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RESEARCH ARTICLE

MYXOID LIPOSARCOMA WITH RARE ENTITY BREAST, THIGH AND PELVIS: CASE REPORT.

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Abstract

Liposarcoma is a malignant tumor of mesenchymal origin, it is the second most frequent histological type of soft tissue sarcomas after fibrous malignant Histiocytoma. A 45-year- old female patient presented with huge intra-abdominal mass, extending into the upper pelvis and right thigh and involving all the posterior thigh muscle. CT scan of both regions "thigh and abdomen" showed a mass extending from diaphragm to the upper pelvis. Needle core biopsies from both masses, abdominal and thigh, showed neoplastic proliferation of small cells. Although Myxoid liposarcoma is a relatively rare malignancy, its diagnosis should be suspected in the presence of lesion of soft tissues and a histological and imaging investigation should be done without delay to confirm the diagnosis.

Introduction:-

Liposarcoma is a malignant tumor of mesenchymalorigin, it is the second most frequent histological type of soft tissue sarcomas after fibrous malignant Histiocytoma. This cancer accounts for up to 18% of all soft tissue sarcoma [1]. Four histological varieties of increasing malignancy are described according to the WHO classification [2]: the liposarcoma, myxoidliposarcoma, pleomorphic liposarcoma, and dedifferentiated liposarcoma. This classifcation has above all a prognostic interest. Myxoidliposarcoma (MLS) is the result of chromosomale rearrangement, t(12;16)(q13;p11) [3] and consists of uniform round to oval shaped primitive non-lipogenicmesenchymal cells and a variable number of small signet-ring lipoblasts in a prominent myxoidstroma with a characteristic branching vascular pattern [4].

MLS represents more than one third of liposarcoma, it is the second most common subtype of liposarcoma and of all adult soft tissue sarcomas [5].

Case report:-

A 45-year- old female patient presented with huge intra-abdominal mass, extending into the upper pelvis and right thigh and involving all the posterior thigh muscle.

On 2013, CT scan of both regions "thigh and abdomen" showed a mass extending from diaphragm to the upper pelvis, more lateralized to the left side and showed heterogeneous density. The abdominal mass measured about 358 × 229 × 202 mm in its CC, AP, SS diameters respectively.

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There were tiny nodular calcification foci. Bone window demonstrated no suspicious lesions

The well-defined mass in the posterior aspect of the right thigh showed heterogeneous density, mainly fat and soft tissue densities with multiple calcification foci. The mass extended all the length of the thigh down to the level of the right knee joint and it measured about 370 × 240 × 250 mm in its CC, SS and AP diameters respectively. The right femur bone showed no cortical erosion and no intramedullary lesion and also no periosteal reaction.

Needle core biopsies from both masses, abdominal and thigh, showed neoplastic proliferation of small cells that had round to oval nuclei and single or multiple cytoplasmic clear vacuoles which indented the nucleus. A chicken-wire capillary network was noted in the background. Mitotic figures were rare, and no necrosis was identified. Focal hypercellularity of lesion was noted with the hypercellular areas comprising approximately 30% of thigh tissue.

Properly controlled immunohistochemical stains showed the neoplastic cells to be positive for vimentin, rarely positive for S-100, and negative for CK (AE1/AE3), desmin, muscle specific actin, and CD 34. The chicken-wire vascular network was highlighted by CD 34. The diagnosis of myxoid / round cell right thigh liposarcoma was then confirmed. The patient received palliative radiotherapy.

An ultrasonography of both breasts revealed scattered fibro-glandular parenchyma. A well-defined hypoechoic lesion was seen in the axillary tail of the right breast approximately at 10 o'clock. This lesion measured about 3.5 × 1.7 cm. The right breast mass was likely a fibroadenoma. No cystic or solid lesions were detected in the left breast.

