British Journal of Medicine & Medical Research
7(8): 711-717, 2015, Article no.BJMMR.2015.380
ISSN: 2231-0614

SCIENCEDOMAIN *international*
www.sciencedomain.org

Extensive Right Pleural and Chest Wall Malignant Fibrous Histiocytoma at an Unusual Age: Computed Tomography Features

Ademola J. Adekanmi[1*] and Mustapha A. Ajani[2]

[1]Department of Radiology, College of Medicine, University College Hospital, Ibadan, Nigeria.
[2]Department of Pathology, University College Hospital, Ibadan, Nigeria.

Authors' contributions

This work was carried out in collaboration between both authors. Author AAJ discovered the case, wrote the first draft of the manuscript and managed the literature searches. Author MAA performed the histopathology and wrote the histopathology report and description. Both authors read and approved the final manuscript.

Article Information

DOI: 10.9734/BJMMR/2015/16463
Editor(s):
(1) Franciszek Burdan, Experimental Teratology Unit, Human Anatomy Department, Medical University of Lublin, Poland and Radiology Department, St. John's Cancer Center, Poland.
Reviewers:
(1) Yoshifumi Nakayama, Wakamatsu Hospital, University of Occupational and Environmental Health, Japan.
(2) Koji Teramoto, Department of Medical Oncology, Shiga University of Medical Science, Japan.
(3) Stathopoulos Konstantinos, Department of Medical Imaging and Interventional Radiology, "Jules Bordet" Institut-Tumors Center of Ulb-Brussels, Belgium.
Complete Peer review History: http://www.sciencedomain.org/review-history.php?iid=948&id=12&aid=8551

Case Study

Received 2nd February 2015
Accepted 7th March 2015
Published 23rd March 2015

ABSTRACT

Aims: To describe a rare case of malignant fibrous histiocytoma at an unusual age of sixteen (16) years. Also to sensitize the medical community to the need for thorough evaluation of an opaque hemithorax and to describe the imaging features of this rare neoplastic disease.
Presentation of Case: This was a 16 year old girl who presented with progressive swelling and recurrent right chest pain of 10 months duration and difficult breathing of 6 weeks duration. There was associated weight loss, dry cough and low grade intermittent fever. She had solitary cervical lymphadenopathy, grade II finger clubbing and low hematocrit.
Discussion: Malignant fibrous histiocytomas (MFHs) are tumors of adulthood with a mean age of 59 years. It has predilection for the extremities, the abdominal cavity and the retroperitoneum. Primary pleural occurrence is relatively rare. This is a rare case of an extensive malignant fibrous

**Corresponding author: Email: kanmiademola@gmail.com, kanmiademola@hotmail.com;*

histiocytoma of the right pleura with chest wall involvement presenting at an unusual young age of 16 years, at variance with the ages documented in the literatures. Imaging findings of the histologically proven tumor were also described on high resolution chest Computed Tomography.
Conclusion: Malignant fibrous histiocytomas may occur much earlier than the age documented in most literatures. This case showed that not all cases of extensive opaque hemithorax are due to massive pleural effusion and further and better diagnostic imaging will be necessary for prompt and proper management.

Keywords: Fibrous; histiocytoma; pleura; computed tomography.

1. INTRODUCTION

Most pleura or chest wall tumors are often secondaries from cancers of the breast, lung, kidney, thyroid and multiple myeloma of the ribs [1,2]. Primary MFHs of the pleura are now called pleomorphic undifferentiated sarcomas by the World Health Organization (WHO) tumor classification [3]. Pleura MFHs are relatively rare tumors, despite the fact that MFHs are the most common soft tissue sarcomas in adults [4], accounting for about 20-30% of all soft tissue sarcomas. MFHs are known to have predilection for the extremities, abdominal cavity, and retroperitoneum [2], they occur in adults with age range of 32-80 years [5]. This is a case report of an extensive MFH of the right pleura (hemithorax) occurring at a rather young and unusual age of 16 years with late presentation. Also described are the Computed Tomographic features.

