Leiomyoma in 16 Years Old Libyan Female in Two Sites

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Abstract

Benign retropertionealleiomyoma orextrauterine leiomyoma is a rare condition that occurs in all age groups and it is particularly prevalent among women of late childbearing age. All patients have a history of uterine leiomyoma and/or myomectomy, often associated with distant metastases from the uterus, which commonly occurs in the lung. We report a case of retroperitoneal leiomyoma with lung liomyoma at the same time, and did not have previous gynecological problem or surgical operation in a 16 year old Libyan female.

Keywords: Benign Leiomyoma, Retropertoneal Leiomyoma**,** Lapratomy.

Abbreviation:

Benign Metastasizing Leiomyoma (BML), Lung Biopsy (LB), Immunohistochemistry (IHC), Benign Leiomyoma (BL), Open Lung Biopsy (OLB), National Cancer Institute Sabratha (NCIS), Ultra Sound Scan (USS).

Introduction

Retroperitoneal leiomyoma is in frequent, and their prevalence among primary retroperitoneal tumors has been estimated as 0.5 -1,2 %.

Leiomyoma is a common benign conditions arising from smooth muscle cells. Approximately 20–30% of women older than 35 years have uterine leiomyoma that is manifested clinically [1]. Retroperitoneal leiomyoma has a rare occurrence and has recently been recognized as distinctive lesions with similar histological features as uterine leiomyoma. Poliquin et al. [2] studied features of about 100 cases of retroperitoneal leiomyoma. This rare entity is usually misdiagnosed preoperatively even with diagnostic imaging. We report a case of large retroperitoneal leiomyoma and Right lung leiomyoma at the same time .

The lungs are the most common site of metastasis, with characteristic scattered nodules [3-4]. The pathological examination of a pulmonary nodule

usually indicate a specific smooth muscle phenotype with a low proliferation index and slow growth. This type of disease is usually called pulmonary benign metastasizing leiomyoma (PBML). Most cases have been discovered by chest X-ray or CT scan during routine examinations. Some patients have symptoms such as cough, dyspnea or chest pain. PBML is quite difficult to diagnose by simple medical imaging or physical examination and is often misdiagnosed as pneumonia, bronchitis, phthisis or metastasizing lung cancer [5]. Open lung biopsy (OLB) is one of methods of diagnostic procedure for PBML.

Extra uterine leiomyoma is uncommon, generally benign and occasionally cause diagnostic dilemmas as they can mimic malignancy. Retroperitoneal leiomyoma is a rare finding and there are very few reported cases of leiomyoma without previous uterine surgeries or concomitant presence of uterine fibroids [6].

Case report

16 years old female referred from a private clinic by history of right chest pain (stabbing in nature and not radiating) which started one year back. Chest X-ray was done (Fig 1) and showed big mass in lower left side of the chest and upper abdomen.

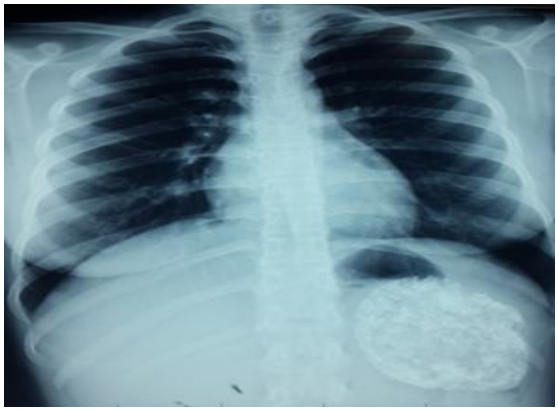

Fig 1: Chest X Ray.

History & Physical Exam

Single Libyan female menarche at age of 14, regular cycle, no history of chronic illness before or surgical interventions, and examination was unremarkable . Abdominal ultrasound & CT scan revealed large soft tissues mass 8x7 cm at left adrenal gland (Fig 2), and multiple nodules at the base of right lung (Fig 3). Radiology report showed Left suprarenal gland nuroblastoma with suggestive of lung mets.

