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Case Report

Intravenous leiomyomatosis

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ABSTRACT

Intravenous leiomyomatosis is a rare benign smooth muscle tumour arising from a venous wall or a uterine leiomyoma. This case highlights that intra-cardiac leiomyomatosis should be considered as a differential diagnosis in addition to extended cancer or thrombus, in female patients, diagnosed with a right sided cardiac mass extending from the IVC. To prevent pulmonary embolism or sudden death in patients with intra-cardiac extension, complete surgical resection is necessary. One stage surgical removal has been recommended for complete resection.

Keywords: IVL, Intra-cardiac extension, IVC

INTRODUCTION

Intravenous leiomyomatosis is a rare neoplasm characterized by a smooth muscle cell tumor with a histologically benign appearance that grows within the uterine and pelvic venous system and inferior vena cava (IVC), even involving the heart.[1] Although histologically benign, IVL can behave clinically in a malignant way, since sudden death can occur as a result of intracardiac leiomyomatosis with subsequent obstruction of venous return to the heart.[3]

There are currently two main theories regarding the cause of this neoplasm. The first suggests that the intima of myometrial sinuses is invaded by leiomyomatosis cells originating from the uterine myometrium. The second contends that the tumor is comprised of proliferating smooth muscle cells arising directly from the venous wall of the uterine or pelvic veins.[4] An intravenous leiomyomatosis typically occurs in parous women and prior to menopause. Surgical outcome is usually good as these patients are relatively young and tolerate the surgery fairly well.[2]

CASE REPORT

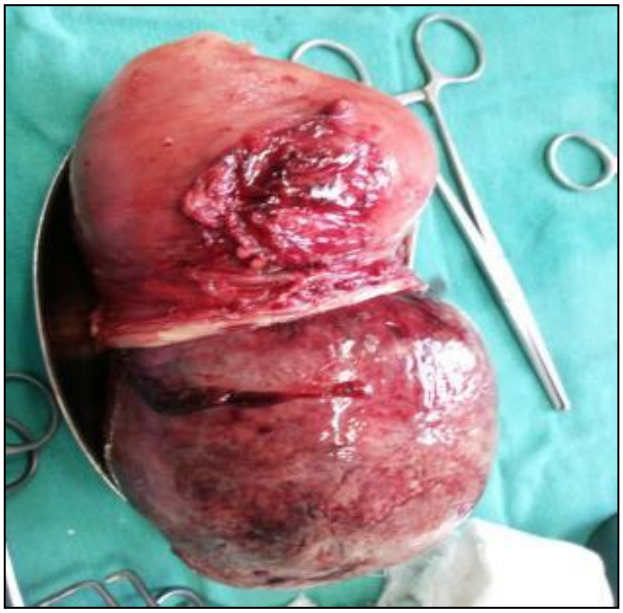

Figure 1: Hysterectomy specimen- uniformly enlarged uterus with huge myomatous fibroid.

40 years P_{2+0} presented in the department of obstetrics and gynaecology. with H/O irregular bleeding pervaginum for about 1 month and severe anaemia (Hb-5 gm%). On examination, uterus was found to be enlarged uniformally to about 20 weeks size because of a big fibroid which was projecting into vagina as a huge myomatous fibroid. She was given 2 units blood transfusions followed by total abdominal hysterectomy as shown in Figure 1.

Her postoperative period was uneventful and she was discharged on 5th POD in good condition and was perfectly normal after 6 weeks of follow up. After about 11 months, she reported with H/O pain epigastrium, shortness of breath and swelling of feet for about 5-6 months. As no gynaecological abnormality was detected, she was referred to medicine department. Investigations (USG and CT) revealed a huge tumour extending from internal iliac vein up to right atrium and provisional diagnosis of intravenous leiomyomatosis was made.

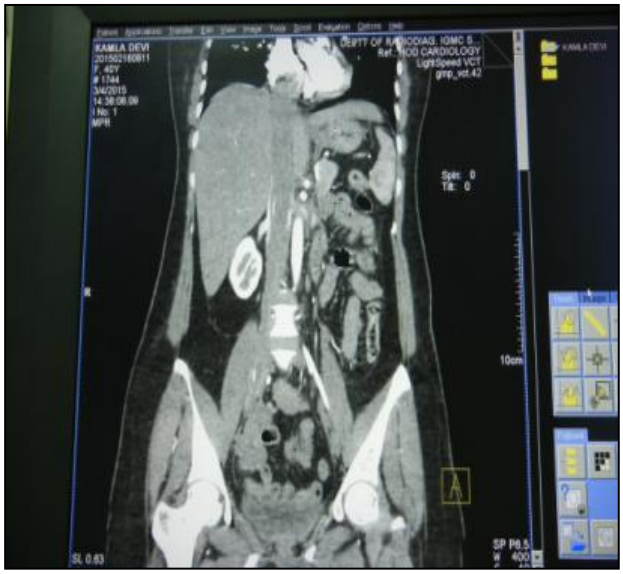

Figure 2: CT scan of a mass extending from the pelvis to IVC and Rt. Atrium.

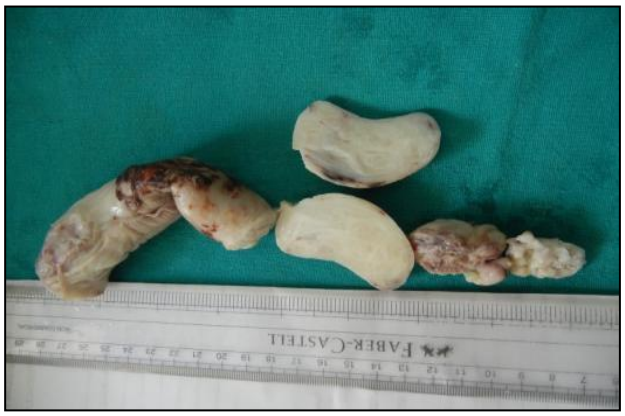

Figure 3: Gross operative specimen of IVL.