In 2015, the patient presented with right breast mass, mammography was done and, as compared to the previous mammography done on 2013, the current one showed the previous reported right breast lesion at 10 o'clock, but with a markedly enlarged diameters; 10 × 8.8 cm, with lobulated out lines. This mass occupied most of the right upper outer quadrant. A change in character was as well noticed; the lesion became more echoic with areas of degeneration and calcifications. A new lesion anterior to the first one appeared and it measured 3.3 × 1.1 cm. The left breast was unremarkable. Right breast core biopsy was done and showed proliferation of malignant small cells in a myxoid background. Tumor cells had ovoid nuclei and single or multiple cytoplasmic clear vacuoles that indent the nucleus. No necrosis was seen. Mitotic figures were rare. A chicken wire network of vessels was evident in the background. The lesion was diagnosed as myxoidliposarcoma.

Discussion:-

Despite that breast is the most common site of primary cancer for women, metastases from primary malignancies outside the breast are rare [6-9].

The cause of this rarity is due to the characteristics of breast tissue, as it contains large areas of fibrous tissue with a relatively poor blood supply [10]. This is particularly seen in older women whose breasts are found to be structurally rich in fibrous tissue with a poor blood supply compared with younger women [11]. This structural difference also explains why the majority of breast metastases occur in woman younger than 50 years of age as in our case [12]. Breast metastases are more frequently occurring in malignant melanomas, sarcomas, lung cancers, ovarian tumors, renal carcinomas, and thyroid tumors [13,14]. As regarding myxoidliposarcoma, about one-third of patients with this malignancy develop distant metastases [15]. Myxoidliposarcoma are known to spread to pulmonary sites [16].In patients with extra pulmonary metastasis, the disease-free interval and the overall survival rate have been shown to be significantly better compared with those with pulmonary metastasis [17,18]. Breast metastasis from myxoidliposarcoma is a rare entity, in fact few cases are reported in the literature [10,19].

The challenge for the physician is to distinguish a multifocal primary sarcoma from a metastasis of a unique tumor. Gharehdaghi et al, 2014 proposed three criteria to differentiate between those two entities; it includes the site of the second tumor, cytogenetic features and the time between the two lesions; 6 months is the average time between the diagnosis of a primary sarcoma and the development of metastasis. In our case the breast metastasis occurred 2 years after the diagnosis of the first tumor [20].The histological examination of a specimen biopsy helps to have an accurate diagnosis of this tumor [21].Furthermore, imaging of the abdomen and chest should be performed for accurate staging [22] and for post treatment follow-up in order to detect an eventual recurrence of the tumor or, even more, the occurrence of metastases [23,24].

For our patient imaging eliminated skeletal extension of the tumor at the time of the diagnosis of the primary tumor, but the patient didn't have a monitoring of her breast lesion and the diagnosis of metastasis was done by mammography because the patient complained from the enlargement of the breast lesion diagnosed earlier as fibroadenoma.

The prognosis of myxoidliposarcoma depends in a major part of the histological characteristics [25].

A study including 20 cases of myxoidliposarcoma, found that after a follow up of 8 years, tumors with more than 5 % of round cells had a greater risk of recurrence [26]. Other authors found the same results [27]. Based on published papers, no factor was found to be a predictor of the site of the myxoidliposarcoma metastasis. In the other hand, most studies agreed that age was independently associated with a lower survival of patient with myxoidliposarcoma [28].