2. CASE REPORT

A 16 year old girl who presented with progressive swelling and recurrent right chest pain of 10 months duration and difficulty in breathing of 6 weeks duration. There was associated weight loss, dry cough and low grade intermittent fever. However, there was no associated hemoptysis. Clinical examination revealed a well oriented, chronically ill-looking young girl. Essentially, she had grade 2 finger clubbing and a 0.5 x 0.5 cm solitary right cervical lymph node. No enlarged lymph nodes were found elsewhere. The Vital signs, with the exception of the respiratory rate, were within normal limits; Pulse rate was 74/min, Blood Pressure was 90/60 mmHg.

2.1 Systemic Examination

Essential systemic findings were in the chest and abdomen. Chest examination revealed a respiratory rate of 26/min, outward bulging of the right hemithorax, tracheal deviation to the left, and dull percussion note also in the right hemithorax with markedly reduced breath sounds. Abdominal examination showed tender hepatomegaly and the liver was down by 9 cm below the costal margin in the right mid clavicular line.

2.2 Laboratory and Imaging Findings

Laboratory results showed reduced haematocrit of 25%, other laboratory parameters were within normal range. She had a chest tube inserted due to the initial chest X-ray finding of an opaque right hemithorax from suspected massive pleural effusion and possible intra thoracic mass at the referring hospital. This was said to have drained some hemorrhagic fluid containing some tissues. Chest X-ray taken at the referring hospital was however not provided.

2.3 Histopathology

The patient had percutaneous transthoracic needle biopsy at our hospital and histopathological findings revealed mesenchymal neoplasm composed of proliferating spindle to polygonal cells, some with wavy spindle nuclei exhibiting moderate pleomorphic. These cells were supported by loose myxoid stroma. Skeletal muscle bundles were invaded and destroyed by the tumor cells. Mitotic figures were present but no definite area of necrosis was noted (Figs. 1A and 1B).

Overall features were those of malignant solitary fibrous tumor. Surgery was not contemplated, due to the extensive nature of the disease, the distant cervical node secondary and the late presentation.

2.4 Treatment

She however had chemotherapy with eight courses of Vincristine, Actinimoycin, Cyclophosphamide and prednisolone combination (VAC-P) and palliative radiotherapy. There was slight improvement in patient's condition over time. Patient however died of

cardiorespiratory arrest after six and a half month on palliative treatment.

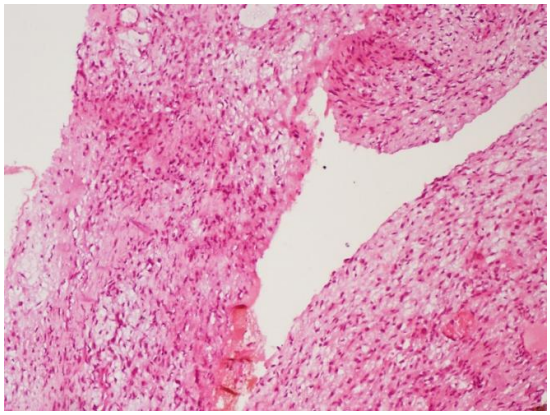

Fig. 1A. Photomicrograph showing a malignant spindle cell neoplasm with increased cellularity supported by loose myxoid stroma. (Haematoxylin and Eosin stain, X100)

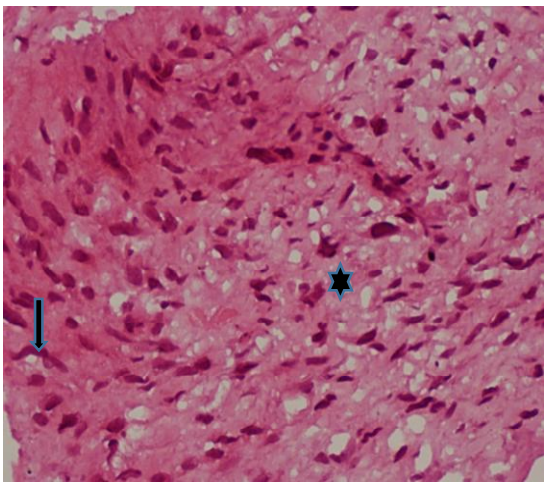

Fig. 1B. Photomicrograph showing a mesenchymal neoplasm composed of proliferating spindle to polygonal cells some with wavy-spindled nuclei (arrow) exhibiting moderate pleomorphism. The cells are supported by loose myxoid stroma (asterisk). (Haematoxylin and Eosin stain, X400)

2.5 Computed Tomography Imaging Findings

High resolution pre and post contrast Computed Tomography scan of the chest was acquired with a 64 slice Multidetector Toshiba Aquilon CT scanner. The plane chest image (scanogram) show complete opacification of the right hemithorax with abnormal lateral bulging of the antero-lateral chest wall (Fig. 2).

