Malignant giant cell type fibrohistiocitoma of the abdominal wall

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Abstract

Malignant fibrous histiocytoma is a condition involving a tumor of the bone or soft tissues. The tumor often appears in the legs or arms, but it can develop in other parts of the body. The disease is the most common soft tissue cancer that is diagnosed in older adults, and is often diagnosed in people between the ages of 50 and 70. We present a case of giant cell malignant fibrohistiocytoma of the abdominal wall. A 77 year old woman was admitted for further evaluation and treatment of tumor in the right inguinal region of the abdominal wall. She claimed that tumor have appeared 3 months earlier and it was characterized by rapid growth and tenderness. NMR demonstrated expansive heterogeneous mass, 18,6x16,2cm of diameter. We performed total extirpation of the tumor. Postoperative period was uncomplicated and she was discharged from the hospital in good condition. Pathohistological examination confirmed malignant fibrous histiocytoma. Depending on the stage of disease and the depth of invasion by the tumor, surgical resection is the treatment of choice. Radiotherapy, chemotherapy, and immunotherapy are other therapeutic modalities. Long term follow-up with regular chest X-rays and CT scans of the abdomen to detect tumor recurrence, metastasis and any lymph node involvement are mandatory.

Keywords: malignant fibrous histiocytoma, anterior abdominal wall, soft tissue

Introduction

Malignant fibrous histiocytoma, a type of sarcoma, is a malignant neoplasm of uncertain origin that arises both in soft tissue and bone. MFH of soft tissue typically presents in a patient that is approximately 50 to 70 years of age though it can appear at any age. There is a slight male predominance¹. Soft tissue MFH can arise in any part of the body but most commonly in the lower extremity, especially the thigh. Other common locations include the upper extremity and retroperitoneum. Patients often complain of a mass or lump that has arisen over a short period of time ranging from weeks to months. The mass does not usually cause any pain unless it is compressing a nearby nerve. Symptoms such as weight loss and fatigue are not typical but can present in patients with advanced disease². Retroperitoneal tumors can become quite large

Maligni fibrohistiocitoma tipa džinovskih ćelija prednjeg trbušnog zida

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Apstrakt

Maligni fibrohistiocitoma je tumor koji napada kosti ili meka tkiva. Najčešće se pojavljuje na nogama ili rukama, ali može da se razvije i u drugim delovima tela. Ovaj tumor je najčešći dijagnostikovani karcinom mekih tkiva kod starijih odraslih i najčešće se dijagnostikuje kod ljudi između 50 i 70 godina. Mi predstavljamo slučaj malignog fibrohistiocitoma tipa džinovskih ćelija prednjeg trbušnog zida. Pacijentkinja stara 77 godina primljena je u bolnicu radi dijagnostike i lečenja tumorske promene u desnoj ingvinalnoj regiji prednjeg trbušnog zida. Navodi da se promena javila 3 meseca pre pregleda i da je karakterišu rapidni rast i bolna osetljivost. NMR pregled trbuha pokazao je ekspanzivnu heterogenu masu veličine 18,6x16,2cm. Izveli smo totalnu ekstirpaciju tumorske promene. Pacijentkinja je nakon nekomplikovanog postoperativnog perioda otpuštena iz bolnice u dobrom opštem stanju. Patohistološka analiza operativnog materijala pokazala je da se radi o malignom fibrohistiocitomu. U zavisnosti od faze bolesti i dubine invazije tumora hirurška resekcija je metoda terapijskog izbora. Radioterapija, hemioterapija i imunoterapija su drugi terapijski moduli. Dugoročno praćenje sa redovnom Rtg i CT dijagnostikom za otkrivanje rekurentnosti, metastaza i zahvatanja limfnih čvorova je obavezno.

Ključne reči: maligni fibrozni histiocitoma, prednji trbušni zid, meka tkiva

before they are detected as patients do not feel a mass per se but rather associated constitutional symptoms such as anorexia or increased abdominal pressure³.

MFH manifests a broad range of histologic appearances with five sub-types described (Table 1.)⁴

Procedures and tests that healthcare provides use to make a diagnosis include X-rays, Computer tomography (CT) scan, Magnetic resonance imaging (MRI) scan, Bone scan and Biopsy⁵. Once the diagnosis of MFH has been confirmed, an individual treatment plan is made for each patient. Sarcoma treatment requires a multimodality approach and hence a team of physicians will participate in a patient's care. There are essentially three main types of treatment that will need to be coordinated to treat the MFH: Surgery, Radiation and Chemotherapy⁶. Prognostic factors that are known to correlate with survival in patients with MFH include tumor grade, depth, size, metastatic status, patient's age and histological subtype⁷.

Case report

A 77-year-old female patient was admitted to the hospital due to tumor in the right inguinal region of the abdominal wall with central necrosis of skin (Figure 1. and Figure 2.), which the patient claimed to have appeared 3 months earlier. She also claimed that it was characterized by rapid growth and tenderness. Blood count and blood chemistry were within normal range.

Figure 1. Right inguinal region of the abdominal wall with tumor

Figure 2. Right inguinal region of the abdominal wall with central necrosis of skin

NMR examination (Figure 3.) of the pelvis verified that it was expansive heterogeneous mass with the largest diameter of 186mmx162mm located in the right groin, in the subcutaneous tissue. The mass put pressure on the front-lateral abdominal wall and laterally compressed tissue. There was no clear delineation of the oblique abdominal muscle. Femoral artery and vein were behind the tumor, without direct contact. Given the findings, it was decided to do the surgical treatment of these changes.

