Malignant fibrous histiocytoma: A case report.

Chayada Kokpol MD,

Kumutnart Chanprapaph MD.

ABSTRACT:

KOKPOL C, CHANPRAPAPH K. MALIGNANT FIBROUS HISTIOCYTOMA: A CASE REPORT. THAI J DERMATOL 2011; 27: 306-311.

DIVISION OF DERMATOLOGY, DEPARTMENT OF MEDICINE, RAMATHIBODI HOSPITAL, MAHIDOL UNIVERSITY, BANGKOK, THAILAND.

Malignant fibrous histiocytoma (MFH) is a high-grade pleomorphic sarcoma. It is one of the most common adult soft tissue sarcoma mainly affecting deep structures. It is quite uncommon for MFH to present as a primary tumor of the skin. We report a case of cutaneous MFH presenting on the arm of 32-year-old woman. Surgical excision was done with complete tumor removal. There was no evidence of recurrence during the period of 2-year follow up.

Keywords: Malignant fibrous histiocytoma (MFH)

บทคัดย่อ :

ชญาดา ก๊กผล กุมุทนาท จันทร์ประภาพ รายงานผู้ป่วยโรค MALIGNANT FIBROUS HISTIOCYTOMA วารสารโรคผิวหนัง 2554; 27: 306-311.

หน่วยโรคผิวหนัง ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ โรงพยาบาลรามาธิบดี มหาวิทยาลัยมหิดล

Malignant fibrous histiocytoma (MFH) จัดอยู่ในกลุ่มมะเร็งเนื้อเยื่อเกี่ยวพัน ที่มีความรุนแรงสูง ถึงแม้ว่า MFH จะเป็นมะเร็งในกลุ่มเนื้อเยื่อเกี่ยวพันที่พบได้ค่อนข้างบ่อยในวัยผู้ใหญ่ แต่ส่วนมากจะพบในรูปแบบที่เป็นอยู่ในชั้นลึกใต้พังผืดลงไปสำหรับ MFH ที่มาด้วยก้อนในชั้นผิวหนังนั้น พบได้ค่อนข้างน้อย รายงายฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงอายุ 32 ปี มาด้วยก้อนที่แขนขวา ผลการตรวจทางพยาธิวิทยาเข้าได้กับ MFH ได้ทำการรักษาด้วยการตัดออก จากนั้นได้ทำการติดตามผู้ป่วยเป็นระยะเวลา 2 ปี ยังไม่พบการกลับเป็นซ้ำของมะเร็ง

คำสำคัญ: Malignant fibrous histiocytoma (MFH), มะเร็งของเนื้อเยื่อเกี่ยวพัน

Case report

A 32-year-old female was referred to the dermatology outpatient clinic for an evaluation of asymptomatic solitary slow-growing erythematous mass at right arm for 3 months. This mass was accidentally hit by a solid material causing it to ulcerated and bleed (Figure1). She lacked other systemic symptoms. Her past medical history disclosed overlapping rheumatoid arthritis with systemic lupus erythematosus since 2007. Her disease was well controlled.

Figure 1 Clinical picture demonstrates solitary well-circumscribed reddish ulcerated nodule, firm consistency with contact bleeding on the right arm

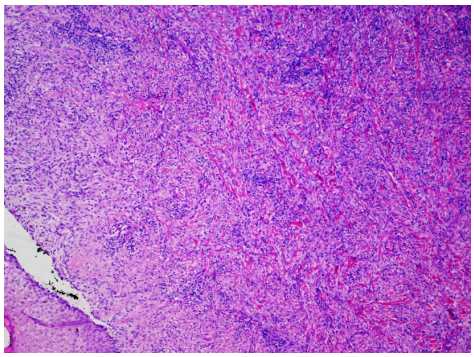

Figure 2 Histopathology (x 40 magnification) shows dense diffuse infiltrate of lymphocytes, histiocytes, fibroblasts admixed with atypical spindle and polyglonal cells in the dermis extending into the subcutis

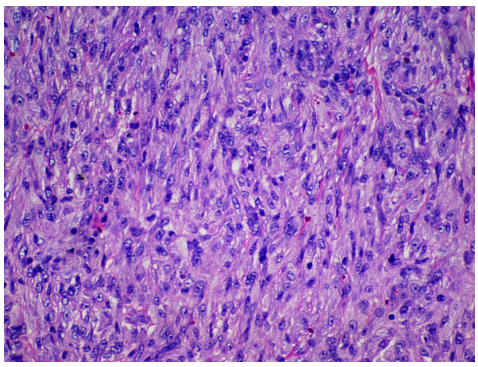

Figure 3 Histopathology (x200 magnification) shows large atypical cells with pleomorphic nuclei. Many are in mitosis and some have atypical mitotic figures.

