Radiotherapy for Kasabach-Merritt (KM) phenomenon in an adult patient with benign vascular neoplasm- a case report

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Abstract

Introduction: Kasabach Merritt (KM) phenomenon is consumption coagulopathy in a vascular neoplasm. The combination of haemangioma, thrombocytopenia, and coagulopathy is termed Kasabach-Merritt phenomenon. Vascular neoplasms are more common in children wherein they may resolve spontaneously. Kaposi form hemangioendothelioma and tufted angioma are most common vascular neoplasms in children associated with Kassabah-Merritt phenomenon but are found with other vascular neoplasms too. These lesions are rare in adults. Radiotherapy was the treatment of choice in the past in children but not considered in the absence of life threatening complication presently due to its late effects. The other modalities that are tried are surgery, embolization, laser surgery, and pharmacologic agents such as steroids, interferons, and cyclophosphamide. We report a persistent 10 years' long response of a benign vascular neoplasm with Kasabach-Merritt phenomenon and life threatening bleeding to radiotherapy in an adult with asymptomatic late side effects. **Case report:** A 35-year-old lady from south India presented with a 15x15 cm left lower back swelling and hemorrhagic pleural effusion and thrombocytopenia. She received radiotherapy of 40 Gy in 20 fractions to the swelling. Swelling, effusion and thrombocytopenia resolved and remain resolved for 10 long years. She did have asymptomatic late side effects of radiotherapy. **Conclusion:** Though radiation therapy is not considered in children with KM phenomenon, it could be considered as an option of treatment in adults with KM phenomenon.

Key words: Coagulopathy, Haemangioma, Kasabach-Merritt phenomenon, Radiotherapy, Vascular neoplasm

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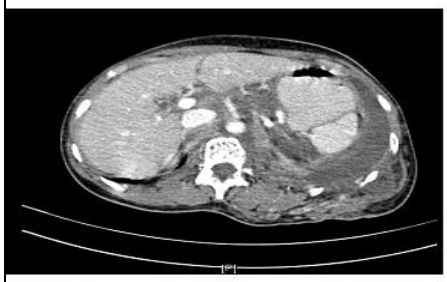
Introduction: Vascular neoplasm is commonly encountered in paediatric age group and majority involutes by 8 to 10 years of age. One of the life-threatening complications arising in these vascular neoplasms is bleeding due to consumption coagulopathy, which is known as the Kasabach-Meritt phenomenon [1]. This phenomenon is mostly observed in neonates and rarely in adults. Historically, radiotherapy was widely used to treat these vascular neoplasms with Kasabach-Merritt phenomenon, but it has been slowly replaced by alternative treatment strategies[2][3][4]. This report describes an adult patient with a giant benign vascular neoplasm presenting with life-threatening bleeding due to Kasabach-Merritt phenomenon and was treated with radiotherapy.

Case presentation: The patient was a 35-year-old lady, presented in January 2003 with a gradually enlarging swelling in the left side of lower back of six years duration. She had no vascular lesion during her childhood. Physical examination revealed a large, tender, compressible, soft tissue swelling involving the entire lumbar region on the left side. The maximum dimension of the lesion was about 15x15 cms. It was not pulsatile, and there was no associated bruit, thrill, skin discoloration or purpura. Respiratory system examination revealed reduced breath sounds in the left base. Rest of the examination revealed no abnormality. Hematologic laboratory results revealed a platelet count of 42000 cell/cumm, haemoglobin of 9.4 g/dL and a minimally elevated prothrombin time. CT imaging revealed multiple vertebral haemangioma and a large mass in the subcutaneous plane of the left lumbar region, extension into the muscles of the posterior abdominal wall with pre-and para-vertebral component in the lower thoracic and lumbar region, involvement of the root of the mesentery and lesser omentum and a large left pleural effusion (Figure1).

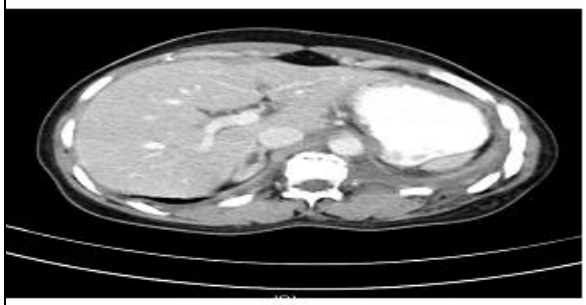
Figure 1: Pre RT CT thorax in 2003



Lobulated mass in the subcutaneous plane in the left lumbar region with left pleural effusion

