Case report

Neonatal Kasabach-Merritt Syndrome (KMS): case report

Rathia SK [1], Phuljhele S [2], Gandhi A [3]

[1]Dr. Santosh Kumar Rathia, Assistant Professor, Department of paediatrics, Pt. JNM Medical College, Raipur; [2]Dr. Sharja Phuljhele, Professor and Head, Department of Paediatrics, Pt. JNM Medical College, Raipur; [3]Dr. Animesh Gandhi, Junior Resident, Department of Paediatrics, Pt. JNM Medical College and Dr. Bhim Rao Ambedkar Memorial Hospital, Raipur (C.G), India.

Address for Correspondence: Dr. Santosh Kumar Rathia, Email: drsantoshrathia84@gmail.com

Abstract

A 2days baby girl presented with congenital reddish-blue swelling over chest and abdomen with thrombocytopenia. The clinical, imagining and laboratory findings suggested the diagnosis of KMS. Oral steroid was started with initially needed platelet and FFP transfusions. Large thoracic hemangioma was non-amenable to surgical and radiological interventions. Vincristine was initiated after 2week trial of Prednisolone for persistent thrombocytopenia and non-regressing haemangioma. Over a week the lesion shrunk with improving platelet count. Vincristine was stopped after 4weeks in view of no further tumour shrinkage. The patient was discharged on steroid advised for 6-12 months and showed significant tumour regression at 3month.

Key words: Kasabach-Merritt syndrome, haemangioma, thrombocytopenia, corticosteroid, vincristine

Introduction

Hemangiomas constitute the commonest paediatric neoplasm. The management of these benign lesions not only depends upon their size and site but also on vascular characteristics or certain complications [1]. The triad of hemangioma, thrombocytopenia and consumption coagulopathy is known as Kasabach-Merritt phenomenon [1]. Hypotheses suggest that exposure of sub endothelial elements or abnormal endothelium within the haemangioma results in aggregation and activation of platelets with a secondary consumption of clotting factors [2].

This case report describes a neonate with Kasabach-Merritt syndrome (KMS). The clinical presentation of the case and its outcome with possible treatment modalities has been discussed in the light of similar cases/studies reported previously [3–8]

Case Report

A female baby born to a non-consanguineous couple presented with a diffuse reddish blue swelling over chest and upper abdomen, present since birth. Although normally delivered at term with no perinatal insults/ trauma, the baby got admitted in private nursery in view of suspected hematoma or ? vascular lump (see figure 1). She was referred from there on 2nd day to our NICU to rule out bleeding diathesis with thrombocytopenia (leading to enlarging hematoma). On examination at admission, baby had mild respiratory distress with RR-68/min, HR- 152/min, CFT<3sec, pulse- normal, Spo2- 99% with O2 with minimal retractions, but no lethargy. On local examination- reddish blue diffuse swelling with few petechiae was seen over chest and abdomen, with predominant bulge over right upper chest; the swelling was smooth, firm, non tender and warm to touch. Abdominal/other systemic examinations including growth parameters were normal. There were no dysmorphism/organomegaly/neurocutaneous markers.

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Figure-1: The 2 days old newborn having reddish blue diffuse swelling of chest and abdominal wall (most prominent bulge over right chest wall).

The initial investigations showed normal liver and renal function; but complete blood counts and coagulogram were suggestive of coagulopathy with negative sepsis screen and sterile blood cultures [normal INR=1.14(on post FFP sample), with FDP>30(raised) and reduced platelet count= 94,000/cumm].

With the clinical picture of hemangiomatous lesion over right chest wall and Ultrasound also confirming it as large heterogenous vascular/capillary hemangioma, possibility of hematoma (due to sepsis or DIC) was ruled out. Considering the case as KMS with large thoracic haemangioma, the baby was initiated on supportive treatment with oxygen, IVF, vitamin K and antibiotics.

