ORIGINAL RESEARCH PAPER

Pathology

A RARE CASE OF RETROPERITONEAL LEIOMYOMA – A HISTOPATHOLOGICAL CASE REPORT

KEY WORDS:
retroperitoneal, leiomyoma, histopathology, intra-operatively.

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ABSTRACT

Uterine Leiomyoma is one of the most common benign tumor in women in which hysterectomy is done. Retroperitoneal leiomyomas are rare entity of benign smooth muscle tumors which may be presented as single or multiple small peritoneal nodules. Because of its rarity and non-specific presentation, the pre-operative diagnosis might be challenging. These neoplasms shows same morphological and microscopic features similar to uterine leiomyoma. Here we present a case of retroperitoneal leiomyoma in a 40 years old woman suspected to be a malignant tumour of unknown nature. After post-surgical resection the retroperitoneal mass was sent to the Department of Pathology, RIMS Ranchi for histopathological diagnosis. After histopathological examination the case was confirmed as leiomyoma but the final diagnosis of retroperitoneal fibroids can be obtained intra-operatively and after histopathological examination. We report the case from a histopathological perspective with a brief review of relevant literature. Retroperitoneal fibroids are rare neoplasm and surgical removal of the tumour is the main treatment.

INTRODUCTION

Leiomyoma is benign tumour with smooth muscle differentiation accounting for the most common gynaeco logical neoplasm. Retroperitoneal growth pattern in leiomyoma is a rare condition as only ~100 cases have been reported in the English literature to date [1]. Most of them are detected in women of middle aged group. These histological benign tumors which originate from smooth muscle cell, usually arises in the genitourinary tract but may arises in nearly any anatomic site[2]. On histology these neoplasms share the same microscopic morphological and phenotypic features with uterine leiomyoma[3]. Retroperitoneal leiomyoma often present a diagnostic challenge as it shares resemblance with most retroperitoneal smooth muscle tumour in terms of clinical presentation, histology, and steroid/oestrogen and progesterone hormone positivity; moreover, they are considered to be malignant whereas retroperitoneal leiomyoma is not[4]. Diagnostic work up should include USG, CT and/or MRI but final diagnosis of retroperitoneal fibroid is obtained intra-operatively and after a histopathological examination.

CASE REPORT

A case of 40 year old female, belongs to low socio-economic background of an interior village of Jharkhand, presented at out patient department of Surgery, RIMS Ranchi, with complain of acute pain abdomen and abdominal swelling. Patient had history of 2 normal vaginal delivery, the age of younger child is 15 years. Patient had history of asymptomatic abdominal lump since past 6 years. There was no menstrual irregularity. There was no previous illness and surgical intervention. Then she experienced gradual increase in size of the lump with symptom of increase frequency of micturition, pelvic pain and discomfort within 6 months and for these she consulted local medical practitioner for symptomatic relief but she did not get relief and referred to higher centre. On general examination she was an average built women with normal vitals. Patient was non-vegetarian, non-alcoholic, non-smoker with normal bowel habits. On per abdomen palpation there was an extremely large, firm, tender palpable abdominal mass of 25cm x20cm size , with smooth surface and restricted motility occupying the right iliac fossa and lower part of the abdominal cavity to the level of umbilicus. All parameters including tumour markers were normal.

Systemic examination including gynaecological examination did not reveal any abnormality. USG revealed a large encapsulated 30x20x10cm solid tumour filing most of the abdominal cavity. Since her definitive diagnosis was not possible and mass was large and rapidly growing, the patient was taken up for surgical exploratory laparotomy with excision of retroperitoneal mass.

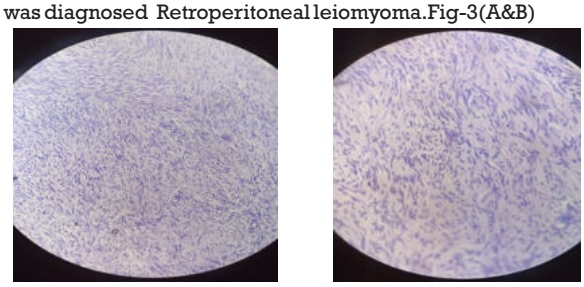
Intra-operative a large encapsulated mass of 30cm x20cm x10cm adherent to the small and large intestine with adjacent structure also involved ureter and inferior venacava. Uterus and bilateral adnexa were normal. After the procedure the specimen was sent for histopathological examination in the department of pathology, RIMS Ranchi. We received tissue specimen of retroperitoneal mass in 2 pieces (mass+capsule) [.Fig-1]

Fig. 1- Macroscopic appearance of the retroperitoneal myoma

The gross appearance of the mass is greyish white in colour, firm in consistency, and measuring 18cm x 14cm x11cm. The capsule measuring 11cm x6 cm. On cut open section of the mass is greyish white in colour, whorled appearance with no apparent degeneration or calcification [Fig-2].

