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Case Study

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Reconstruction of the Portal Vein After Resection of A Large Hepatic Tumor Due to Tangier Disease : Case Study and Literature Review

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Summary
The reconstruction of the portal vein following the resection of a hepatic tumor associated with Tangier disease is a delicate and relatively rare surgical procedure. Tangier disease is a genetic disorder that affects lipid metabolism, resulting in cholesterol accumulation in tissues and vascular function anomalies. This study presents a clinical case where the resection of a large hepatic tumor and the reconstruction of the portal vein were successfully performed. We will discuss the importance of this rare intervention, the management of risks, and recommendations for long-term follow-up.

Keywords: Portal Vein, Dacron Graft, Tangier Disease, Hepatic Tumor

Introduction
Tangier disease is a rare genetic disorder characterized by a significant reduction in high-density lipoprotein (HDL) levels in plasma due to mutations in the ABCA1 gene, lipid accumulation in tissues, and hepatic, cardiac, and neurological anomalies. The diagnosis is often challenging due to its varied clinical presentation. The incidence of vascular complications in Tangier disease is underestimated, and the management of these complications remains a major challenge. The literature concerning portal vein surgery in this specific context is limited, and even more so following the resection of a large tumor [1,2].

Clinical Case
This is a 46-year-old patient, known to have Tangier disease since childhood, with a history of a myocardial infarction treated with stent placement, who has been diagnosed with a large liver tumor compressing the portal vein. The discovery was incidental during the exploration of digestive disorders associated with intermittent pain predominantly in the right hypochondrium persisting for one month. The physical examination noted isolated hepatomegaly. Imaging studies (CT angiography and ultrasound) revealed an 8 cm mass in the right lobe of the liver compressing the portal vein (Figure 1), and the extension assessment did not detect any abnormalities. Following discussion in a multidisciplinary consultation meeting, an enlarged right hepetectomy with resection of the mass followed by reconstruction of the portal vein using a Dacron graft was performed (Figure 2). The postoperative course was uncomplicated, and the patient is currently under follow-up at 12 months with no local recurrence.

Discussion
Described for the first time in 1961 on Tangier Island (USA), the disease is caused by a mutation in the ABCA1 gene, responsible for a defect in the transport of lipids to HDL. In exceptional cases, it may be associated with severe hepatic involvement, including massive tumors requiring resection with reconstruction of the portal vein [3-5].

Frequent comorbidities include peripheral neuropathy (60%), hepatosplenomegaly (70%), early cardiovascular diseases (30%), and chronic pancreatitis. The predominant clinical manifestation remains hepa¬tomegaly in our case. This condition has an estimated prevalence of less than 1 in 1,000,000, with fewer than 200 reported cases. It affects both sexes with a slight male predominance [6-8]. Manifestations appear as early as childhood, but severe hepatic complications arise in adulthood.

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The literature aligns with our patient, who is 46 years old and presents with cardiovascular comorbidities against a backdrop of Tangier disease [7,9].

The mechanisms leading to tumor formation in Tangier disease are still being explored. Chronic inflammation, oxidative stress, and lipid overload may play a role in this process. Tumor progression is exceptional, with fewer than five similar cases described [10,11].

Hepatic magnetic resonance imaging (MRI) with angiography allows for the assessment of vascular involvement. Portal involvement in these patients is reported in 20–30% of cases of hepatic tumors. These data highlight the necessity for proactive evaluation of the vascular status in these patients to optimize their clinical management [12].

Portal reconstruction in the face of a massive hepatic tumor and a systemic pathology such as Tangier disease represents a major surgical challenge [4,5,13]. In our case, a multidisciplinary approach facilitated tumor resection followed by portal reconstruction using a Dacron graft. The choice of graft (autologous iliac vein versus synthetic prosthesis) depends on the clinical background [14,15]. In our case, the synthetic choice aimed to minimize surgical risks in a cardiac patient [16,17]. Mid-term results demonstrate good tolerance provided the anastomosis is patent [18].

The management of this complex pathology is primarily preventive. Our patient did not exhibit any notable extrinsic factors. It is important to closely monitor the disease by conducting systematic familial genetic screening6, annual hepatic surveillance through Imaging starting from adolescence, regular lipid follow-up, multidisciplinary management4, avoidance of hepatotoxic substances, and vaccination against hepatitis. The literature notes that in the long term, there is an improvement in the quality of life of operated patients, stability of Tangier disease through regular monitoring of its markers, as well as patency of the portal vein with normal blood flow [19-23].

Figures

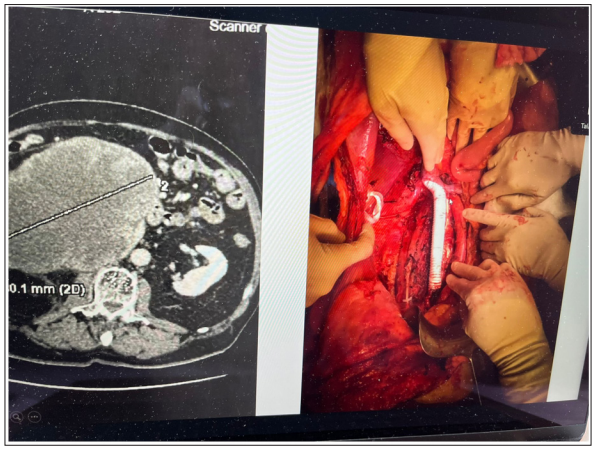

Figure 1: the abdominal angiogram shows a large mass affecting the liver, compressing the portal vein ; the abdominal aorta remains intact and patent (on the right)

Figure 2: image of the reconstruction of the portal vein by a Dacron prosthesis (on the left).

Conclusion

Tangier disease, which is relatively rare, can present with severe hepatic complications including portal compression. Reconstruction of the portal vein, although complex, is feasible with good results if risk factors are well identified and managed. Synthetic grafts may be indicated in specific cases. Future studies are needed to compare long-term outcomes based on the types of grafts used.

Author contribution

All authors have contributed to this work and have read and approved the final version of the manuscript.

Declaration of Competing Interest

The authors declare no conflict of interest.

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