Although the patient had received FFP outside and PT/INR was normal at admission, later in NICU course, she required frequent platelet transfusions for recurrent thrombocytopenia along with few plasma. Meanwhile, on day 3, the baby developed deep jaundice (indirect NNHB, DCT-ve) which resolved after phototherapy. On day4, the child had developed increased purpuric rash on swelling with reduced platelet (5000/cumm), deranged PT/APTT and reduced fibrinogen.

Platelets and FFP were transfused for worsening coagulopathy (DIC). The relevant haematological investigations carried out during 8 weeks of hospital stay have been shown in Table 1(annexure). CT findings of chest swelling were suggestive of a large cavernocapillary haemangioma. The overall clinical course, imaging and laboratory findings confirmed the diagnosis of KMS. USG cranium/abdomen and echocardiography ruled out other systemic vascular malformations.

Once CT angiography also failed to locate definite feeding vessel, the intervention radiologist denied for arterial embolization and in view of diffuse haemangioma with risk of fatal hemorrhage, pediatric surgeons and radiotherapist also denied for surgical or laser therapy. Finally, we decided to watch its course on medical management only. On drug

therapy, first tried was 2week trial of oral Prednisolone (2mg/kg/d) and observation was static size of haemangioma with somewhat improved platelet.

But, later in view of non regression of tumour size and recurrent thrombocytopenia, after consulting oncologist for chemotherapy options, intravenous Vincristine (0.5mg/kg/day once weekly) was opted along with oral steroid. One week after the start of Vincristine, the size of the lesion decreased almost to half and platelet count increased to 50,000/cumm, which over 2-3 weeks improved up to 65000/cumm. Vincristine, in the same dose, was continued for total 4 weeks and then stopped as later no further improvement in lesion size or platelet count was noticed.

Table : 1

Days of life (age)	Platelet count	Hemoglobin	WBC	Other specific lab investigations
Day2 (morning sample; at admission)	94000	13.6	14800	Initial PT=15sec, INR=1.14(normal value ?post FFP), but FDP(fibrin degradation products)=>30(raised), CRP=12.2mg/L, uESR=10mm at 1Hr
Day2 (evening)	47000	12.3	10600	Serum urea=33mg/dl, creatinine=0.9 mg/dl, SGOT=24 IU/L, SGPT=10 IU/L, ALP=105 IU/L, total protein=5.1g/dl, albumin=3.4g/dl, serumCa+=10.6mg/dl, sodium=130meq/l, potassium =3.7 meq/l
Day3	14000	8.9	16700	S.Bil- T=21.5 mg/dl(Direct=1.8 mg/dl), DCT- Negative, Reticulocytes=2%
Day4	5000	10.4	12500	PT=25sec(control: 13sec), APTT=60sec(control: 25-30sec) Fibrinogen=19.7mg/dl(low)
Day7	35,000	12.5	8400	BLOOD C/S – sterile
Day15	5500	8.3	7400	-
Day21	50000	10.8	8000	
Day28	54000	8.2	9300	-
Day45	65000	9.9	19700	-
2months (at Discharge)	115000	12.3	10000	-