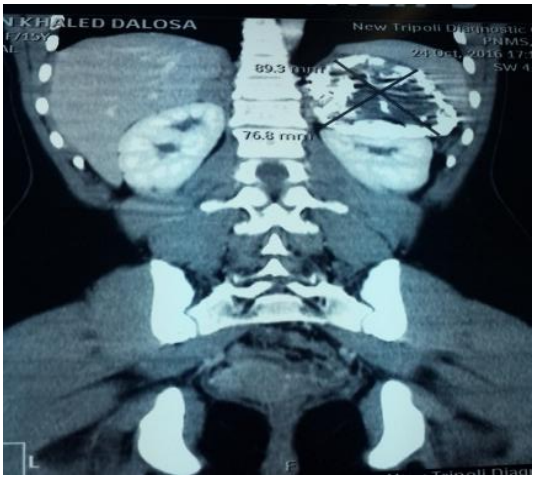

Fig 2: Suprarenal Mass

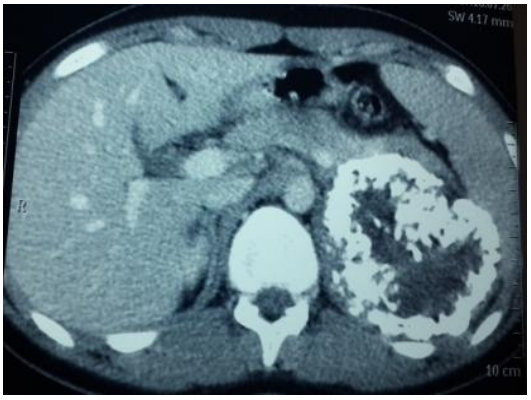

Fig 3: Multiple nodules at the base of right lung.

She underwent laparoscopic resection intraoperative finding big mass measured 9.5x9x7.2 cm, retroperitoneal adherent to left adrenal gland (LT adrenactomy) and excision of the mass enblock was done on 20 Nov 2016.

Histopathology Results

Microscopy: showed well circumscribed encapsulated benign mesenchymal tumor composed of interlacing bundles of smooth muscle cell separated. (Fig 4)

IHC: SAM :Postive

Desmin: Focaly Positive

Myogenin : Negative

S100 :Negative

Cd117 :Negative

Ki67 : Positive

It is benign leiomyoma as it was well circumscribed; no mitosis, no pleomorphisn (Fig 5), with metaplastic calcification; no necrosis bland cytomorbhology.

IHC rulled out leimyosarcoma, rhabdomyosarcomaand neuroblastoma. Epithelial neoplasm is ruled out by negative cytokeration (Fig 6 – Fig 7). On 21 Nov 2016 she underwent right thoracotomy (OLB). Histopathology showed benign leiomyoma, no evidence of malignancy (Fig 8). IHC showed spindle cells which were negative for HMB 45.

Figure 4: Smooth muscles, actin immunostain.

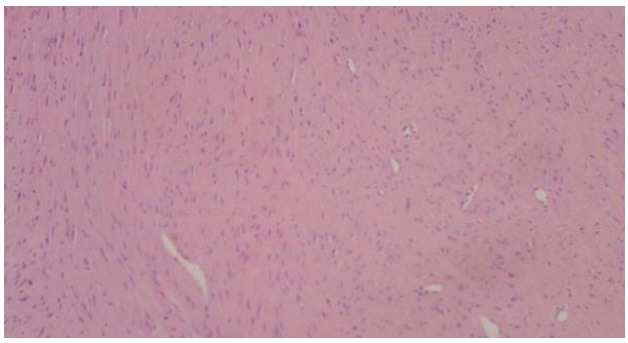

Figure 5: Benign spindle cell, No pleomrphism or mitosi.

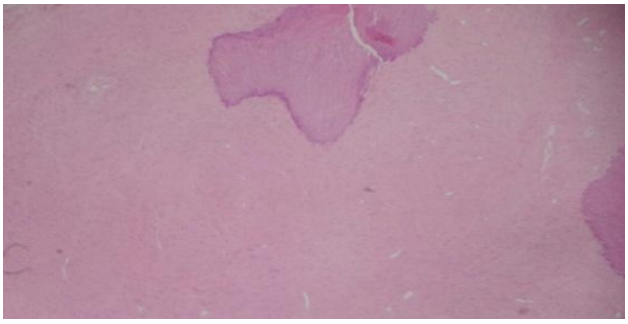

Fig 6: Leiomyoma with metaplastic calcification.

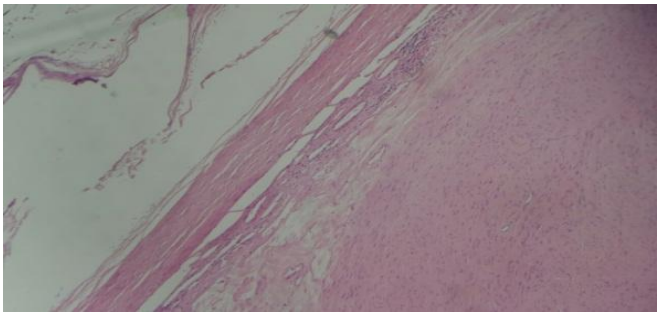

Fig 7: Entirely encapsulated benign leiomyoma f.