Figure 3. Expansive heterogeneous mass with the largest diameter of 186mmx162mm (NMR examination)

Surgical treatment and findings

After adequate preoperative preparation the patient underwent surgery. An elliptical incision was made to cut the skin and subcutaneous tissue around tumor borders.

Preparation of tumor borders was done followed by partial resection of fascias m.externus abdominis and m.internus abdominis, and after hemostatic control extirpation of the tumor was done. The size of tumor was 25x30cm (Figure 4.). The extirpation of the tumor was done partly through manual digitoklasis, partly through thermocauter. The tumor was macroscopically partly necrotised with fields with purulent secernation, and partly was encapsulated. Elements of the Scarpa's triangle were prepared, and there was no evidence of infiltration a or v femoralis. Tumor and the surrounding tissue samples were sent to HP analysis in order to confirm the possible tumor dispersion. Lavage was done after checking hemostasis, 2 Redon drains were placed, and surgical wound was reconstructed in layers.

Figure 4. The size of tumor was 25x30cm

Postoperative course

The patient Early postoperative course of spends in the ICU where significant decline in CSF was recorded and it was solved with the compensation of blood products. Further postoperative course was normal in surgery ward and therefore the patient was discharged in good condition. The patient was observed in the normal controls for the next 3 months, and no recurrence of the tumor has been recorded.

Pathological findings

HP analysis of tumor formation verified the proliferation of pleomorphic cells, together with an increased hyperchromatic nuclei and numerous mitotic activities. The large cells with multiple nuclei of malignant morphological features can be seen. Immunohistochemical analysis (vimentin +, CD68 +, S100 +, CEA, PanCK-, NF. Actin + / -, desmin-, SMA-, CD34-), proved it to be a malignant fibrous histiocytoma. HP analysis of the snippets of surrounding tissue verified fragments of connective, muscle and adipose tissue in the absence of tumor proliferation.

Discussion

Malignant Fibrous Histiocytoma is a sarcoma of mesenchymal origin affecting soft tissues of the body, particularly the extremities and retroperitoneum². Rarely, it may affect intra-peritoneal organs^{8,9}. Its occurrence has been reported in almost all parts of the body including head and neck, intracranial, intra-abdominal organs and heart muscle. It is considered as the most common soft tissue sarcoma of the adults, but may occur in children as well as infants. There are approximately 12.000 new adult sarcoma cases each year, encompassing seventy different histologic types of mesenchymal tumors that arise from bone, cartilage, fat, muscle, blood vessels, or other connective or supportive tissue¹⁰. MFH is an aggressive tumour with a high potential of metastasis to the other parts of the body. The vast majority of metastatic disease from sarcomas including MFH present as pulmonary disease (90%). Involvement of extra-pulmonary sites is uncommon: lymph nodes (10%), bone (8%), liver (1%). The current treatment of choice for primary malignant fibrous histiocytomas is surgical resection, which involves wide excision of the tumour with an aim for tumour free margins¹⁰. Recurrence of the tumour is not uncommon even when resection margins are tumour free. Metastasis may present months or years after resection of the primary lesion. Treatment for metastatic disease is surgical where possible. Palliative surgery may be carried out if complete resection is not possible. The role of adjuvant radiotherapy and chemotherapy is not clear in the case of retroperitoneal and visceral sarcomas. There are studies that suggest no improvement in overall survival after systemic chemotherapy^{11,12,13}. Some advocate the use of chemoembolization for unresectable metastatic sarcomas, which can provide durable tumour response¹⁴. Operative planning needs to detail carefully the extent of disease at the initial surgical treatment and more specifically to document the size and depth of the tumor with respect to muscle and deep fascia. Radioisotope scanning, computer tomographic scanning and arteriography are techniques that can assist the operative plans. Any surgeon who undertakes the excision of a soft-tissue mass should have the experience and technical ability to proceed with definitive ablation of the lesion. The surgical pathologist should be expected to render a definitive working tissue diagnosis. Radical surgical removal of the primary tumor is an essential first step, but that is not enough-there is an urgent need for an effective postsurgical adjuvant treatment program¹⁵.

Factors of prognostic importance in patients with soft tissue sarcomas are histologic grade, site (proximal versus distal location), size, and lymph node involvement¹⁶. Lot of study demonstrates that the major prognostic factors for MFH are size and grade of the primary tumor. Most reports suggest that the prognosis associated with abdominal MFH is poorer than tumors in the extremities, due to late detection owing to location.

Berton reported 78 cases of MFH "located and deeply seated in the extremities" and found the prognosis to be worse for tumors larger than 5cm in diameter, tumors that recurred early (less than 1 year after surgery), and tumors that had inadequate surgical treatment (intralesional or marginal excision). The overall 5-year survival rate was only 36%^{17, 18}. Kearney reported 167 cases of MFH from the Mayo Clinic and found the most important prognostic factors to be the depth of the tumor and proximal versus distal location. Their study showed no difference

in prognosis between the different histologic subtypes¹⁸. In a review of 200 cases of MFH, Weiss and Enzinger found that larger and more deeply located tumors metastasized most frequently. Their analysis of MFH showed that the 2-year survival rate with the storiform-pleomorphic type of MFH is 60% and the rate of metastases is 42%. Myxoid tumors have low metastatic propensity (13%, 10-year metastatic rate) compared to non-myxoid tumors (40%, 10-year metastatic rate)⁵.

Depending on the stage of disease and the depth of invasion by the tumor, surgical resection is the treatment of choice. Radiotherapy, chemotherapy, and immunotherapy are other therapeutic modalities. Long term follow-up with regular chest X-rays and CT scans of the abdomen to detect tumor recurrence, metastasis and any lymph node involvement are mandatory⁸.

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