A 62-year-old patient was studied after an episode of fainting with oppressive pain that appeared and recurred during exercise.

Abdominal-pelvic CT revealed a heterogeneous retroperitoneal mass from the infrarenal distal third of the inferior vena cava (IVC) with calcifications that was highly vascularized, partially extra- and endoluminal, with cranial extension up to the right auricle, that was implanted in the septum and up to the tricuspid valve to the RV (Figs. 1 and 2).

Given the obstruction of the IVC and the existence of collateral venous return, and considering the differential diagnosis between leiomyosarcoma and angiosarcoma with associated thrombosis, surgery was performed and the IVC was resected in 2 stages using laparotomy and sternotomy.

Final pathology diagnosis: high-grade leiomyosarcoma.

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* Corresponding author.
E-mail address: mar_eugg@hotmail.com (M.E. Gómez García).
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