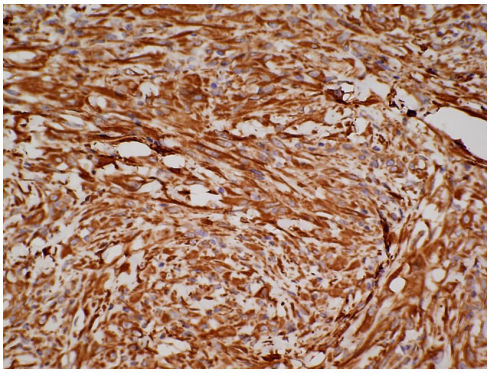

Figure 4 Immunohistochemistry: staining positive with Vimentin

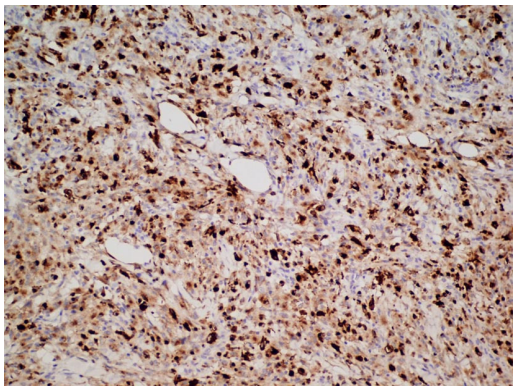

Figure 5 Immunohistochemistry: staining positive with CD68

โฆษณา 4 สี

Examination demonstrated a solitary well-circumscribed reddish ulcerated nodule, firm consistency with contact bleeding at the right arm. No regional lymphadenopathy was found. A 4 mm punch biopsy was obtained from the mass and revealed dense diffuse infiltrate of lymphocytes, histiocytes, fibroblasts admixed with atypical spindle and polyglonal cells in the dermis extending into subcutis. Atypical cells showing large pleomorphic nuclei, many in mitosis and some have atypical mitotic figures. The tumor was further investigated with immunohistochemistry. These atypical cells had positive staining with vimentin, focal positive with CD68 but negative with S100. (Figure 2-5) Clinicopathologic correlation of the biopsy findings resulted in a diagnosis of malignant fibrous histiocytoma. This tumor had been completely excised with wide surgical margin and no recurrences were noted during the 2 years of follow-up.

Discussion

Fibrohistiocytic tumors are heterogeneous groups of soft tissue neoplasms composed of cells that resemble fibroblasts and/or histiocytes. They can be classified according to their malignant potential. Malignant fibrous histiocytoma (MFH) is considered to be the most aggressive form of the fibrohistiocytic tumors.[1]

Malignant fibrous histiocytoma (MFH) is defined as a high-grade pleomorphic sarcoma. It is the most common soft tissue sarcoma of late adult life, occurring between the fifth and seventh decade and rarely occurs in young individuals.[2] It occurs more commonly in whites and have a slight male predominance.[1] It involves the deep soft tissue and the striated muscles of the extremities or the retroperitoneum. However, it is uncommon as a primary tumor of the skin.[3] The pathogenesis and etiology of this soft tissue sarcoma has not been well defined. In the past, the tumor was thought to be of histiocytic origin. But now there are some evidence to point out that MFH may arise from a primitive mesenchymal cell with potential to differentiate in a fibroblastic or histiocytic response.[3,4]

MFH could be classified as either superficial or deep. Superficial variant is usually confined to the subcutaneous tissue, extend only down to the fascia. In contrast to deep variant which can go below the fascia into muscle and bone. The majority of MFH (>90%) are deep tumor located beneath the fascia.[1,5]

The superficial variant of MFH presents as a painless enlarging nodule, which may be 5-10 cm in size with or without ulceration at the time of presentation. The tumor is usually solitary but it can be multinodular, occurring most commonly on the extremities of the elderly (70-75%). The head and neck region is the second most common location for MFH (10%).[1,6]

