

# Massive Splenomegaly Secondary to Prolidase Deficiency

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FIGURE 1

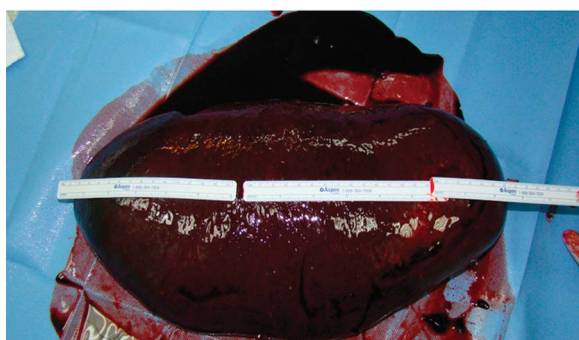
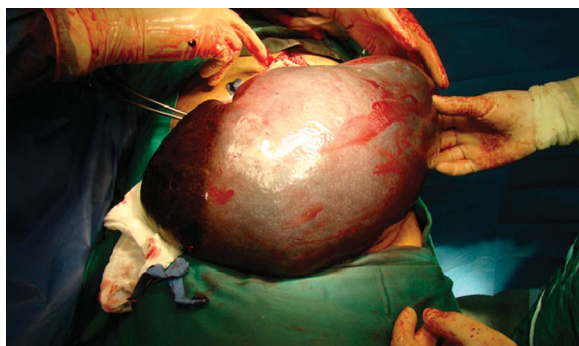


FIGURE 2

## CLINICAL PRESENTATION

A 64-year-old gentleman was referred by hematology-oncology service for elective splenectomy. The patient is known to have prolidase deficiency with recurrent skin ulcerations (Figure 1) and splenomegaly, causing early satiety, sensation of abdominal fullness and worsening pancytopenia. Prolidase deficiency is a rare autosomal-recessive inborn error of amino acid metabolism secondary to mutations of the human prolidase gene with few cases reported in the literature.<sup>1</sup> Clinical features include skin ulcerations, recurrent infections and hepatosplenomegaly.<sup>2,3</sup> Open splenectomy was performed, and massive splenomegaly was evident with the spleen measuring around 35 cm in its largest dimension (Figure 2). The patient had an uneventful postoperative course

and was discharged home after 4 days. On follow-up a month later, the patient reported marked improvement in his gastrointestinal symptoms and his pancytopenia had resolved.

## REFERENCES

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