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Discordant splenic uptake of Tc-99m colloid and Tc-99m denatured RBC in candidiasis-endocrinopathy syndrome

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We report discordant splenic uptake of Tc-99m colloid and Tc-99m heat-denatured red blood cells (RBC) which occurred in a 21-year-old female with candidiasis-endocrinopathy syndrome. Tc-99m colloid liver-spleen imaging showed no splenic uptake, suggesting the presence of functional asplenia. A subsequent Tc-99m heat-denatured RBC study clearly revealed a small spleen with preserved sequestrating function. These results may demonstrate that the qualitative dissociation of splenic functions in processing colloid and denatured RBC in functional asplenia: the sequestration function remains while the reticuloendothelial system is impaired.

Key words: Tc-99m colloid, Tc-99m denatured RBC imaging, functional asplenia, candidiasis-endocrinopathy syndrome

INTRODUCTION

CANDIDIASIS-ENDOCRINOPATHY SYNDROME, also termed autoimmune polyendocrinopathy-candidiasis syndrome was first described in 1956 in patients with the triad of hypoparathyroidism, adrenal insufficiency, and mucocutaneous candidiasis.[1] Since then various other manifestations have been described, including insulin-dependent diabetes mellitus, gonadal failure, hypothyroidism, gastrointestinal malabsorption, alopecia areata and totalis, pernicious anemia, vitiligo, chronic active hepatitis, keratopathy, and hypoplasia of dental enamel and nails.[2-4] Although there were no reports of a- or hypo-splenism in these reports, an acquired functional asplenia in a sibship has been described in a recent report,[5] but to our knowledge, discordant splenic uptake of Tc-99m colloid and Tc-99m denatured red blood cells (RBC) has not been reported in this syndrome. Presented here is a case of candidiasis-endocrinopathy syndrome, in which liver-spleen imaging with Tc-99m colloid showed no splenic uptake suggesting the presence of functional asplenia, and spleen imaging with Tc-99m heat-denatured RBC revealed uptake by a small spleen.

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CASE REPORT

A 21-year old female was admitted with mucocutaneous candidiasis and hepatic dysfunction. She was first seen at age 2 months with chronic mucocutaneous candidiasis. At age 12 years, she developed hypothyroidism and since then has been treated with thyroid hormone replacement therapy. Laboratory investigation revealed slight hepatic dysfunction: GPT, 41 IU/m*l* (normal range, 0–37 IU/m*l*); GOT, 49 IU/m*l* (normal range, 0–49 IU/m*l*); ALP, 305 IU/m*l* (normal range, 96–284 IU/m*l*). The serum IgG level of 1,966 mg/d*l* (normal range, 607–1621 mg/d*l*) was high. There were no Howell-Jolly bodies on the peripheral circulating blood smear. The diagnosis of candidiasis-endocrinopathy was made on the basis of the clinical picture. Radiograph CT (Fig. 1) showed a small spleen.

Liver-spleen imaging was performed to evaluate splenic function. An initial 500,000-count anterior image was obtained 30 minutes after intravenous injection of 148 MBq of Tc-99m tin colloid. Thereafter, posterior, right lateral, and left lateral images were recorded in the same acquisition time as the anterior image. And a subsequent SPECT study was done as follows. Sixty-four projection

data were acquired in a 64 × 64 matrix, in a 360° degree step rotation mode with an acquisition time of 10 seconds each. The image reconstruction was performed by a filtered back-projection method with a Ramp filter after pre-processing with a Butterworth filter to obtain 11.2 mm thick transaxial images. Planar and SPECT images (Fig. 2A, B) showed no splenic uptake, which would suggest the presence of functional asplenia despite deliberate overexposure of both planar and SPECT images.

Fig. 1 Radiograph CT shows a small spleen.