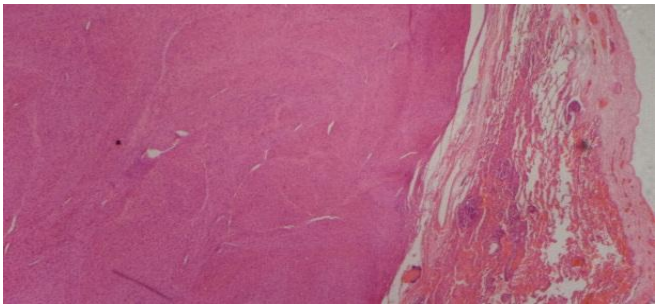

Figure 8: Lung Biopsy (Same tumor that seen in lung).

Discussion

Most retroperitoneal smooth muscle tumors are believed to be malignant, and Leiomyomas are considered very rare.

Leiomyoma is defined by the World Health Organization (WHO) as a ‘‘circumscribed benign, often tumor composed of intersecting bundles of mature smooth muscle cells.”

Based on the histopathologic findings, the WHO classified Leiomyoma into three groups: Solid Leiomyoma, Vascular Leiomyoma (angioleiomyoma) and Epithelioid Leiomyoma (Leiomyoblastoma). Our case is the solid variants of Leiomyoma [7].

The extra uterine Leiomyoma presentations mentioned in the literature are benign metastasizing Leiomyoma, disseminated peritoneal Leiomyomatosis, intravenous Leiomyomatosis, parasitic Leiomyomata (Leiomyoma adherent to surrounding structures acquire an auxiliary blood supply and become detached from the uterus such Leiomyomas have been termed Parasitic), with retroperitoneal growth [5]. Furthermore the unusual sites of origin include the vulva, ovaries, urinary bladder, and urethra. Some rare locations are sinonasal cavities, orbits, kidneys, and skin [8].

Retroperitoneal fibroids are rarely diagnosed preoperatively even with imaging techniques like USS, CT, and MRI. And in most case reports of retroperitoneal Leiomyoma, etiology and pathogenesis of Leiomyoma is still poorly understood. Since these tumors probably arise from smooth muscle cells including those blood vessels, Leiomyomas are common tumors that may originate from anywhere that smooth muscle

exists. The most common site for a Leiomyoma is in the uterus. Other Leiomyoma sites have been presented as case reports.

Horstmann et al. reported that the radiological presentation of pulmonary benign metastasizing Leiomyoma is as multiple nodules in 87% of cases (70% bilateral nodules and 17% unilateral nodules) or as a solitary nodule in 13% of cases [9]. However, it was the first case at NCI-Sabratha reported Retroperitoneal Leiomyoma.

There are several main hypotheses for the pathological origin of PBML [10-11] to be confirmed: (I) it is derived from low-grade Leiomyomatosis; (II) it is derived from benign metastasizing hysteromyoma; or (III) it is derived from a multicenter-growing Leiomyomatosis. Additionally, some researchers hypothesize that BML is related to angioleiomyoma [12]. The main pathological PBML lesions show no significant differences from Leiomyomatosis in other parts of the body. The characteristic findings are well-defined nodules varying from miliary size to 10 cm, interwoven with a pale solid ductile cut surface with no hemorrhage or necrosis.

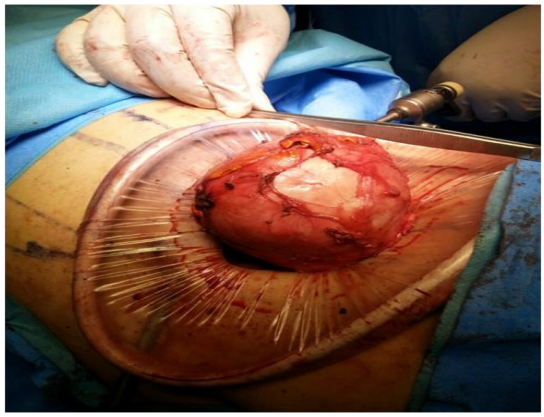

Figure 9: Intra-operative Finding.

Immunohistochemical staining of tumor issues in the lung showed that the tumor cells are characteristic of smooth muscle cell differentiation (α-SMA+, Desmin+), and the low expression of Ki-67 indicates a low tumor cell proliferation index, which indicates that the likelihood of malignancy was not high.

Making the diagnosis of benign leiomyoma is very difficult, and it has strict pathological diagnostic criteria. It is based on the results of the immunohistochemical staining and without history of Leiomyoma. In our case the pathological examination showed spindle-shaped cells without mitotic activity or nuclear atypia. The tumor cells were strongly positive for SMA, confirming the smooth muscle origin of this benign tumor. In addition to these features, immunohistochemical staining was negative for CD117, ruling out an extra-gastrointestinal stromal tumor, and all of the cells were negative for S100, myogenin, positive for desmin , ki67 2% very low.

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