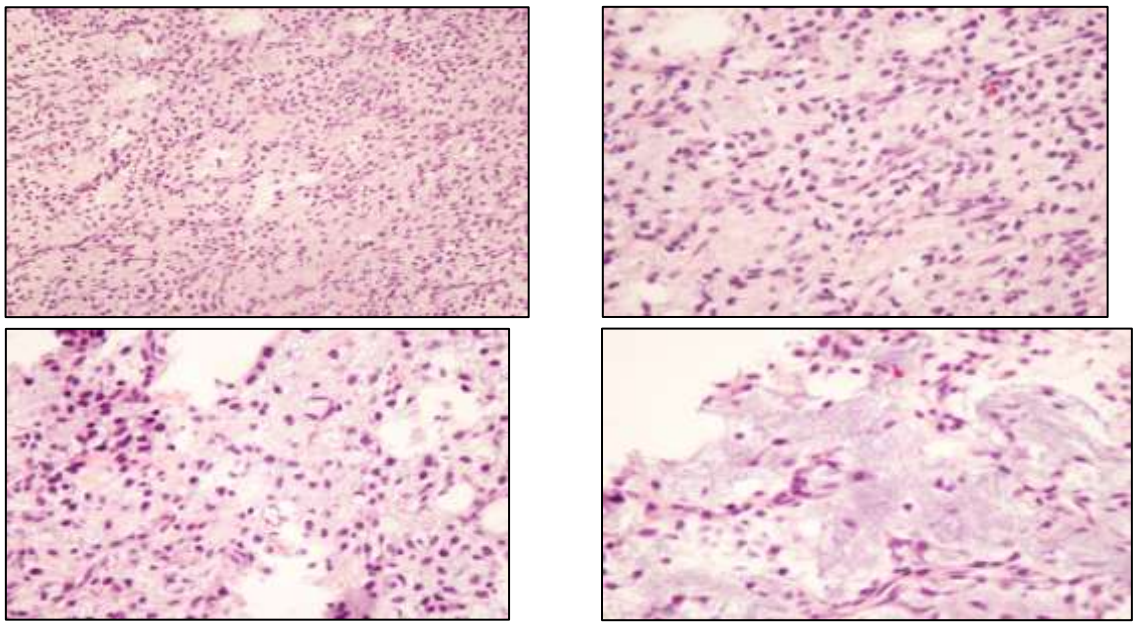
In addition to age, tumor size was found to be associated with a bad prognosis, in our patient the tumor exceed the threshold of 15 cm cited by some authors as a factor that increase the risk of death in patient diagnosed with liposarcoma [29].

Myxoidliposarcoma, like other soft tissue sarcomas, are treated mainly by surgery. The main purpose of the surgery is to remove the tumor in its entirety and to prevent its reappearance. The most reliable method is a radical resection [30].But in the case where the tumor was unresectable, like in our case, a palliative radiotherapy is proposed [31].In fact myxoidliposarcoma is known to be particularly sensitive to radiotherapy [32]. Choi et al showed that low fraction-sized intraperitoneal radiotherapy allowed a durable intraperitoneal control for metastatic peritoneal myxoidliposarcoma [33].