Fig. 2 – Cut section of retroperitoneal myoma

Haematoxylin Eosin stain section revealed spindled mature smooth muscle bundle running in different direction with some blood vessel seen with abundant eosinophilic cytoplasm arranging in intersected fascicles and the case was diagnosed Retroperitoneal leiomyoma.Fig-3(A&B)



A-20 X view B. 40 X view

Fig-3(A&B) Haematoxylin and eosin showing the 20X and 40X view of mature smooth muscle bundle with abundant eosinophilic cytoplasm arranging in intersected fascicles.

DISCUSSION

Uterine fibroid are most common benign solid pelvic tumours in women and are present in about 80% of all hysterectomy specimen [5]. Billings et al.[6] have described two distinct subsets of deep soft tissue leiomyoma: (i) leiomyomas of somatic soft tissue which develop in extremities and (ii) Retroperitoneal -abdominal leiomyoma. The later occurs almost exclusively in women during the premenopausal period[6] with(i) concurrent uterine leiomyoma or (ii) history of hysterectomy for uterine fibroid being, reported in up to 40% of cases[7]. Clinically, presenting symptoms of retroperitoneal leiomyomas are often non-specific (including discomfort, fatigue, back pain) or are related to compression of adjacent structures[1,7]. As these tumour may extend to the upper retroperitoneal, being as a high as the level of the renal hilum[7]. Pathogenesis of these lesions remains unclear. In fact, retroperitoneal leiomyomas might arise from the harmonically sensitive smooth muscle elements of the embryonal remnants of Mullerian and Wolffian duct might the cell of origin. They might also represent metastatic or synchronous primary lesion because only nine cases of this condition have been reported in men [1]. Since retroperitoneal smooth muscle tumour are more often malignant than benign prompt and accurate pre-operative radiological assessment is of paramount importance. Ultrasonography provides good localisation for retroperitoneal masses, though CT and specially magnetic resonance imaging (MRI) are most useful screening tools in evaluating and distinguishing exact nature of the tumour and its relations with its adjacent organ and vascular structure. However not diagnostic modality appears highly sensitive or specific in ruling out malignancy and differential diagnosis on the basis of radiological finding alone is difficult[8].

Kho and Nezhat [9] studied 12 cases of parasitic leiomyoma distinct from uterus which included two retroperitoneal fibroids. They suggested iatrogenic parasitic myoma formation as the cause of retroperitoneal fibroid.With regard to their pathological origin, it is unclear whether these retroperitoneal lesions represent metastatic or synchronous primary lesions and whether these arises from the hormonally sensitive smooth muscle elements[6].

Histologically, the distinction of benign leiomyoma and malignant leiomyosarcoma (especially low grade) may also difficult. The histopathological parameters used for differential diagnosis include gross tumour size, the presence of nuclear atypia, pleomorphism and necrosis and the mitotic activity as the most useful guide to prognosis. On light microscopy, leiomyoma consists of monomorphic spindle cells arranged in interweaving fascicles which are separated by variable amounts of hyalinized collagen. Smooth muscle cells are elongated with eosinophilic cytoplasm and uniform, cigar shaped nuclei. Usually, there is no cytologic atypia or necrosis and mitotic index is less than 5 per 10 high-power fields. It is very important to distinguish coagulative from hyalinizing necrosis as the presence of coagulative necrosis, even in absence of significant atypia would lead to a diagnosis of sarcoma[10].

Molecular genetics of these tumour have not been enough studied. To the best of our knowledge, the first cytogenetical analysis of retroperitoneal leiomyoma has been conducted by Panagopoulos et al.[3], it showed a fusion of the genes KAT6B(10q22) with KANSL1(17q21) as a result of translocation t(10;17).

Surgical excision of the mass is the gold standard treatment in respect of benign or malignant nature of the mass and can be done by laparotomy or laparoscopic[11]. Konda et al.[12] reported a case of retroperitoneal fibroid where resection was done by laparoscopic approach.

Abdominal hysterectomy along with the resections depends on the age of the patients and her symptomatology and associated uterine myoma. The overall prognosis of patient with retroperitoneal leiomyoma is good with small potential with local recurrence[4].

CONCLUSION

Retroperitoneal leiomyomais rare neoplasm because of its unusual location their occurrence in retro peritoneum is however extremely rare. Poorly accessible location and rarity often make retroperitoneal tumour a clinical puzzle. Thorough imaging may also miss the exact site of this fibroid. Final diagnostic is made intra operatively on direct visualisation as well histopathological examination of the specimen. Complete excision with or without abdominal hysterectomy is the treatment of retroperitoneal leiomyoma.

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