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Retroperitoneal Liposarcoma-Excision of Retroperitoneal Tumor and Right Nephrectomy: A Case Study

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Abstract

A 64 year old woman was referred to hospital with complaints of abdomen right side tightness with discomfort since one month. Computed tomography scan of abdomen investigation revealed large retroperitoneal mass measuring 18.9*12.1cms displacing the inferior vena cava right kidney and renal vessels in right hypochondriac and lumbar regions of the abdomen with large fat components. She underwent surgery which involves the resection of tumor mass with non-affected surgical margins. These tumors tend to be resistant to radiotherapy or chemotherapy. Among the most important prognostic factor related to survival is surgery with non-affected margins. A review on etiology, pathophysiology, pathological classification and grading is explained in literature review.

Keywords: Liposarcoma; Retroperitoneal; Sarcoma; Well differentiated; Radiotherapy; Chemotherapy

Introduction

Retroperitoneal liposarcoma refers to liposarcoma arising from abdomen and pelvic retroperitoneal adipose tissue [1]. According to Windham and Pisters (2005) liposarcoma accounts for less than 1% of systemic malignant tumors retroperitoneal liposarcoma is the most common type (41%) of retroperitoneal soft tissue sarcoma, followed by leimyosarcoma and malignant fibrous histiocytoma. Onset of age is 55-75 years, slightly more common in men than in women with a ratio of 1.3:1 for men to women consisting of 68.3% retroperitoneal tumor and 11.6% liposarcoma [2]. Among 119 cases of retroperitoneal liposarcoma the ratio of male to female incidence was 1.9:1 and the median age at onset was 58 years old [3].

Etiology

The pathogenesis of retroperitoneal liposarcoma remains unclear, which is possibly associated with:

(a) Immunosuppressant

(b) Genetic factors

(c) Environmental factors

(d) Other factors

Immunosuppressant

Immune deficiency and immunosuppressive drugs are associated with pathogenesis of retroperitoneal soft tissue sarcoma [4]. It is reported that patients with systemic lupus erythematosus developed diffuse infiltrative retroperitoneal mucinous liposarcoma [5] after treated with steroid hormone for 13 years.

Genetic factors

Individuals with family history of lipoma or liposarcoma are more susceptible to developing retroperitoneal liposarcoma [6]. Retroperitoneal liposarcoma has been reported to occur successively in two compatriots with family history of malignant fibrous histiocytoma.

Environmental factors

Environmental carcinogens, Phenoxy acid herbicides, chlorophenols, and contaminant 2-, 3-, 7-, 8-TCDD may be related to retroperitoneal sarcomas [7].

Other factors

The changes in levels of insulin receptor and postreceptor in adipose tissue and decrease in biological activity of insulin may be involved in retroperitoneal liposarcoma [8].

Pathogenesis

The pathogenesis of retroperitoneal liposarcoma remains unclear [9] and may be related to molecular mechanism [10].

Mechanism of MDM2-p53

MDM2 gene (human homologue of the murine double minute type2) located at 12q13-15 region shows constant amplification

in well differentiated liposarcoma[11].MDM2 is a p53-specific E3 ubiquitin ligase and principle cellular antagonist of P53,acting to limit the p53 growth suppressive function in unstressed cells[12]. In healthy body, the precise balance between p53 and MDM2 guarentees the normal proliferation and differentiation of tissue cells. If MDM2 is overamplified, p53 activity is inhibited, resulting in uncontrollable cell proliferation. This may be related to the pathogenesis of retroperitoneal liposarcoma [13].

Mechanism for prune-nm23-H1

Prune the human homologue of Drosophilia prune gene, located in 1q21-23, encodes a protein that can bind to nm23-H1[14] (nucleoside diphosphate kinase) to downregulate its activity. The nm23-H1 may inhibit cell proliferation and tumor metastasis. The balance and precise coordination between prune and nm23-H1 expression present in healthy human bodies in contrast, overexpression of prune gene is found in liposarcoma, with downregulation of nm23-H1 activity [15]. This may be one of the molecular mechanisms responsible for the pathogenesis of liposarcoma [16].

Staging of Retroperitoneal Liposarcoma

Currently used staging system for soft tissue sarcomas is the TNM system developed by the American joint committee on cancer [17] (AJCC) (2010 7th edition). In this system clinical staging is based on histology, size, depth, lymph node, distant metastasis [18].