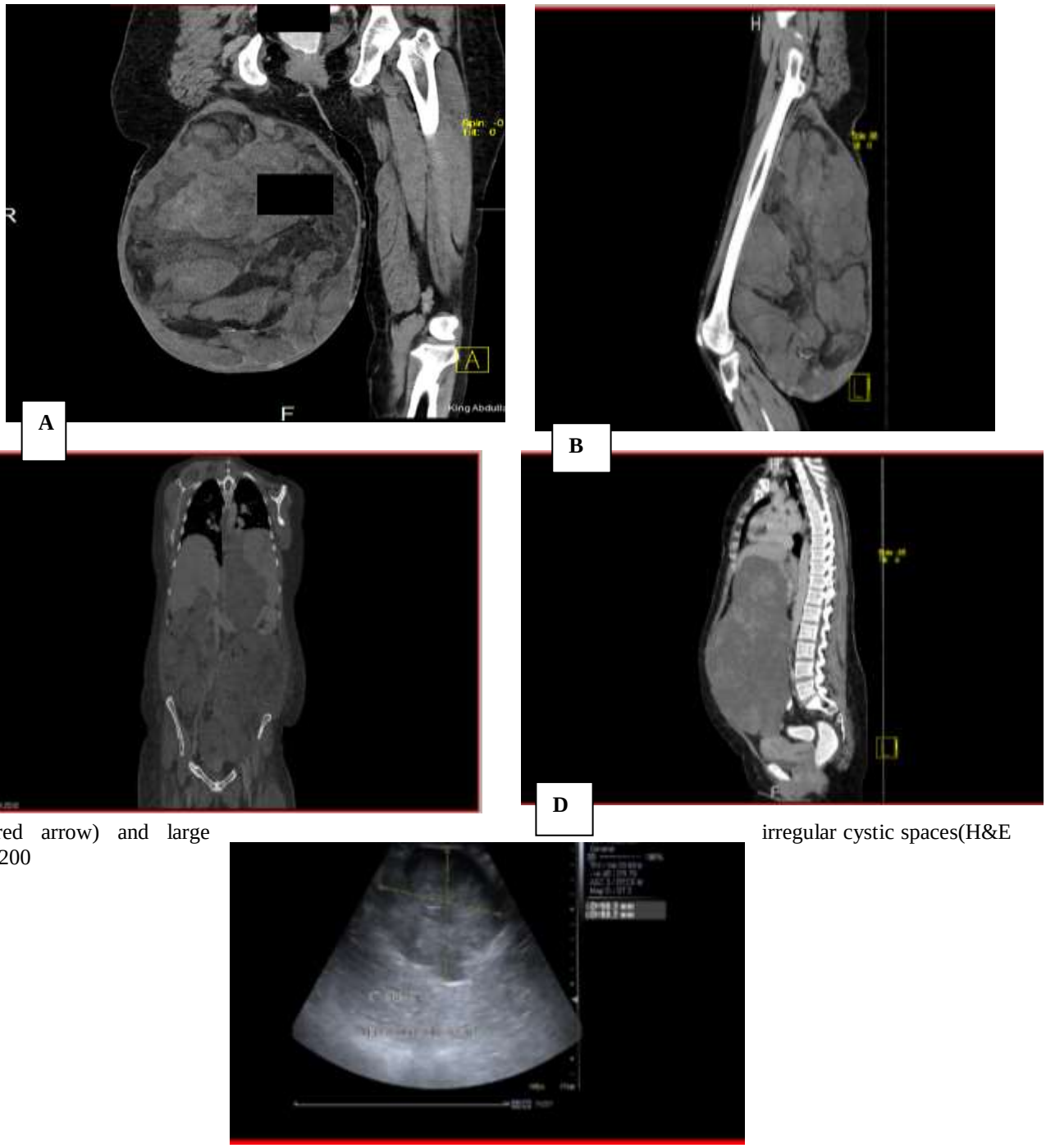
Besides radiotherapy, our patient received doxorubicin and ifosfamide. The combination has proven it efficacy in managing myxoidliposarcoma by a clinical trial [34]. But in our case, the patient died after being diagnosed with her breast metastasis.

Conclusion:-

Although Myxoidliposarcoma is a relatively rare malignancy, its diagnosis should be suspected in the presence of lesion of soft tissues and a histological and imaging investigation should be done without delay to confirm the diagnosis. Patient diagnosed with MLS should beneficiate of a careful follow up to enable an early diagnosis of metastasis. Although surgery is still the gold standard treatment of this kind of tumor, recent advance in managing MLS could improve the prognosis of unresectable lesions



MyxoidLiposarcoma (A) showing proliferation of monomorphic or fusiform cells (H&E x100)(B) no atypia and no mitotic figures(H&E x200. (C) MyxoidLiposarcomashowingprominent chicken wire vasculature (delicate thin walled arborizing and curving capillaries) in a mucoid matrix (H&E x200). (D) Numerous signet ring lipoblasts (red arrow) and large irregular cystic spaces(H&E x200



A. Right thigh CT scan showing huge mass at the posterior aspect of the right thigh
B. Right thigh CT scan: lateral view of the right thigh huge mass.
C. Abdominal CT scan showing huge abdominal mass extending from the diaphragm to the upper pelvis.
D. Abdominal CT scan: lateral view of the abdominal mass.
E. Ultrasonography of the right breast: Large right breast lesion measuring 10 × 8.8 cm

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