Endometrial Stromal Sarcoma: An Unusual Bone Metastasis in the Proximal Humerus

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Abstract

Myeloma, breast, prostate and lung cancer, are related to most of skeletal lesions in people over 40 years of age, specially at axial skeleton. According to the American Cancer Society, bone metastases are more common than primary bone tumours. Endometrial Stromal Sarcoma (ESS) is a rare tumour (less than 10% of uterine sarcomas and 0.25% of all malignant uterine tumours), affecting women with an average age of 45 years. This disease has a slow clinical development, being classified in low or high grade depending on its behaviour. Treatment is hysterectomy combined with adjuvant radiotherapy to reduce the number of recurrences in the most aggressive types; several authors agree that radiotherapy decreases relapses but does not have a significant impact on survival rate, as well as hormone therapy. Many reported cases describe the appearance of secondary metastases of endometrial stromal sarcoma mainly at the belly (guts), pelvis and lung. Bone metastases are rare and the few cases reported refer to metastasis that take place in the axial skeleton (dorsal and lumbar vertebrae, sacrum, and iliac bones). A woman of 40 years old with a lytic bone lesion at the left proximal humerus came to the office; the lesion was diagnosed by an open biopsy in another Institution. The outcome of this biopsy was a primary humerus tumour (myxofibrosarcoma), but final diagnosis after resection of the whole tumour was bone metastasis of endometrial stromal sarcoma. The purpose of this clinical case is to highlight the possible, although uncommon, involvement of bone tissue by endometrial stromal sarcoma, even in young patients. Therefore we would like to draw attention to complexity of diagnosis of this disease that at first glance was interpreted as a primary bone disease.