T Staging (primary tumor T)

TX primary tumor cannot be assessed

T0 No evidence of primary tumor

T1 Tumor < 5cm in maximum diameter

T1a superficial tumor

T1b Deep tumor

T2 5cm tumor > 5cm in maximum diameter

T2a superficial tumor

T2bDeep tumor

N Staging (Regional lymph nodes N)

Regional lymph nodes that cannot be assessed

No regional lymph node metastasis

Regional lymph node metastasis

M staging (distal metastasis)

M0 No distal metastasis

M1 Distal metastasis

G Histological grade (G)

GX Grade that cannot be assessed

G1 Grade 1 well differentiated

G2 Grade 2 moderately differentiated

G3 Grade 3 poorly differentiated

Pathological Classification and Grading

Since 2000, liposarcoma has been classified into four categories by WHO according to immuno histochemistry (IHC) and molecular and cytogenetic characteristics based on conventional histopathological findings [19].

- Nonclassic liposarcoma/ high grade differentiated liposarcoma
- Well differentiated liposarcoma/ dedifferentiated liposarcoma
- Myxoid/ round cell liposarcoma
- Pleomorphic liposarcoma

Pathological grading of LS is currently determined by the grading system of soft tissue malignant tumor [20]. which has been recently modified by the French Federation of cancer centers sarcoma group with a new scoring classification method [21]. Histological grading is calculated as the total score for three parameters, including tumor differentiation, degree of necrosis, and mitotic count in the new classification system [22]

Differntiation

(a) Score sarcomas closely resembling normal adult mesenchymal tissue (eg: well differentiated LS)

(b) Score 2 sarcomas with confirmed histological typing (eg: Myxoid liposarcoma)

(c) Score 3 embryonal and undifferentiated sarcomas and sarcomas of uncertain types

Mitotic figures

a. 0-9/10 HPF score 1

b. 10-19/10 HPF score 2

c. ≥20/10 HPF score 3

Tumor necrosis (Under microscopy)

a. Score 0: no necrosis

b.Score 1: ≤50% tumor necrosis

c.Score 2: >50% tumor necrosis

Grading system

Grade 1 total score of 2-3

Grade 2 total score of 4-5

Grade 3 total score of 6-8

The accuracy of histologic grading directly predicts the prognosis of patients with retroperitoneal liposarcoma [23]

Histologic grade

Grade 1: total score 2-3

Grade 2: total score 4-5

Grade 3: total score 6-8

Case Report

A 64 years female patient admitted in hospital with chief complaints of abdomen right side tightness with discomfort since one month and decreased appetite and weight loss. She had a past history of diabetes mellitus, Hypertension on regular medication

Table: 1: Medication history

Inj mixtard	subcutaneous
T.Diamicron-XR	60mg
T.Metformin	500mg
T.Telsartan-H	40mg/12.5mg
T.Atocor	5mg OD
T.Shelcal	OD

Table: 2: Physical examination.

Temperature	98.6℉
Blood pressure	140/90
Pulse rate	80
Respiration rate	20
Spo2	98%

Table 3: Hematology.

Test	Result	Units	Values
Hb	11.5	g/dl	12-15
MCHC	31.9	g/dl	32.0-37.0

Table 4: Biochemistry.

Test	Result	Units	Values
Bilirubin-Total	1.26	mg/dl	0.00-1.20
Bilirubin-Direct	0.41	mg/dl	0.00-0.30
Bilirubin-Indirect	0.85	mg/dl	0.10-0.80

CT Scan of Chest

Impression

Known case of retroperitoneal tumor

Small patchy fibrotic lesions are seen in right lower lobe

No soft tissue density nodules seen in both lungs

CT Coronary Angiogram

Impression

Total calcium score of 294 multiple calcified and mixed plaques in proximal mild left anterior descending. Moderate stenosis is seen in proximal left anterior descending (52% diameter stenosis) Calcified plaques in proximal left circumflex artery and in obtuse marginal causing mild stenosis (29% diameter stenosis) discrete calcified plaques in proximal and mild right coronary artery no significant stenosis

Scan of Abdomen (Plain and Contrast

Impression

Large retroperitoneal mass measuring 18.9*12.1 cm Right kidney and right vessels in right hypochondria and lumbar regions of the abdomen with large fat components multiple small calcified fibroids in the uterus