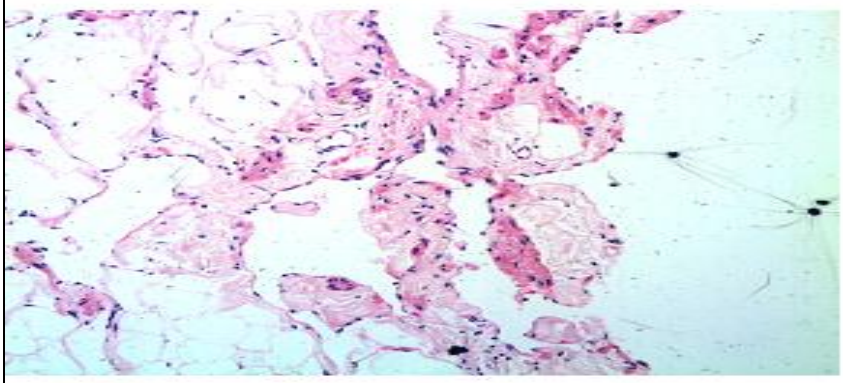
The arterial supply was from lumbar & intercostal arteries. Venous drainage was into paravertebral venous plexus, azygos & hemiazygos vein. Pleural aspirate revealed hemorrhagic fluid indicating bleeding into pleural cavity. Biopsy was deferred in view of risk of life-threatening bleeding. Radiotherapy was given to the swelling using telecobalt megavoltage beam to a total dose of 40 Gy in 20 fractions. Two days after completion of radiotherapy, she presented with increasing breathlessness. Laboratory results showed further decrease in platelets and anaemia. She was supported with packed red cells, fresh frozen plasma and platelet transfusions. She required an intercostal drainage to relieve breathlessness due to hemorrhagic effusion. Thereafter, platelet counts increased to normal values in 5 weeks' time and the lesion showed regression. Two years after the treatment, she remained well with no clinically palpable swelling though CT imaging showed residual mass and the pleural effusion had resolved (Figure 2). Biopsy of the residual lesion showed a benign angiomatous lesion. (Figure3)

Figure 2: Post RT CT thorax in 2005



Significant reduction in the soft tissue component

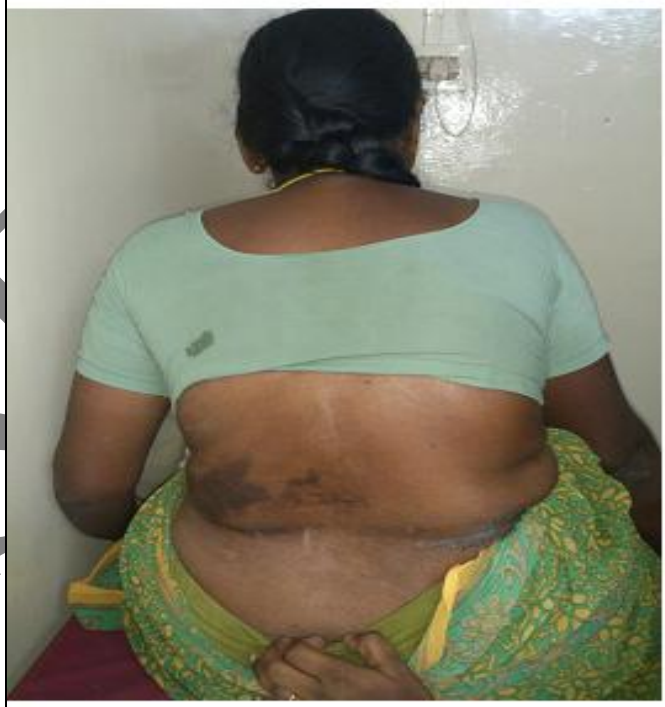
Figure 3: Histopathology



Lesion showing vascular proliferation with smooth muscle in the wall. HE 200x.

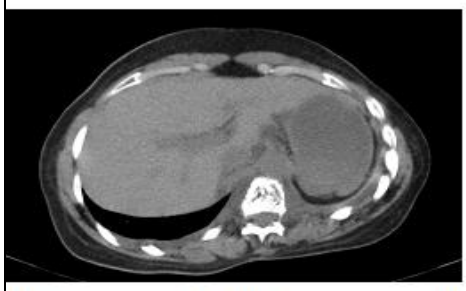
She was asymptomatic with no swelling palpable and normal platelet count in her last followup in March 2013(Figure 4). CT scan showed similar findings as before along with further increase in volume loss, pleural thickening and post inflammatory changes in left lung (Figure5).

Figure: 4 Clinical photograph 2013



Skin discoloration and no palpable swelling

Figure: 5 CT thorax March 2013



Volume loss in left hemithorax with pleural thickening and postinflammatory changes left lung

She was found to have small left kidney, volume loss of left hemi-thorax and minimal kyphosis as probable late effects of radiotherapy but she was asymptomatic and these did not affect her quality of life and her serum creatinine was normal. She was brought dead to emergency room following a seizure of unknown cause in July 2013.

