Partial splenic embolization in beta - thalassemia major

A case report

Adalet Meral[1], Betül Sevinir[1], Yurtkuran Sadıkoğlu[2], Ergun Nacarküçük[1], Ünsal Günay[1]

[1]Departments of Pediatrics, and [2]Department of Radiology, Uludağ University Faculty of Medicine, Bursa, Turkey

SUMMARY: Meral A, Sevinir B, Sadıkoğlu Y, Nacarküçük E, Günay Ü. Partial splenic embolization inbeta-thalassemia major: a case report. Turk J Pediatr 2000; 42: 76-79.

Partial splenic arterial embolization was performed in a thalassemic child for hypersplenism manifested by splenomegaly, leukopenia, thrombocytopenia, and anemia requiring frequent erythrocyte transfusion. During a follow-up period of 11 months, his hematological parameters improved significantly and the spleen size decreased. Partial splenic embolization could be an alternative therapy to surgical splenectomy for thalassemic children with hypersplenism.

Key words: splenic embolization, thalassemia.

Thalassemia is considered to be the most common genetic disorder world-wide. It occurs in a high frequency in a broad region from the Mediterranean through Asia. Good transfusion programs may retard the development of splenomegaly. There is usually increasing hypersplenism and physical discomfort because of the grossly enlarged spleen. Progressive splenic sequestration of transfused cells is observed by 1-8 years of age. Splenic trapping of platelets, leukocytes and erythrocytes produces thrombocytopenia, leukopenia, anemia and an increased transfusion requirement which accelerates iron loading. The spleen is removed when the red cell transfusion requirement exceeds 200-250 ml packed red cell/kg/year[1].

Surgical anatomy may be distorted secondary to enlarged spleen and dissection can be difficult in massive splenomegaly. Additionally, thrombocytopenia increases the hemorhagic risk during surgery. Therefore, surgical removal of the spleen may carry a significant mortality and morbidity[2]. Laparoscopic splenectomy, which is often used in congenital spherocytosis or idiopathic thrombocytopenia, may also be difficult to perform due to increased collaterals and spleen size[3]. For this reason, partial splenic embolization (PSE) has been accepted as an alternative procedure to splenectomy in patients with splenomegaly is performed in children with beta-thalassemia major as will as with other diseases causing hypersplenism[4-6].

Case Report

A 17-year-old boy had been followed for beta-thalassemia major since he was six months old. A splenectomy could not be performed until 1993 when the patient was 13 years old because of the family's disagreement on the matter. In 1993, the surgical procedure was not successful due to massive splenomegaly with distorted collaterals. PSE was planned three years later when he developed severe pancytopenia and his transfusion requirement increased to six units of erythrocyte infusion per month. Before PSE, he developed local hyperemia and enduration in the periumbilical region. Subcutaneous abscess formation was determined by ultrasonography. The drainage of the abscess material revealed Escherichia coli and Klebsiella pneumonia, which were treated with a combination of penicillin, clindamycin and amikacin. He responded to treatment in 20 days and there was no further growth in the cultures.

He was vaccinated with 23 valent pneumococcal polysaccharide vaccine (Pasteur Merieux^{R}) and Hamophilus influenzae vaccine (Pasteur Merieux^{R}) before embolization. Penicillin (200,000 U/kg/day) and amikacin (15 mg/kg/day) intravenously were given 24 hours before the procedure and the following seven days. The procedure was performed under sterile conditions and intravenous sedation and local anesthesia were used.

A preliminary aortogram to view the celiac axis was obtained by approaching the femoral artery with a

5F catheter. A selective splenic arteriography was then performed (Fig. 1). The catheter was repositioned in the main splenic artery to determine its exact configuration and its branches. Controlled embolization of the peripheral splenic branches. Controlled embolization of the peripheral splenic branches and main splenic artery was performed using particles measuring 200 μ (IVALON^{R}). A postembolization arteriogram was obtained to show the occluded splenic artery (Fig. 2). Although the splenic artery seemed to be fully occluded by angiography, computerized tomography (CT) of the same region documented that the collaterals from the diaphragm and lumbar arterias flowed into the splenic parenchyma. The embolized splenic parenchyma was seen as hypodense regions by CT (Figs. 3, 4). It was determined that approximately 40 percent of the spleen was infarcted.

