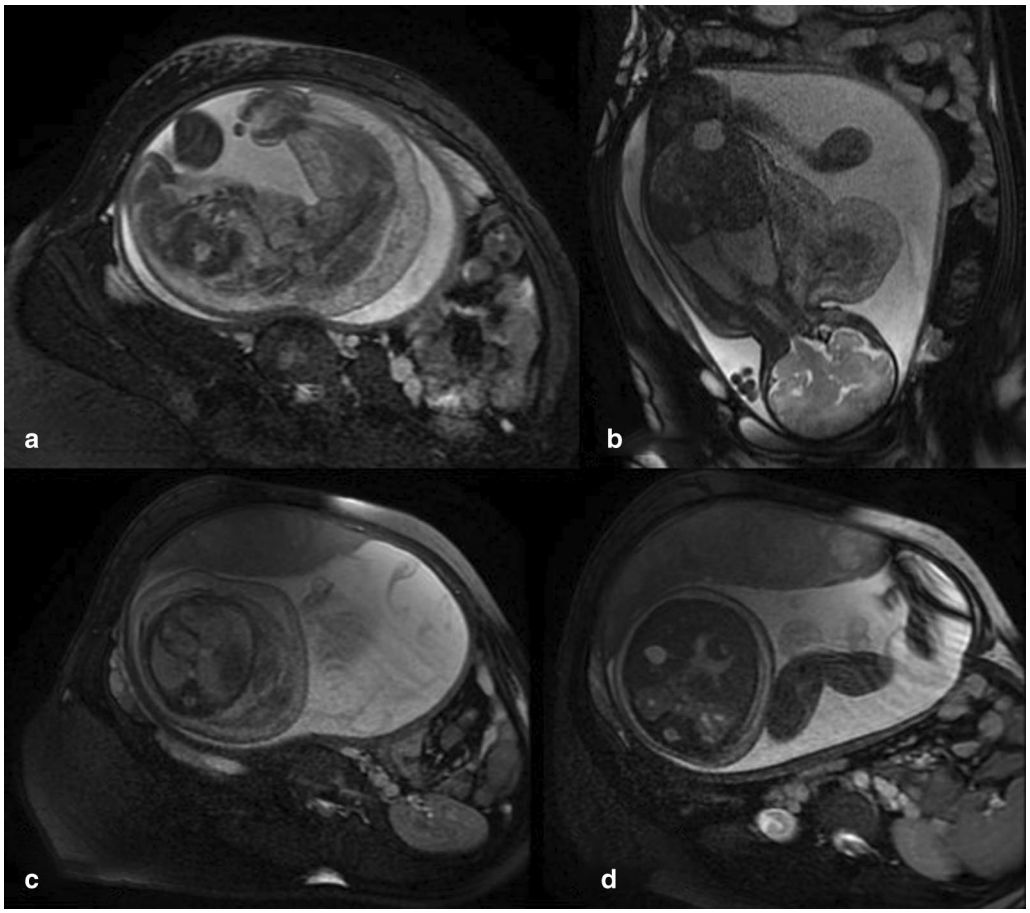

Fig. 3 **a**, **b**, **c** and **d** Fetal MRI showing hypertrophy and lymphedema of right upper limb and trunk (**a** and **b**) with subcutaneous edema of chest wall and abdomen (**c** and **b**)

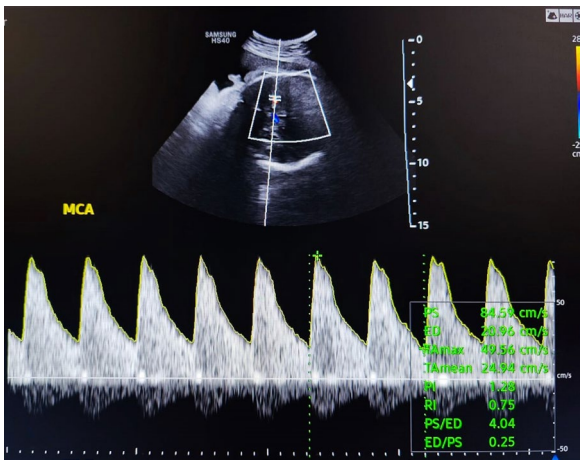

Fig. 4 Fetal MCA Doppler spectral waveform with peak systolic velocity of 84.59 cm/s which was 1.73 times the MoM for the gestational age

Discussion

Kaposiform Hemangioendothelioma is a disease of childhood which is rare and presents usually in neonatal period and infancy. It has been classified as locally aggressive vascular tumor by International Society for the Study of Vascular Anomalies. Clinically, children present with cutaneous swelling which may vary from an erythematous papule to a large soft tissue swelling involving the entire extremity or trunk which has a purplish appearance [3]. When extensive, KHE may be associated with Kasabach–Merritt phenomenon which is characterized by life-threatening consumptive coagulopathy, thrombocytopenia and microangiopathic hemolytic anemia.