Subsequent spleen imaging with Tc-99m heat-denatured RBC was performed 7 days after liver-spleen imaging to evaluate the splenic sequestration function. Tc-99m heat-denatured RBC were prepared according to the modified *in vivo* method reported by Armas et al.[6] 0.5 mg of stannous ion as stannous pyrophosphate was injected intravenously. 15 minutes later, a 6 m*l* blood sample was drawn into a heparinized syringe. 111 MBq of Tc-99m pertechnetate was added to the blood and mixed gently. The mixture was heated for 30 minutes at 49.5 ± 0.5°C. Tc-99m heat-denatured RBC was injected, and the imaging was done 60 minutes later. An initial 500,000-count posterior image was obtained. Thereafter, anterior, right lateral, and left lateral images were recorded in the same

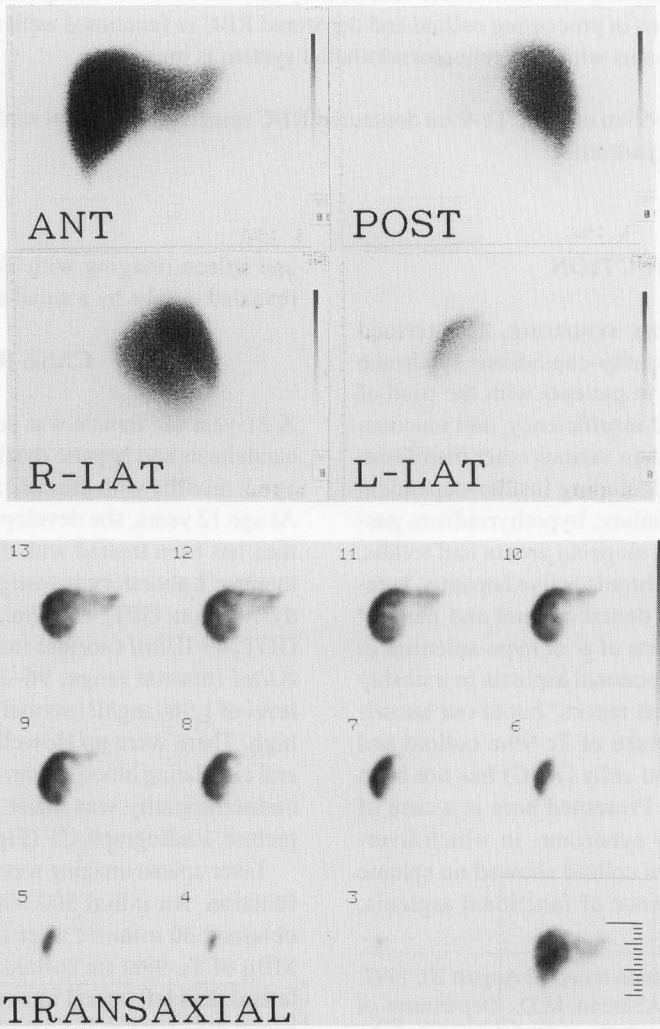

Fig. 2 Tc-99m tin colloid planar (A) and SPECT (B) images show no splenic activity.

