

## Hybrid SPECT-CT with $^{99m}\text{Tc}$ -labeled red blood cell in a case of blue rubber bleb nevus syndrome: added value over planar scintigraphy

Kalpa Jyoti Das, Punit Sharma, Niraj Naswa, Ramya Soundararajan, Rakesh Kumar, Chandrasekhar Bal, Arun Malhotra

### ABSTRACT

Blue rubber bleb nevus syndrome (BRBNS) is a rare clinical entity characterized by multiple venous malformations (hemangiomas) of the skin and gastrointestinal tract. These hemangiomas usually cause episodes of occult gastrointestinal bleeding leading to iron deficiency anemia, and also carry a significant potential for serious hemorrhage. The  $^{99m}\text{Tc}$ -labeled red blood cell scintigraphy has traditionally been utilized in the localization of occult bleeding sites in patients with suspected vascular malformations, angiodysplasia, and Meckel's diverticulum. We report the incremental value of  $^{99m}\text{Tc}$ -labeled red blood cell hybrid single-photon emission computed tomography-computed tomography (SPECT-CT) over planar scintigraphy alone in a 12-year-old female patient with BRBNS.

**B**lue rubber bleb nevus syndrome (BRBNS) is a rare vascular anomaly consisting of multifocal venous malformations most commonly involving the skin, soft tissues, and gastrointestinal tract. The gastrointestinal lesions of BRBNS are the most clinically relevant malformations and carry a significant potential for serious bleeding. William Bennet Bean first coined the term "BRBNS" in 1958, and only 200 cases have been described since then (1). A few reports have described the use of  $^{99m}\text{Tc}$ -labeled red blood cell (RBC) planar scintigraphy in BRBNS patients to localize the site of bleeding (2–5). To the best of our knowledge, however, the role of single-photon emission computed tomography-computed tomography (SPECT-CT) in examining such patients has not been evaluated. Here, we report the value of  $^{99m}\text{Tc}$ -labeled RBC hybrid SPECT-CT over planar scintigraphy alone in a patient with BRBNS.

### Case report

A 12-year-old girl presented with repeated episodes of lower gastrointestinal bleeding that had required multiple blood transfusions in the past. She also had a history of multiple cutaneous swellings since birth. A hemogram analysis revealed features of severe iron deficiency anemia. A capsule endoscopic examination of the gastrointestinal tract revealed multiple vascular blebs arising from the second part of the duodenum and extending until the caecum.

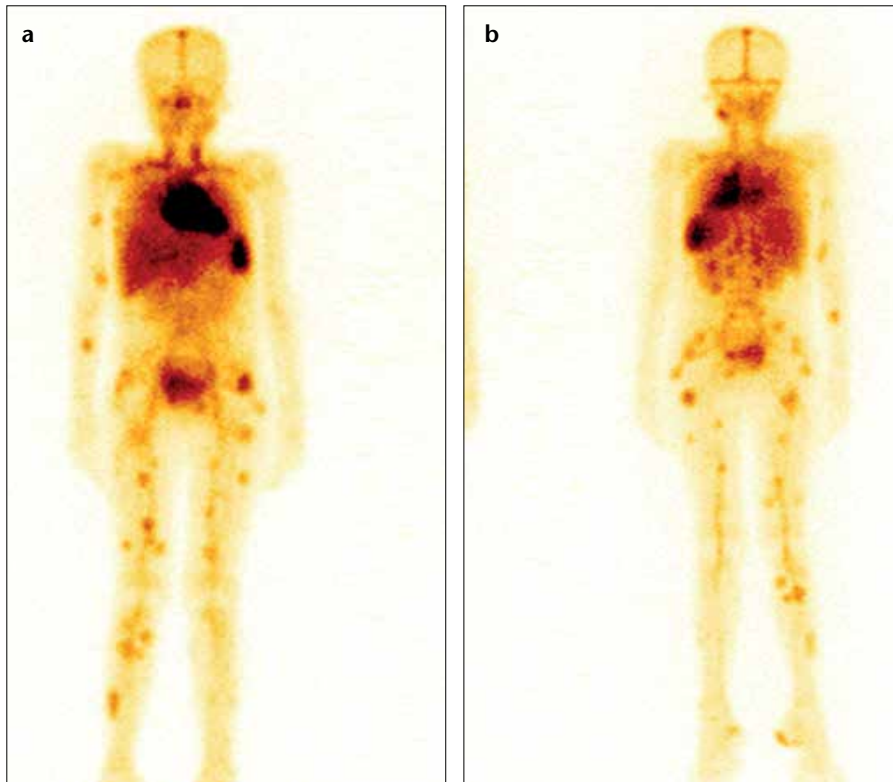
Subsequently, she underwent multiple enterotomies and colostomies with excision of blebs and wedge resection of multiple small bowel segments. Multiple blebs were found on various organs: a total of 54 blebs in the small intestine, 12 in the large intestine, three on the surface of liver, one involving the lesser curvature of stomach, and two involving the pancreas. Histopathological assessment of the lesions revealed features of venous cavernous hemangiomas. In addition, the colonic lesions showed areas of thrombosis and recanalization, a characteristic feature of BRBNS.