Since KHE is a vascular tumor of neonates and infancy, it can also present in prenatal period as was seen in our case. The literature has many case reports of cases presenting in childhood, but very few cases have been reported in prenatal period [4]. The clinical appearance of these vascular lesions is so striking that it is not difficult to diagnose these cases in post-natal period. However, in the prenatal period they may not manifest in the

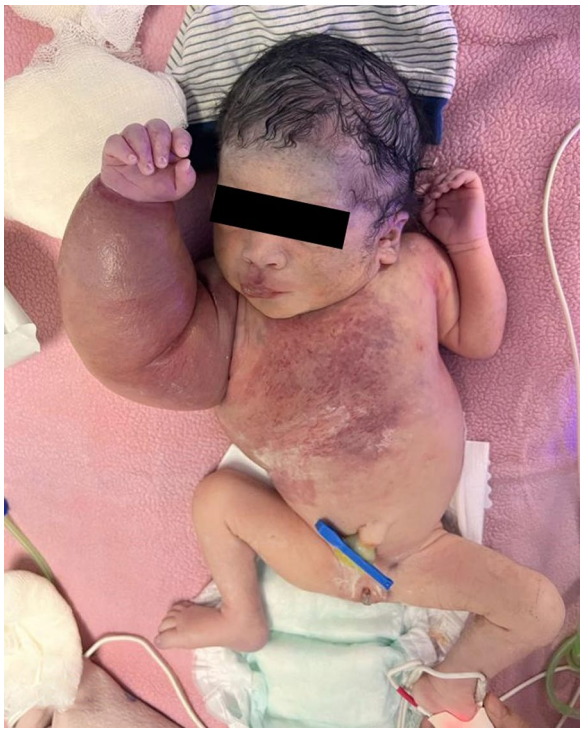

Fig. 5 Image of neonate showing marked hypertrophy and edema of right upper limb and upper trunk with shiny purple discoloration of skin

2nd trimester and remain undetected during level II scan as happened in this case. As these tumors have propensity to show rapid growth, they tend to manifest in 3rd trimester.

The prenatal findings on ultrasound are rapid enlargement of extremity or body part with increased soft tissue thickening involving the subcutaneous tissue and sometimes the muscular compartment (Fig. 1). The lesion shows hypervascularity with high flow on color Doppler (Fig. 2). These findings were seen in this case as the level II scan done at 20 weeks was normal and the follow-up scan done at 34 weeks showed the above-mentioned findings. If the lesion is extensive, these cases may be complicated by Kasabach–Merritt phenomenon in utero. The KMP is characterized by consumptive coagulopathy and severe thrombocytopenia due to trapping of platelets within the tumor and secondary development of hypofibrinogenemia. Hemolytic anemia develops due to sequestration of red blood cells within the tumor. All these findings were seen in our case postnatally. Prenatally, the KMP can be suspected if the fetus demonstrates features of anemia and cardiac failure with development of non-immune hydrops as was seen in this case. The fetal anemia can be suspected by studying the peak systolic velocity of MCA as seen in this case with increased PSV of 84.59 cm/s (Fig. 4).

MRI is the imaging modality of choice for prenatal cases as it can demonstrate the true extent of the lesion. MRI in this case showed extensive involvement of right upper limb and trunk with better delineation of the subcutaneous and muscle involvement (Fig. 3). It also demonstrated the subcutaneous edema due to non- immune hydrops better than the ultrasound.

When abnormality of unilateral hypertrophy of limb with hypervascularity and Kasabach–Merritt phenomenon is encountered in prenatal scans the differential diagnosis of KHE, Klippel Trenaunay syndrome, Kaposiform Lymphangiomatosis and tufted angioma should be considered. The Klippel Trenaunay syndrome is associated with venous phlebectasias, cutaneous capillary malformations and limb overgrowth which includes bony overgrowth [5]. Kaposiform Lymphangiomatosis presents with lymphatic malformations which are large and cystic in nature and most of the times involves mediastinum or retroperitoneum [6]. Tufted angiomas are morphologically similar in appearance in postnatal period but are less extensive and slow growing and less likely to present in prenatal period [7].

There is no consensus on prenatal management of these cases. If the lesion is detected early, then abortion can be offered to the patient after counseling. If it is detected in the last trimester, depending on the associated findings of KMP the pregnancy should be terminated and focus should be on postnatal management. These cases have high mortality with rates ranging from 20 to 40% according to available literature [4].

Conclusion

Kaposiform Hemangioendothelioma is a tumor of neonates and infants which is locally invasive and can present with Kasabach–Merritt phenomenon. The diagnosis of KHE in prenatal period is uncommon with very few cases reported in the literature. The diagnosis of KHE should be suspected in prenatal period if there is unilateral hypertrophy of limb which has evolved rapidly over a short interval of time and shows findings suggestive of KMP. Prenatal presentation is life-threatening if it is associated with poor fetal hemodynamics. Prenatal counseling in such cases should include advice on termination of pregnancy with an emphasis on neonatal management.

Abbreviations

KHE	Kaposiform Hemangioendothelioma
TORCH	Toxoplasmosis, Rubella, Cytomegalovirus and Herpes simplex virus
KMP	Kasabach–Merritt phenomenon
MRI	Magnetic resonance imaging

T2WI	T2 weighted imaging
MCA	Middle cerebral artery
MoM	Multiples of median

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Author contributions
SJS and SS performed the patient's ultrasound and color Doppler. PS and VM performed the patient's MRI. AG was involved in history taking and postnatal follow-up. All the five listed authors have been involved in drafting the paper and revising it critically for important intellectual content, and all were involved in the final approval of the version to be published. All the authors agree to be held accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors read and approved the final manuscript.

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Availability of data and materials
The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
The inclusion of patient data was done only after acquiring approval on the Institutional Ethical Committee of Base Hospital Delhi Cantt.

Consent for publication
Written and informed consent was obtained from the patient before use of the data pertaining to the patient in the study.

Competing interests
The authors declare that they have no competing interests.

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