Discussion: Kasabach- Merritt phenomenon is characterised by profound thrombocytopenia, microangiopathic hemolytic anemia, a consumptive coagulopathy and an enlarging vascular lesion either a Kaposiform haemangioendothelioma or a tufted angioma. Consumption coagulopathy in a vascular neoplasm was first described by Kasabach and Merritt in 1940, and they also reported striking response to radiotherapy[1]. Since then, consumption coagulopathy occurring in diverse clinical settings, involving a variety of vascular neoplasms have been broadly designated as Kasabach-Merritt syndrome. It is now known that Kasabach-Merritt syndrome is clinically heterogeneous, commonly associated with kaposiform hemangioendothelioma, tufted angioma and less frequently in other vascular neoplasms[5][6]. Due to this heterogeneity, it is no longer considered a syndrome but a phenomenon[7].

Radiotherapy was once a widely used treatment for vascular neoplasms, but it is no longer considered appropriate in the absence of life-threatening complications due to the late effects[8] associated with it. Other treatment modalities include surgical excision, embolisation, laser surgery and pharmacological agents like steroids, interferon alpha, vincristine and cyclophosphamide. There are no prospective studies or randomized control trials comparing the different modalities, hence, treatment for Kasabach-Meritt phenomenon has been largely empirical [5].

In literature, the radiotherapy dose used ranges from 8 Gy to 20 Gy and 10 Gy is reported to be sufficient by many [2][3][4]. Response to radiotherapy is 60-100% [4]. This dose response has been predominantly observed in neonates with Kasabach-Merritt phenomenon and the dose response in adults is not clear since this phenomenon is rare in adults. A trend towards greater percentage of complete response with higher doses of radiotherapy (30 Gy and greater) has been observed for the treatment of vascular neoplasms in adults[9]. Vascular neoplasms in older patients have been treated with higher doses since they are regarded less responsive to radiotherapy[8][10]. We used higher doses since dose response has not been studied in adults and due to the presence of life-threatening intrapleural bleeding, which could become catastrophic if adequate response was not achieved with lower doses of radiotherapy. The objective response to radiotherapy in our patient was slow and so was the platelet recovery and restoration of coagulation profile. Complete resolution of the thrombocytopenia and coagulopathy occurred with good regression of the neoplasm, but residual lesion remained despite the higher doses of radiation used.

Conclusion

We conclude that in appropriately selected patients, radiotherapy is an effective and a valuable treatment for benign vascular neoplasms with Kasabach-Merritt phenomenon.

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References

1. Kasabach HH and Merritt KK. Capillary hemangioma with extensive purpura: report of a case. Am J Dis Child 1940; 59: 1063-70.
2. Mitsuhashi N, Furuta M, Sakurai H, Takahashi T, Kato S, Nozaki M, Saito Y, Hayakawa K, Niibe H. Outcome of radiation therapy for patients with Kasabach-Merritt syndrome. Int J Radat Oncol Biol Phys 1997; 39:467-73.
3. Ogino I, Torikai K, Kobayasi S, Aida N, Hata M, Kigasawa H. Radiation therapy for life-or function-threatening infant hemangioma. Radiology 2001:218:834-9
4. Hesselmann S, Micke O, Marquardt T, Baas S, Bramswig JH, Harms E, Willich N. Kasabach-Merritt syndrome: a review of the therapeutic options and a case report of successful treatment with radiotherapy and interferon alpha. Br J Radiol. 2002; 75(890): 180-4.
5. Hall GW. Kasabach-Merritt syndrome: pathogenesis and management. British Journal of Haematology 2001; 112(4):851-862.
6. Choi JW, Na JI, Hong JS, Kwon SH, Byun SY, Cho KH, Youn SW, Choi HS, Park KD, Park KC. Intractable tufted angioma associated with Kasabach-merritt syndrome. Ann Dermatol. 2013 Feb;25(1):129-30
7. Sarkar M, Mulliken JB, Kozakewich HPW, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg 1997; 100:1377-86
8. El-Dessouky M, Azmy AF, Raine PA, Young DG. Kasabach-Merritt syndrome. J Pediatr Surg 1998;23: 109-11.
9. Schild SE, Buskirk SJ, Frick LM, Cupps RE. Radiotherapy for large symptomatic hemangiomas. Int J Radat Oncol Biol Phys 1991; 21: 729-735.
10. Dutton SC, Plowman PN. Paediatic hemangiomas: the role of radiotherapy. Br J Radiol 1991;64:261-269