A follow-up  $^{99m}\text{Tc}$ -labeled RBC study was planned three months later to assess any residual lesions. In vitro  $^{99m}\text{Tc}$ -labeled RBC (370 MBq; 10 mCi) prepared using a standard kit was injected intravenously, and serial images were acquired (6). At 1.5 hour, images showed multiple foci of abnormal  $^{99m}\text{Tc}$ -labeled RBC accumulation in the abdomen, pelvis, right upper extremity, and both lower extremities, consistent with BRBNS (Fig. 1). SPECT-CT was performed to delineate the abdominal lesions and differentiate musculoskeletal lesions from intra-abdominal vascular lesions, if any. SPECT-CT revealed these lesions to be confined to muscles only (Fig. 2). The gastrointestinal lesions were successfully resected.

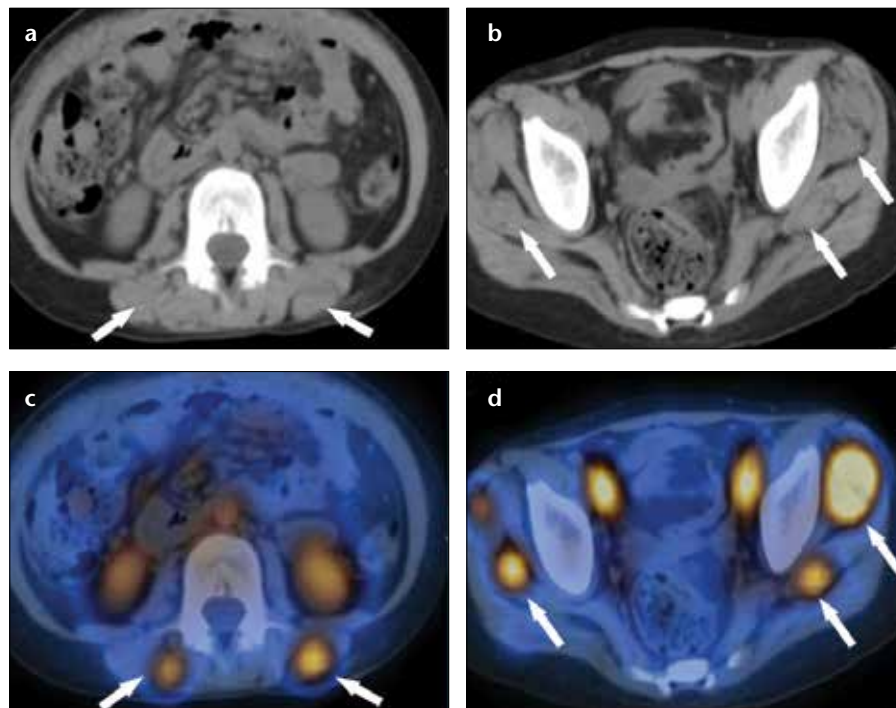
From the Department of Nuclear Medicine (K.J.D., P.S., N.N., R.S., R.K. ✉ [rkphulia@yahoo.com](mailto:rkphulia@yahoo.com), C.B., A.M.), All India Institute of Medical Sciences, New Delhi, India

Received 8 March 2012; revision requested 29 March 2012; revision received 20 May 2012; accepted 9 June 2012

Published online 13 December 2012  
DOI 10.4261/1305-3825.DIR.5811-12.2



**Figure 1. a, b.** Anterior (a) and posterior (b)  $^{99m}\text{Tc}$ -labeled red blood cell planar scintigraphy images. Multiple focal areas of increased  $^{99m}\text{Tc}$ -labeled red blood cell accumulation equal to that of cardiac blood pool activity were noted in the body, including the abdomen and pelvis. These findings were suggestive of hemangiomas of blue rubber bleb nevus syndrome. However, it was difficult to accurately localize the site of hemangiomas on planar scintigraphy.