Case Presentation

A 40 - year - old female, clinical assistant in a residential care institution for elderly people, came to our office on June 6th, 2014. She reported a history of pain in her left shoulder after exertion, of approximately 10 months of development (September 2013). Pain improved after arm resting, did not restrict her activity nor caused insomnia. She was diagnosed of left shoulder tendinitis and analgesics/anti - inflammatories pills were given, advising her a restriction of certain movements. She was treated with pills for pain control and physical therapy to alleviate the inflammatory phase of the rotator cuff. Among her records, she had undergone a hysterectomy with annexectomy of the left ovary in 2006 due to the existence of calcified myoma. For that reason, she was subsequently treated with hormone replacement therapy. After an episode of deep vein thrombosis in lower limbs (and later pulmonary thromboembolism), she received anticoagulant treatment (Acenocoumarol), which was later replaced by antiplatelet therapy (acetylsalicylic acid). The patient also suffered from thrombophilia and was given aspirin as well, due to the expression of a genetic mutation, C677T of the MTHFR gene (methylenetetrahydrofolate reductase) of homocysteine; on the other hand she did not express the G20210A mutation of the prothrombin gene. As a family history of cancer, his father had been diagnosed of prostatic tumour. Physical examination revealed in duration of soft tissues and local swelling on the left shoulder. Axillary lymphadenopathy was not observed. Active mobility was restricted, while passive activity was complete but extremely painful. A scar of about 4 cm was seen on the lateral side of the left shoulder because the patient had undergone an open biopsy 15 days ago in another institution. The patient had an MRI of the left shoulder before the incisional biopsy, and a thoraco - abdominopelvic CT. MRI showed a big not homogeneous mass (with intra and extra osseous component), of 45 x 43 x 82 mm (AP, lateral, craniocaudal) in the proximal humerus. The tumour was placed in the metaphyseal region with extension to the humeral head and...
The patient had a 10 mm hepatic cyst, without splenomegaly, and lobe (endothelial nodule). No pulmonary nodules were observed. There was an increase in the size of the left thyroid at the mediastinum and no signs of pleural or pericardial effusions were observed. The tumour was diagnosed as a myoma (Figure 2). No lymphadenopathy was identified topographic region of the uterus (the patient had an antecedent of hysterectomy) close to the remnant of the uterine cervix and that was well surrounded and partially calcified of 6 x 10 cm was found in the humerus epiphysis. In addition, a heterogeneous solid pelvic mass, bone and infiltration of soft tissues), in metaphysis and proximal diaphysis, without sclerotic rim and reaching the subchondral bone of the humeral head. The biceps tendon was in its place and without signs of disruption. Axillary lymph nodes were not infiltrated (Figure 1). A big expansive lesion was observed in CT scan, with aggressive features (not-homogeneous, large volume, rupture of the cortical bone and infiltration of soft tissues), in metaphysis and proximal humerus epiphysis. In addition, a heterogeneous solid pelvic mass, well surrounded and partially calcified of 6 x 10 cm was found in the topographic region of the uterus (the patient had an antecedent of hysterectomy) close to the remnant of the uterine cervix and that was diagnoses as a myoma (Figure 2). No lymphadenopathy was identified at the mediastinum and no signs of pleural or pericardial effusions were observed. There was an increase in the size of the left thyroid lobe (endothelial nodule). No pulmonary nodules were observed. The patient had a 10 mm hepatic cyst, without splenomegaly, and absolutely normal adrenal glands and kidneys. Tc99 scan was used to identify a hyperactive lesion in the proximal third of the left humerus, without detecting other sources of uptake. X - ray showed an osteolytic image breaking cortical bone, located in the proximal third of the humerus with the previously described measurements. Possible diagnoses were plasmacytoma, aggressive chondral lesion, aggressive giant cell tumor and proximal humerus osteogenic sarcoma. However, we waited for the outcome of the initial biopsy to define our surgical planning. The outcome of the biopsy described the tumor as an atypical cellular lesion of small and monomorphic cells, scant and clear cytoplasm, oval nuclei with 3 mitosis per 50 fields and mild pleomorphism. There was no presence of osteoid tissue or chondral matrix, as well as residual bone trabeculae. The immunohistochemical study revealed intense vimetin expression, decreased expression of CD99 and focal expression of desmin. The results were negative for osteocalcin, osteonectin, S100, enolase, EMA, CKAEI-AE3, smooth muscle actin, Myo D1 and myogenin. The proliferative index was low, about 3%. With the results of the tests requested diagnosis of malignant mesenchymal neoplasia was established, specifically “myxofibrosarcoma of the proximal humerus”. Extended resection was planned according to the biopsy outcome, which informed of a low malignancy disease. Surgery was performed on June 24th, 2014 by deltopectoral approach after general anaesthesia. A proximal resection of approximately 12 cm of left humerus with free-lesion edge was performed and a reverse shoulder arthroplasty was implanted (Figure 3). Excision of the previous biopsy pathway was performed, as it could not be included in any usual approach. Analysis of the tumour was negative for osteogenic sarcoma (lack of osteoid tissue and bone cells). Low primary grade sarcoma was diagnosed (leiomysarcoma; desmin +). The piece resected showed free-disease edges. On June 25th, she started physiotherapy at our institution and was discharged from the hospital 4 days after surgery with a schedule of exercises suggested by the physiotherapist. The patient came to the outpatient on July 25th, 2014. ROM was 75° of abduction, 70° of forward flexion, with internal and external rotation limited because of the muscular resection performed. The patient did not report pain at rest or during physical activity. She was assessed in our Oncology department, and a thoraco - abdominopelvic CT scan was requested. Multiple pelvic and peritoneal masses were noticed, as well as a mass in liver and in the vicinity of the anterior rectus. The largest mass was about 13 cm. After these new findings, was set up the diagnosis of uterine leiomyosarcoma with bone metastases. She was referred to Gynecology department and started cytoreductive treatment with Gem and Taxotere. A new MRI of the liver was performed and subcapsular metastases were detected. The patient was referred to the Surgery department and a laparotomy with resection of multiple nodules in peritoneum, omentectomy, resection of cervix plus right annexectomy was performed. A new biopsy of the resected mass was requested, and it was finally diagnosed of diffuse low - grade Endometrial Stromal Sarcoma (ESS). The tumour was positive for vimentin, CD10, progesterone and estrogen, and negative for desmin, actin and caldesmon. The expression of Ki67 was around 5%. Follow - up showed good outcome with no signs of local relapse of the disease and with a left shoulder ROM of 95° of abduction, 110° of forward flexion, limited external and internal rotation, and backward flexion of 20°. She did not report joint pain and PET and control CT scan showed no evidence of disease. A colloid cyst in the left lobe of the thyroid (4 x 2 x 2 cm) was detected as an incidental finding.
Appropriate management of the sampling procedure ensures a better therapeutic planning of the disease and its possible complications. Specialized institutions in tumor pathology from the beginning. Therefore, we insist on the efficiency of performing these studies in biopsy placement, which can jeopardize the viability of the extremity. Treatment. Stress must be done in the significance of the initial biopsy that could have compromised definitive treatment. It should be emphasized that initial biopsy was done by a more anatomical reconstruction in a young patient could have been aware of the definitive diagnosis prior to surgery, despite the spreading of the lesion and the degree of joint involvement detected, a more anatomical reconstruction in a young patient could have been performed. It should be emphasized that initial biopsy was done by an incision in a wrong area that could have compromised definitive treatment. Stress must be done in the significance of the initial biopsy placement, which can jeopardize the viability of the extremity. Therefore, we insist on the efficiency of performing these studies in specialized institutions in tumor pathology from the beginning.

Conclusion

A proper preliminary diagnosis is basic to address for a precise therapeutic planning of the disease and its possible complications. Appropriate management of the sampling procedure ensures a better prognostic result for the patient. Sometimes, the analysis of the pathologist (gold standard technique for the diagnoses of tumours) is not an easy practice because of the complexity of some cases and the limitation of the sample obtained during the biopsy procedure. It is essential to know how to analyse the battery of diagnostic tests we have available, as well as to take into account all the possible diagnoses, even if they are uncommon.

References