Exploratory laparotomy with mid sternotomy with single stage removal of venous leiomyomatosis (tumour) from right iliac vein, IVC and right atrium along with right salpingoopherectomy was done. Tumour was firm in consistency, involving right internal iliac, common iliac vein and IVC. Multiple nodules were present in right iliac fossa (about 1 × 1 cm size), rest of the pelvis and upper abdominal organs were normal. Postoperative period was uneventful and diagnosis was confirmed on HPE.

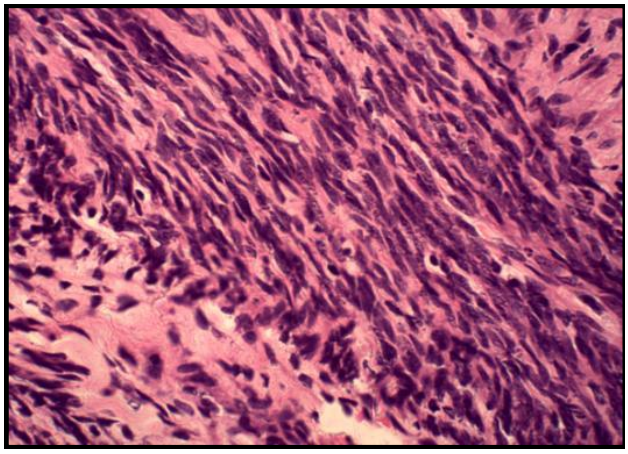

Figure 4: HPE specimen of IVL of smooth muscles.

DISCUSSION

Intravenous leiomyomatosis is defined as one of the unusual growth patterns of histologically benign uterine leiomyoma. Clinically, the tumour is considered malignant because of the style of progression and the occurrence of pulmonary metastasis.[2] IVL may also embolise to the right cardiac chambers or pulmonary artery and cause obstruction .It may be diagnosed as a primary cardiac tumour or a venous thrombus.[5]

Intra-venous leiomyomatosis, benign metastasizing leiomyoma and malignant leiomyosarcoma are three rare associated variants of uterine leiomyoma. Recognition of the potential of fibroids for venous invasion was accredited to Birch Hirschfeld who described four cases in 1896.[6] However, it was in 1959 when microscopic features were described by Marshall and Morris. Lam et al have recently reviewed the literature and identified 200 reported cases of leiomyomatosis, 68 of which had intra-cardiac extension.[5] They identified signs of cardiac failure, venous obstruction or abdominal distension (in women with previous history of hysterectomy done for fibroid uterus) as being the most common mode of presentation in those with involvement of the right atrium as in our case. In the early stages of venous extension, the diagnosis is often unrecognized and further extension of intravenous leiomyomatosis is not prevented by resection of the primary tumour.[6]

This condition is mostly diagnosed in the patients who are present in the 5th and 6th decades of life. Female

gonadal steroids play a major role in the pathogenesis as the condition is associated with high estrogen levels. Pathogenesis is unclear, although it is postulated to be arising from uterine leiomyoma invading into the uterine vein and extending along the lumen of the major veins. The incidence of IVL is thought to be underestimated because, in the early stages, the tumor extension remains in the vessels of the myometrium and cannot be detected on imaging. The correct diagnosis relies on a high index of suspicion and should be suspected whenever an intravascular tumor is noted during surgery or microscopic analysis.[7] Multi-detector computed tomography and ultrasound plays a vital role in arriving at the diagnosis. Immunohistochemistry aids in the diagnosis and differentiating it from other tumours. Most of the tumours are +ve for smooth muscle specific Actin, Desmin and Caldesmon. The intravascular presence of the tumour is demonstrated by vascular markers like CD34.R-1

Leiomyomatosis is thought to be a progression of simple fibroma from its origin in the uterine vein in the direction of its venous drainage involving either the uterine or ovarian vein. Involvement of both iliac and ovarian veins is extremely rare. Recurrence rate of 30% has been reported in patients with intravenous extension.

Treatment is primarily surgical but as the tumour is hormone sensitive , antioestrogen therapy has been used both preoperatively and as an adjunct to surgery and to prevent progression of residual disease.[6] Surgery can be performed as both single and staged procedures and cardiopulmonary bypass is advocated for resection of tumours with intra-cardiac extension.[5]

Although there may be some difficulties in distinguishing such lesions from low-grade sarcomas, they are different histologically from sarcomatosis uteri because the intravenous plug is mainly smooth muscle in origin. The tumor behaves like a benign tumor, and the mitotic index is low with most active lesions showing only one mitosis per 15 high-power fields. Conservative surgery is usually adequate as the intravenous extensions are probably incapable of independent parasitic existence and remain dormant after removing the uterus. The prognosis is excellent even when tumor is left in pelvic veins.[8]

CONCLUSION

Intravenous leiomyomatous may be diagnosed with a high index of suspicion, thus avoiding potentially fatal delay. IVL should be considered in females with an H/O uterine leiomyoma or with a H/O hystrectomy for leiomyomas, presenting with signs of venous obstruction. IVL are histologically benign tumours with quasimalignant behaviour with potential to spread to distant organs along the blood vessels giving rise to life threatening complications. Careful follow up of these patients throughout life is essential.

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