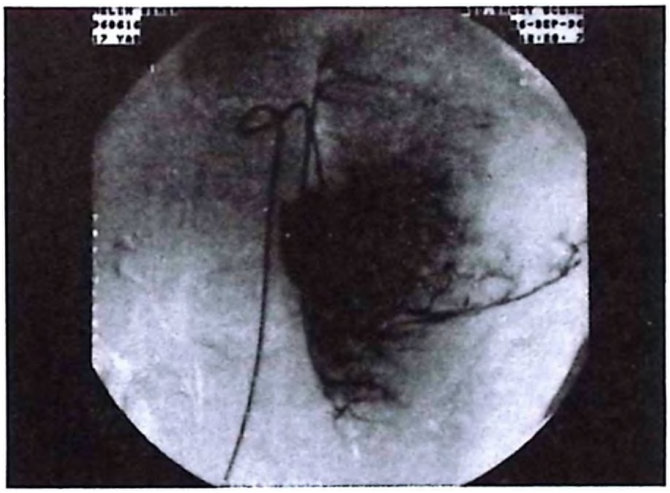

Fig. 1. Splenic arterial angiography before splenic embolization.

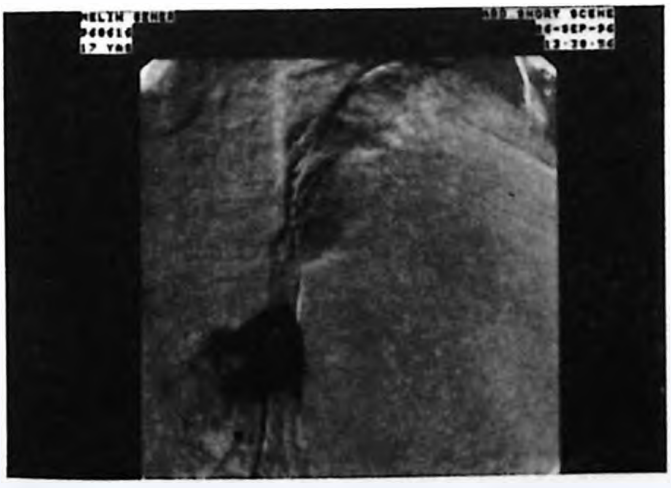

Fig. 2. Postsplenic embolization.

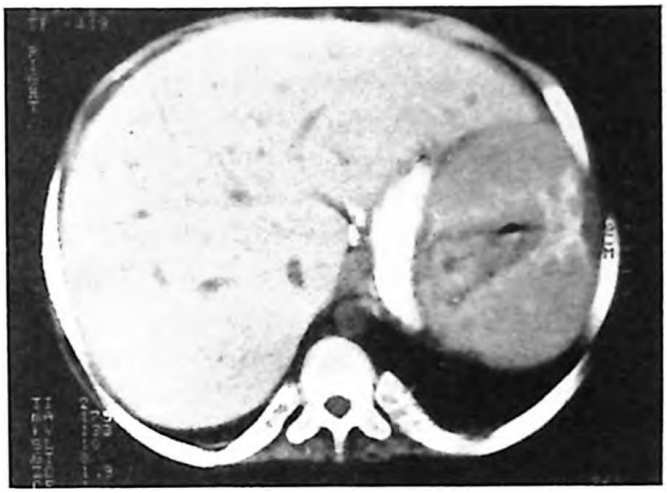

Fig. 3. Abdominal computed tomography showing the normal and necrotic splenic parenchyma.

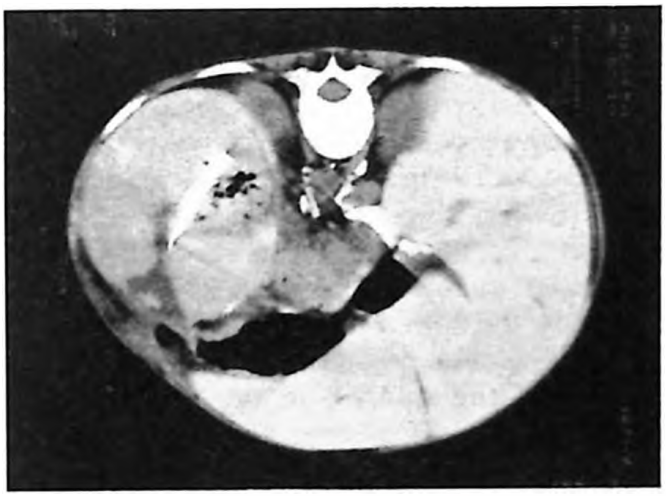

Fig. 4. Abdominal computed tomography showing the normal and necrotic splenic parenchyma.

Postoperative hemorrhage from the incision scar of the previous abscess occurred 48 hours after the embolization. It was attributed to the increased pressure in the collaterals following PSE. Somatostatin was thus administered to decrease the enhanced pressure of the splenic arterial bed. Active hemorrhage stopped after 72 hours of this treatment. However, serohemorrhagic and necrotic material continued to drain from the same region. It was determined by contrast radiography that this material was coming from the infarcted splenic parenchyma. A catheter was placed into the cavity. Irrigation with normal saline four times a day was performed through the catheter. Imipenem and tobramycin were started since Enterobacter boumani was grown in the cultures taken from the catheter.