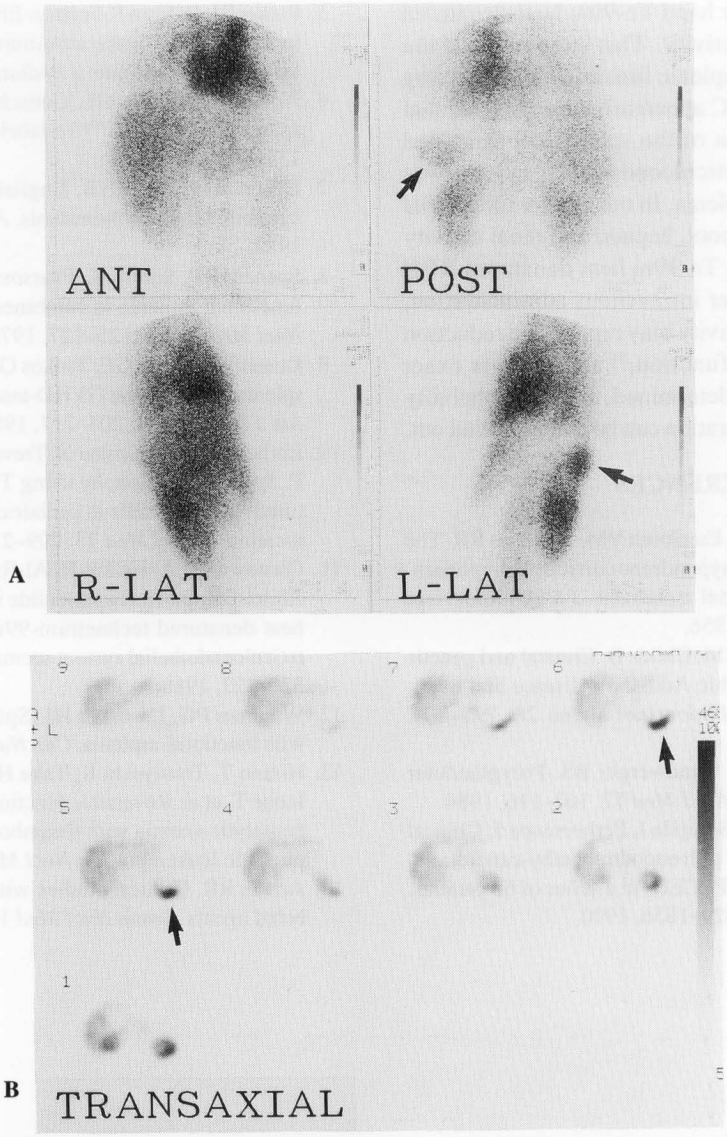

Fig. 3 Tc-99m heat-denatured RBC planar (A) and SPECT (B) images taken at 1 hour after intravenous administration show an obvious splenic activity (arrow). A substantial amount of cardiac blood pool, hepatic and renal activity is also noted.

acquisition time as the posterior image. And a subsequent SPECT study was performed in the same manner as that of live-spleen imaging. Planar and SPECT images (Fig. 3A, B) clearly revealed a small spleen with preserved sequestration function, although a substantial amount of cardiac blood pool and hepatic and renal activity was noted.

DISCUSSION

Functional asplenia, the failure to visualize the spleen with radioactive colloid even when the spleen is anatomically present, has been reported in a variety of disease states associated with abnormal immunoregulation, such as systemic lupus erythematosus, severe combined immunodeficiency, and graft-versus host disease.[7–9] This phenomenon has also been reported in a sibship with candidiasis-endocrinopathy syndrome.[5] The mechanism of functional asplenia which occurred in these conditions has not been proven, but an autoimmune disorder has been suggested.

Discordant splenic uptake of Tc-99m colloid and Tc-99m denatured RBC has been described in many conditions.[10–13] These conditions have demonstrated that Tc-99m denatured RBC imaging can still image the spleen even in those patients with functional asplenia on the Tc-99m colloid image. In the present case with candidiasis-endocrinopathy syndrome, a similar finding was observed: Tc-99m colloid liver-spleen imaging showed no splenic uptake, which would suggest functional

asplenia, and on the other hand Tc-99m heat-denatured RBC revealed splenic activity. This suggests that the qualitative separation of splenic functions in processing colloid and denatured RBC apparently occurred and that the sequestration function of the spleen still remained even when the splenic reticuloendothelial system was impaired in functional asplenia. In our case, a substantial amount of cardiac blood pool, hepatic and renal activity was also observed on the Tc-99m heat denatured RBC image taken at 1 hour after intravenous administration, This high extra-splenic activity may represent a reduction in splenic sequestration function,[14] although its exact mechanism remains to be determined, and the probability of insufficient RBC denaturation can be entirely ruled out.

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