Discussion

The first description of a retroperitoneal lipomatous tumor excision was made in 1761 by Giovanni Battista Morgagni during the autopsy of a 60 year old woman. These tumors are uncommon malignant representing between 0.07 and 0.2% of all neoplasm. They have an incidence of approximately 2.5 inhabitants per 100,000 with an average age of presentation between 40 and 60 years, with a distribution in both sexes equally. Most of them are diagnosed incidentally when performing an imaging test for another reason since most are asymptomatic. They can produce during their growth usually when they exceed 20 cm non specific abdominal pain, early satiety, neurological or obstructive symptoms (urinary or digestive) by compression. For its diagnosis the test of choice is computed tomography with intravenous contrast, since it allows in most cases an adequate staging and preoperative evaluation. The retroperitoneal tumors appear as a large homogenous encapsulated mass of fatty tissue with fine septa displacing the renal parenchyma or even the intestinal bundle. Abdominal ultrasound may conform the presence of a hyperechoic mass and may be useful at the beginning of study. The (WHO) World Health Organization has classified the liposarcomas into two groups according to the degree of differentiation in low grade and high degree. In our case patient came with complaints of abdomen right side tightness with discomfort, loss of appetite, weight loss these are the symptoms or etiological factor for this patient which result from toxins produced by necrotic tissue, metabolites, and cachexia. She had a past history of Hypertension and Diabetes on regular medication since 10 years adequate urine output and bowel and bladder habits are regular. The mainstay therapy for retroperitoneal liposarcoma is complete surgical excision. Patient underwent surgical procedure of complete excision of existing tumor and right nephrectomy. Retroperitoneal tumors grow in an occult manner most patients are asymptomatic in early stage. Often manifested as painless mass and has grown into a very large size before

being detected. Symptoms and signs may not be obvious until the tumor has grown large and compressed adjacent organs or tissues. Retroperitoneal tumors that grow in loose connective tissue space of retroperitoneum usually don't cause obvious symptoms when they are small. When they grow to large size the tumor may result in symptoms by compressing and invading blood vessels, nerves, other vital organs.

Surgical Procedure

35*30 cm tumor noted in right side of retroperitoneal adherent to right kidney Retroperitoneal tumor was neutralized Area adhere to kidney was sharply discreted Tumor was well differentiated liposarcoma by frozen section Right nephrectomy was done as the tumor was adherent to right kidney Hemostasis checked drain was placed

Preoperative

T.PANTOP HS

INJ.TRIKA 0.25mg

SODIUM PHOSPHATE ENEMA

INJ.TAXIM 1gm

INJ.AMIKACIN 500mg

Post operative

IV fluids dextrose normal saline / ringers lactate 120m/hr

Vitals checked

IJ.MAGNEX FORTE 1.5gm in 100ml

INJ.METROGYL 500mg

INJ.TRAMADOL 50 mg in 150 ml

INJ.PANTOP 40 mg

Epidural analgesia when ever needed

Buvalor patch

DUOLIN nebulizer

INJ.ZOFER 4mg

Watch drain for bleeding

Table:5: Drug chart.

Drugs	Dose
Inj.Magnex forte	1.5gm TID
Inj.Metrogyl	500mg TID
T.Tramadol	50mg TID in 100ml NS
Inj.Pantop	40mg BD
Bovular patch	10mg to be applied
Neb.Duolin	Respulses BD
Inj.Zofer	4mg TID

Pharmacist Intervention

Untreated condition

*Multiple small calcified fibroids in uterus.

Conclusion

In this case study patient came with complaints of abdomen right side tightness with discomfort since one month necessary investigations were done and diagnosed as retroperitoneal liposarcoma underwent surgical procedure of excision of tumor and right nephrectomy was done. During the hospital stay patient is treated accordingly with symptomatic and supportive measures. We apprehend that Surgery is the mainstay of treatment of non-metastatic retroperitoneal liposarcoma. The fat content accounts for >75% of well differentiated liposarcoma tissue while the non-fat content is generally manifested as nodule or mass. In conclusion liposarcomas are rare tumor that due to its retroperitoneal location does not present specific symptoms, being diagnosed when they present a large size and produce compressive symptoms. The use of the chemotherapy or radiotherapy controversial due to the low sensitive of these types of tumors.

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