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ABSTRACT N. PO87

INTRAVASCULAR LEIOMYOMATOSIS EXTENDING TO THE RIGHT ATRIUM: A CASE REPORT WITH A MULTIDISCIPLINARY MANAGEMENT.

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BACKGROUND

Intravascular leiomyomatosis (IVL) is a rare condition, with approximately 400 cases reported in the literature since its first description in 1896. It is characterized by the intravascular proliferation of smooth muscle cells, often with a branching pattern that can extend from pelvic veins to thoracoabdominal vessels. Although benign in histological terms, IVL can clinically mimic a malignancy and lead to potentially fatal complications. The majority of cases of IVL have been reported in women, especially in the fourth decade of life, frequently associated with uterine leiomyomas. The treatment of choice is surgical resection, aiming to reduce morbidity and mortality associated with vascular involvement and minimize the risk of recurrence.

CASE REPORT

We report the case of a 48-year-old woman who presented in December 2022 with dyspnea and palpitations. A transthoracic echocardiogram revealed a right atrial mass, prompting further evaluation. A total-body CT scan identified a large pelvic mass, likely of uterine origin, with intravascular extension involving the ovarian veins, the right internal and common iliac veins, and the inferior vena cava (IVC), reaching the right atrium. Pelvic mass biopsy confirmed a benign leiomyoma and intravascular leiomyomatosis. Following multidisciplinary discussion, the patient underwent an initial surgery to remove the tumor from the vascular structures. In June 2023, resection was performed from the atriocaval junction up to the confluence of the suprahepatic veins. Histological examination confirmed IVL. A radiological follow-up in May 2023 revealed an increase in pelvic mass size and recurrent intravascular extension to the right atrium. After re-evaluation, a three-stage surgical procedure was performed, including complete hysteroannessiectomy and tumor removal from the right iliac vein, IVC, and right atrium. The surgery was completed without complications. Postoperative histology confirmed IVL in all specimens. A follow-up CT in July, 2023 showed no residual disease, although a focal filling defect in the IVC-just above the iliac vein confluence and at the renal vein junction-suggested chemical thrombosis. The patient was started on anticoagulant therapy, which was well tolerated, without thromboembolic recurrence or major bleeding. Complete recanalization of the thrombosis was achieved.

CONCLUSIONS

This case illustrates the complex and rare nature of IVL, highlighting the critical role of a multidisciplinary management for diagnosis, treatment and follow-up. Postoperative clinical and radiological follow-up is essential to detect complications such as thrombosis and especially recurrence, which occurs in 16-30% of cases, particularly when complete surgical excision is not achieved. Additional studies are needed to define standardized surgical and therapeutic protocols for the management of IVL.

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