Acute Abdomen as an Unusual Presentation of Broad Ligament Angiomyolipoma. A Case Report and Review of Literature

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Case Report

A 55 year old postmenopausal woman presented with complaints of sudden onset of right sided abdominal pain restricting her daily activities. Physical examination showed a large, smooth and tender immovable mass, extending from right lumbar region to the right anterior superior iliac spine and extending beyond the midline of the abdomen. On per speculum examination the cervix could not be visualised and to the extent vagina was seen, appeared normal. On per vaginal and rectal examination, cervix was pulled up. Uterus felt atrophic and retroverted, both the fornices were free. On the left in a plane above the adnexa, a firm mass could be tipped on downward displacement of the abdominal mass. On per rectal examination, rectal mucosa was free, bilateral parametrium were supple, pouch of Douglas was free and the same mass could be tipped with the middle finger. Complete haemogram and biochemical investigations were within normal limits. CA-125 was 8 IU/L. A CT scan of abdomen and pelvis was done with contrast. It showed a large retroperitoneal mixed attenuation mass $18.7 \times 12.7 \times 15.5$ cms with density value of +20–30 Hounsfield units (Fig. 1).

During surgery, minimal haemorrhagic ascites was seen in the peritoneal cavity. A well encapsulated retroperitoneal mass between the leaves of the broad ligament was

completely resected, clamping its attachments to the parametrial and lateral pelvic wall and other soft tissues (Fig. 2). Frozen section was reported as lipomatous tumour. Total hysterectomy with bilateral salpingo-oophorectomy was then performed.

Pathology

Grossly the tumour was irregular, whitish yellowish, bosselated with surface vessels and measuring $30 \times 23.5 \times 25$ cms. Cut section showed lobules of fatty tissue, white fibrous and myxoid areas (Fig. 3). Uterus and bilateral tubes and ovaries were normal grossly and histologically.

Histology of the tumor consisted of interlacing bundles of spindle cells and foci of thick and thin walled blood vessels in a background of mature adipocytes suggestive of angiomyolipoma (Fig. 4). Interestingly immunohistochemistry with antihuman melanoma HMB-45 was negative.

Post operative period was uneventful. Patient is on follow up for the last 13 months and does not have a recurrence clinically or sonographically.

Discussion

The incidence of renal and extra renal angiomyolipoma is 0.3–3 % [1] and that of angiomyolipoma occurring between the leaves of the broad ligament is very rare, only one case by Rajan and et al has been published [2]. Probably ours is the second case reported till date. Acute pain could be due to ischemia of the tumour mass or an impending rupture of the vessel. On reviewing the literature extra renal angiomyolipoma occurring in the female genital tract has been reported

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Fig. 1 CT scan showing a large retroperitoneal mixed attenuation mass (AML) $18.7 \times 12.7 \times 15.5$ cms with density value of +20–30 Hounsfield units. Uterus (U) is pushed to the left side

Fig. 2 Intra operative findings of well encapsulated and vascular angiomyolipoma (AML). The round ligament (RL) is stretched and divided. Normal looking ovary (O)

Fig. 3 Cut section showed lobules of fatty tissue (F), areas of white fibrous and myxoid areas (M)

Fig. 4 Microphotograph showing spindle shaped fascicles (F) surrounding blood vessels (V) in the background of adipose tissue (A)

in the uterus by Yaegashi H et al [3]. Ovary by Anderson AE et al [4], Vagina by Chen KT et al, and Peh SC et al [5, 6], and fallopian tubes Katz DA et al [7]. Therefore total of six cases arising from the female genital tract has been reported in the English medical literature and the present case will be the seventh case and the first to be reported from this country. Tuberosclerosis is associated with renal angiomyolipoma but has not been reported in patients with extra renal angiomyolipoma and in the present case also. [1].

The differential diagnosis of retroperitoneal fat containing tumours includes lipomatosis, lipoma, lipoblastoma, hibernoma, leiomyoma with fatty degeneration, liposarcoma and angiomyolipoma. Despite numerous preoperative imaging modalities a final diagnosis is made only after surgical excision and hence the need for on table frozen section facilities.

During surgery the tumour was found completely encased in a capsule, attached to the soft tissues between the two leaves of the broad ligament on the right side and the lateral pelvic wall Figs. 5 and 6. Gross and microscopic evaluation ruled out any invasion of the tumour to the uterine wall, fallopian tube and ovary, and had no multicystic spaces as earlier described by Rajan et al in 2003 [2].

Fig. 5 Microphotograph showing predominantly adipose tissue (A), spindle shaped fascicles (F)

Fig. 6 Microphotograph showing predominantly spindle shaped fascicles (F) and Sparse adipose tissue (A)

It is important to establish a histological diagnosis by frozen section, since a malignant neoplasm like liposarcoma would need a more radical surgery. The present case is unique in that, the occurrence in the broad ligament previously is undocumented and differs from the previously reported case in the literature by the absence of entrapment of paramesonephric remnants and antihuman melanoma-HMB 45 –negativity. It is of particular interest that non-vascular smooth muscle cells were negative for HMB45, in contrast to renal and other extra renal angiomyolipoma in which HMB45 immunoreactivity has been demonstrated in these cells. Similar experience has been reported by Yaegashi H et al [3]. On follow up, ultrasound of abdomen and pelvis did not show any recurrence for the last 13 months. The definitive treatments are surgical excision and close follow up. We report this case because of its rarity and its acute presentation and hence consider it in differential diagnosis of acute mass abdomen. Preoperatively anticipate the

possibility of malignant retroperitoneal tumours and stress the need for availability of frozen section to decide the extent of surgery.

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