The diagnosis of MFH is difficult. Since clinical finding is not specific, therefore, diagnosis relies on histopathology. MFH can be divided into five histological subtypes.[1] (Table 1) The storiform-pleomorphic MFH is the most common variant (65%). This type of MFH looks similar to atypical

fibroxanthoma (AFX).[7] The histopathology of storiform-pleomorphic MFH and AFX has interlacing fascicles of pleomorphic spindle-shaped cell. Mitotic figures are common and often atypical. However, the picture of aggressive behavior helps us to differentiate between these tumors. MFH have deeper invasion (deep subcutis/fascia), necrosis and perineural/perivascular invasion when compared to AFX.[6]

Immunohistochemistry of MFH stain positive for vimentin, smooth muscle actin and CD68, which is similar to AFX. Nevertheless, strong positivity of CD74 was shown only by MFH. While MFH is negative for CD99, AFX is positive for CD99 in 35-75% of cases.[6,8,9]

MFH is an aggressive sarcoma; tumor location, size, and histological grading directly influence prognosis.[10] Superficial tumors are generally smaller than deep tumors, has fewer rate of metastasis and therefore, have better prognosis.[3,5]

Lung is most common organ for metastasis (90%), followed by lymph nodes, bone and liver. Cutaneous metastasis have been reported.[11] The overall 5-year metastatic free survival rate of MFH is around 60%.[3]

The standard treatment of MFH is surgical resection. As with other aggressive tumor, MFH has a tendency to invade through the fascial plane, beyond the gross tumor margin. A clearance rate with wide local excision is low. Despite wide excisional margin of 3-5 cm, recurrence rate remains high (40-50%) especially for larger and deeper tumors.[6,7] Mohs micrographic surgery (MMS) could be the best surgical option for MFH because it allows immediate evaluation of tumor margin, yields a higher clearance rate and offers tissue conservation.[12]

Since MFH has high rates of local recurrence and metastases, therefore, adjuvant radiotherapy and/or chemotherapy are recommend in case of unresectable or high risk MFH.[1,3]

Table 1 MFH subtypes[3,13]

MFH subtype	Histopathologic features
Storiform-pleomorphic	Variable appearance spindle, ovoid, giant cells in whorls (fascicles), frequent and abnormal mitoses
Myxoid	>50% myxoid material arcs of blood vessel
Giant cell	Osteoclast-like giant cells, foci of osteoid or mature bone
Inflammatory	Xanthoma cells, inflammatory neutrophilic infiltrate
Angiomatoid	Tumor containing blood filled cystic space

References

1. Stadler FJ, Scott GA, Brown MD. Malignant fibrous tumors. Semin Cutan Med Surg 1998;17:141–52.
2. Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer 1978;41:2250–66.
3. Rothman AE, Lowitt MH, Pfau RG. Pediatric cutaneous malignant fibrous histiocytoma. J Am Acad Dermatol 2000;42:371–3.
4. Cooper JZ, Brown MD. Malignant fibrous tumors of the dermis. In: Wolff K, et al., editors. Fitzpatrick's Dermatology in general medicine. 7th ed. US: McGrawHill 2008. 1162-3
5. Enzinger FM, Weiss SW, editors. Soft tissue tumors. 3rd ed. St. Louis: CV Mosby; 1995.
6. Withers AHJ, Brougham NDL, Barber RM, Tan ST. Atypical fibroxanthoma and malignant fibrous histiocytoma. J Plast Reconstr Aesthet Surg 2011;64:e273–8.
7. Love WE, Schmitt AR, Bordeaux JS. Management of unusual cutaneous malignancies: atypical fibroxanthoma, malignant fibrous histiocytoma, sebaceous carcinoma, extramammary Paget disease. Dermatol Clin 2011;29:201–16, viii.
8. Fletcher CDM. Diagnostic histopathology of tumors. 3rd ed. US: Churchill Livingstone; 2007.
9. Weedon D, Williamson R, Mirza B. CD10, a useful marker for atypical fibroxanthomas. Am J Dermatopathol 2005;27:181.
10. Kearney MM, Soule EH, Ivins JC. Malignant fibrous histiocytoma: a retrospective study of 167 cases. Cancer 1980;45:167–78.
11. Lew W, Lim HS, Kim YC. Cutaneous metastatic malignant fibrous histiocytoma. J Am Acad Dermatol 2003;48:S39–40.
12. Brown MD, Swanson NA. Treatment of malignant fibrous histiocytoma and atypical fibrous xanthomas with micrographic surgery. J Dermatol Surg Oncol 1989;15:1287–92.
13. Rosenberg AE. Malignant fibrous histiocytoma: past, present, and future. Skeletal Radiol 2003;32:613-618.