Hybrid SPECT-CT with 99mTc-labeled red blood cell in a case of blue rubber bleb nevus syndrome: added value over planar scintigraphy

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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 99mtechnetium (99mTc)-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 99mTc-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.

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 99mTc-labeled RBC study was planned three months later to assess any residual lesions. In vitro 99mTc-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 99mTc-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 musculocutaneous 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.
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 99mTc-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 musculoskeletal 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 99mTc-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

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Figure 1. a, b. Anterior (a) and posterior (b) 99mTc-labeled red blood cell planar scintigraphy images. Multiple focal areas of increased 99mTc-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 99mTc-labeled red blood cell accumulation to lobulated lesions seen in muscles (arrows), suggesting intramuscular hemangiomas. No abnormal 99mTc-labeled red blood cell accumulation was seen in the abdominal viscera, suggesting complete removal at surgery.