Both leukocyte and thrombocyte counts increased during the first month of embolization. Necrotic drainage through the catheter ceased three months after PSE. Computed tomography controls demonstrated that the cavity size decreased from 9 cm to 4 cm. Antibiotherapy was discontinued since there was no further growth in the cultures.

Six months later, the spleen size decreased from 14 cm to 10 cm, determined by physical examination and ultrasonography, transfusion requirement decreased from 6 units to 2 units of erythrocyte infusions, and leukocyte (4,600-6,400/mm^3) and platelet counts (150,000-180,000/mm^3) both increased. The patient was lost due to severe myocardial dysfunction 11 months after PSE. During that period, his blood counts were stable as described above.

Discussion

Partial splenic embolization is accepted as a safe and effective procedure to surgical splenectomy for children with hypersplenism[6,7]. Hematological parameters improve for a long period of time and the spleen size decreases as a result of an infarction of greater than 40 percent of splenic mass[8]. However, the spleen size does not decrease to normal even after an infarction of more than 80 percent of the spleen[9]. In the present case, approximately 40 percent of the spleen was infarcted and thrombocyte and leukocyte counts increased two weeks after the procedure. Five months later, the increase of hematological parameters and decrease in transfusion requirement suggested that the PSE was successful.

The most important complications of PSE have been reported to be pleural effusion, pneumonia and spleen abscess[6]. In the present case, necrotic material drained from the infarcted region through the fistula connected to the previous periumbilical abscess cavity. The periumbilical abscess that developed before the procedure could be due to impairment of chemotaxis and netrophil migration, which may partially account for the increased susceptibility to infection in such children[10,11]. No serious infection following PSE was observed. The noninfarcted spleen might still continue to provide immunologic functions preventing the occurrence of overwhelming infections[12]. The hospitalization period was longer than expected due to prolonged resolution of the necrotic region. However, computed tomography in the third month after PSE showed that the resolution and organization of the infarcted region was completed.

Although repeat embolization is recommended in the literature for patients with a subnormal platelet count in whom the infarcted region has been less than 80 percent, our patient did not require a second procedure after eleven months of follow-up[6-9].

The patient died of severe myocardial infarction 11 months after PSE. This was due to previous increased transfusions which caused excess iron loading. However, during eleven months of follow-up, he had a better life quality since transfusion frequency decreased and he had no thrmbocytopenic complication. This outcome was achieved by 40 percent infarction of the splenic mass by arterial embolization.

Therefore, we believe that the morbidity and mortality associated with surgical splenectomy could be avoided using PSE in apropriate pediatric cases.

REFERENCES

1. Lukens JN. The thalassemias and related disorders: quantitative disorders of hemoglobin synthesis. In: Lee RG, Bithell TC, Foersterr J, Athens JW, Lukens JN (eds). Wintrobe's Clinical Hematology (9th ed) Vol. 1. London: Lea and Febiger; 1993: 1102-1145.
2. Schiller M. The spleen. In: O'Neill JA, rove MI, Grosfeld JL, Fonkalsrud WE, Coran AG (eds). Pediatric Surgery (5th ed) Vol. 2. St. Louis: Mosby; 1998: 1545-1554.
3. Tulman S, Holcomb GW 3d, Karamanoukian HL, et al. Pediatric laparoscopic splenectomy. J Pediatr Surg 1993; 28: 689-692.
4. Stanley P, Shen TC. Partial embolization of the spleen in patients with thalassemia. J Vasc Interv Radiol 1995; 6: 137-142.
5. Pinca A, Di Palme A, Soriani S, et al. Effectiveness of partial splenic embolization as treatment for hypersplenism in thalassemia major: a 7 year follow up. Eur J Haematol 1992; 49: 49-52.
6. Shah R, Mahour GH, Ford EG, Stanley P. Partial splenic embolization. An effective alternative to splenectomy for hypersplenism. Am Surg 1990; 56: 774-777.
7. Muguerza R, Lasseletta L, Vazquez J, et al. Partial splenic embolization in the treatment of hypersplenism. Long term results. Cir Pediatr 1995; 8: 11-16.
8. Brandt CT, Rothbarth LJ, Kumpe D, et al. Splenic embolization in children. long term efficacy. J Pediatr Surg 1989; 24: 642-645.

9. Watanabe Y, Todani T, Noda KT. Changes in splenic volume after partial splenic embolization in children. J Pediatr Surg 1996; 31: 241-244.
10. Matzner Y, Goldfarb A, Abrahamov A, et al. Impaired neutrophil chemotaxis in patients with thalassemia major. Br J Haematol 1993; 85: 153-158.
11. Kütükçüler N, Kutlu O, Nişli G, et al. Assessment of neutrophil chemotaxis and random migration in children with thalassemia major. Pediatr Hematol Oncol 1996; 13. 239-245.
12. Israel DM, Hassall E. Partial splenic embolization in children with hypersplenism. J Pediatr 1994; 124: 95-100.