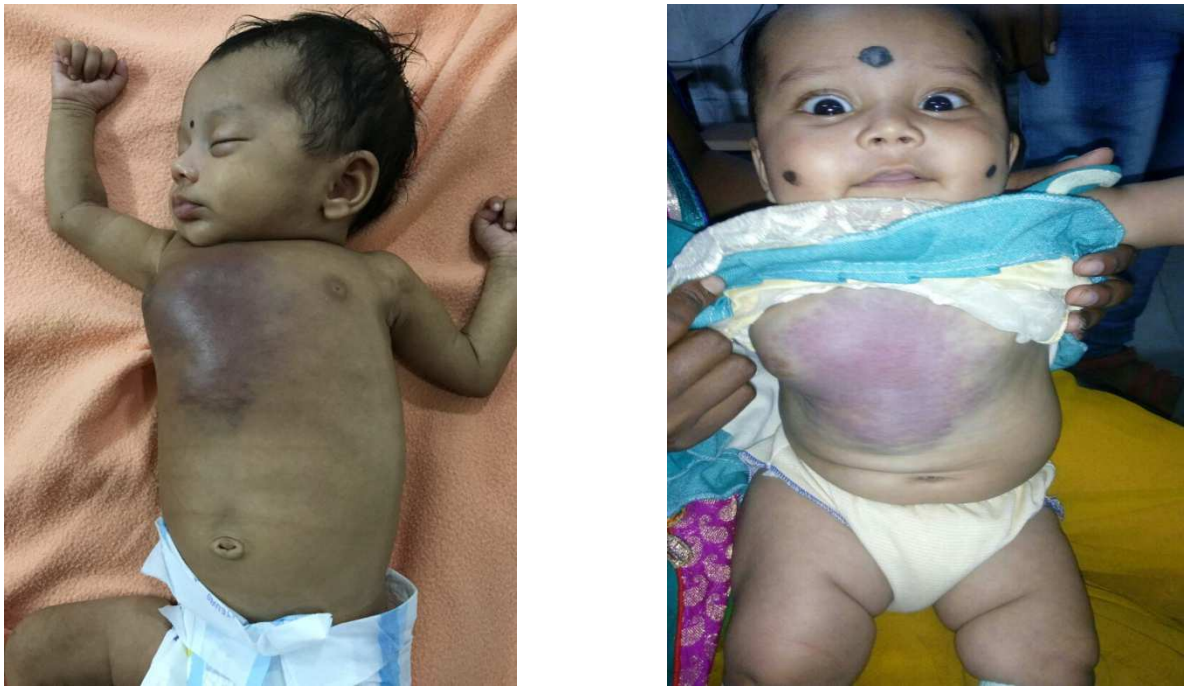

Figure 2: same baby at discharge (2months) and at 4months follow-up

Over 2 months of NICU stay, the baby had considerable regression of haemangioma confined to right upper chest wall (figure 2) and better platelet count persistently recorded >50000/cumm for last 2-3 weeks before discharge. We planned to continue oral Prednisolone for 6-12 months and on follow up more significant tumour regression was observed by 4 months.

Discussion

Kasabach-Merritt syndrome shows wide variation in its response to different treatment modalities including compression, steroids, chemotherapy, interferon, embolization, sclerotherapy, laser, radiation or surgery [1–6, 9–12]. Currently there is no specific guidelines [13], but various multimodal (stepwise) approaches have been suggested till date [4,7,8]. Several researchers agree that most patients with KMS start responding to steroids within few days of treatment [4]. The angiogenetic character of the syndrome indicates that chemotherapy is a logical treatment. Enjolras and associates have reported that several steroid non responders show dramatic response to vincristine [5].

A multicenter study conducted in US showed that vincristine is a safe and effective treatment in the management of KMS [6].. Haisley-Royster reported that the increase in platelet count precedes the regression in tumor size [6].

Usually the mainstay of the treatment is clinical stabilization, tumour regression by medical therapy and/or radio-surgical ablation of hemangioma, the primary culprit. Surgical excision is recommended for single cutaneous lesions or multiple lesions in the spleen (splenectomy) or liver (wedge resection/hepatectomy) [2,11,12].

Any large congenital vascular tumour in newborn with thrombocytopenia with/without significant bleeds must be differentiated from hematoma secondary to sepsis/DIC or liver dysfunction, as hemangioma-thrombocytopenia (KMS) syndrome is not very rare; which warrants early appropriate multidisciplinary management. In the index case, unusually diffuse chest-wall lesion with complex vascularity made it non amenable to invasive radio-surgical interventions; so steroid and vincristine only could be started. Even then considerable response on both platelet count and tumour regression was observed by 4 months follow up.

Conclusion

Initial four weeks vincristine with prolonged oral steroid may be the suitable option in KMS for expectant therapy in selected cases with non-feasibility of surgical/radiological interventions.

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