**Figure 2. a–d.** Axial noncontrast CT images of abdomen (a) and pelvis (b) show ill-defined lobulated lesions in the muscles (arrows), which are more clearly shown in SPECT-CT. Corresponding axial SPECT-CT images of the abdomen (c) and pelvis (d) localize the focal  $^{99m}\text{Tc}$ -labeled red blood cell accumulation to lobulated lesions seen in muscles (arrows), suggesting intramuscular hemangiomas. No abnormal  $^{99m}\text{Tc}$ -labeled red blood cell accumulation was seen in the abdominal viscera, suggesting complete removal at surgery.

## Discussion

BRBNS is a congenital cutaneous and gastrointestinal hemangiomatosis. Treatment of this syndrome is based on pharmacological or surgical intervention to remove the bleeding source. Prognosis depends on the extent of visceral organ involvement, particularly the gastrointestinal tract. Appropriate detection and treatment of culprit lesions is therefore important in such patients to avoid potential bleeding catastrophes (7).

The  $^{99m}\text{Tc}$ -labeled RBC nuclear scintigraphy has been traditionally utilized to locate the site of active bleeding in patients with gastrointestinal hemorrhage (2–5). The added advantage of this technique is the detection of sites other than the gastrointestinal tract (e.g., viscera, musculoskeletal) that may also be involved (5). RBC scintigraphy reveals abnormal areas of tracer uptake in the involved regions; however, it is difficult to discriminate between visceral and cutaneous lesions based on planar scintigraphy alone. Addition of SPECT-CT assisted in exact anatomical localization and in differentiating vascular from musculo-cutaneous lesions, thereby preventing patients from undergoing unnecessary surgical interventions.

In conclusion, even with appropriate surgical treatment, the possibility of incomplete resection and residual disease remains in patients with BRBNS, which could continue to produce symptoms. In addition, new gastrointestinal lesions continue to evolve during the course of disease; hence patients need periodic gastrointestinal monitoring and hematologic follow-up (8). Findings presented in this case demonstrate that  $^{99m}\text{Tc}$ -labeled RBC hybrid SPECT-CT appears to be best suited for both purposes: the detection of residual lesions and the localization of possible recurrence.

## Conflict of interest disclosure

The authors declared no conflicts of interest.

## References

1. Elsayes KM, Menias CO, Dillma JR, Platt JF, Willatt JM, Heiken JP. Vascular malformation and hemangiomatosis syndromes: spectrum of imaging manifestations. *AJR Am J Roentgenol* 2008; 190:1291–1299. [CrossRef]
2. Yarlagadda R, Menda Y, Graham MM.  $\text{Tc-}^{99m}$  red blood cell imaging in a patient with blue rubber bleb nevus syndrome. *Clin Nucl Med* 2008; 33:374–376. [CrossRef]
3. Gallego Peinado M, de Arcocha Torres M, López Casado MA, Custosio Rebollo Aguirre

- A, Santiago Chinchilla A, Llamas Elvira JM. Usefulness of labelled red-cell scintigraphy in Blue Rubber-Bleb Nevus syndrome. *Rev Esp Med Nucl* 2009; 28:18–21. [\[CrossRef\]](#)
4. Hales K, Connolly LP, Drubach LA, Mulliken JB, Fishman SJ. Tc-99m red blood cell imaging of blue rubber bleb nevus syndrome. *Clin Nucl Med* 2000; 25:835–837. [\[CrossRef\]](#)
  5. Adams BK, Attia HMA. Technetium-99m labelled red blood cell blood pool imaging versus contrast venography in a patient with extensive blue rubber bleb naevi. *Postgrad Med J* 2003; 79:935. [\[CrossRef\]](#)
  6. Smith TD, Richards P. A simple kit for the preparation of 99mTc-labeled red blood cells. *J Nucl Med* 1976; 17:126–132.
  7. Dobru D, Seuceha N, Dorin M, Carienu V. Blue rubber bleb nevus syndrome: case report and literature review. *Rom J Gastroenterol* 2004; 13:237–240.
  8. Crosher RF, Blackburn CW, Dinsdale RC. Blue rubber-bleb naevus syndrome. *Br J Oral Maxillofac Surg* 1988; 26:160–164. [\[CrossRef\]](#)