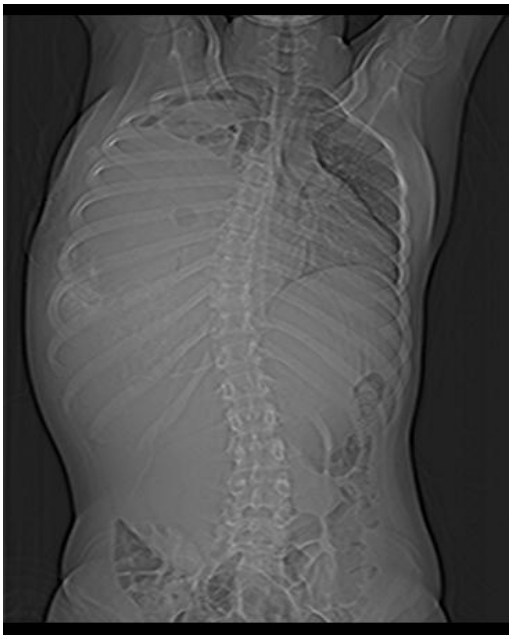

Fig. 2. Plain chest image (CT scanogram) shows almost complete opacification of the right hemithorax, widened intercostal space, medistinal shift to the left and inferior displacement of Right hemidiaphgram and liver inferiorly

Axial (Figs. 3A and 3B) and multiplanar reconstructed Computed Tomography images confirmed complete opacification of the right hemithorax with abnormal lateral bulging of the anterolateral chest wall.

There is an extensive solid heterogeneous mass occupying almost the entire right hemithorax. The mass showed isodense to slightly hypodense mass to the muscle with heavy as well as amorphous calcification, majorly in the periphery of the mass (Fig. 4).

Irregular air pockets were seen within the mass presumably due the chest tube inserted during the course of treatment at the referring hospital and also in the upper thoracic cavity. There was associated expansion of the hemithorax with some extension to the anterolateral chest wall. In addition, there was atelectasis of the right lower lobe and middle lobe, with displacement of the mediastinum to the left and some extension of the right hemithorax massacross the midline to the left were noted. The aforementioned mass also showed inferior extension with displacement

of the ipsilateral hemidiaphgram, the liver and the kidney inferiorly. Associated widening of the intercostal spaces with destruction of the 6th to 8th rib lateral aspect were also noted (Fig. 5).

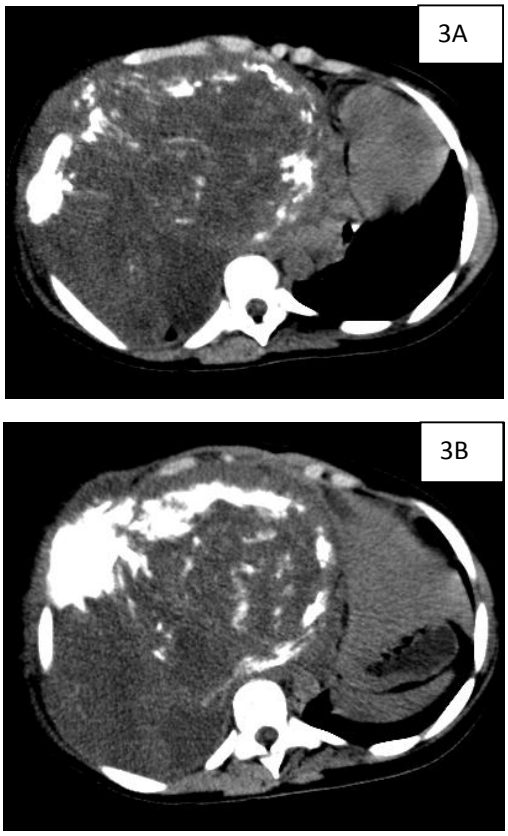

Fig. 3A & B. Non contrast Axial CT images showing the extensive heterogeneous mass with heavy coarse calcification and extension across the midline to the left

3. DISCUSSION

MFHs are deep seated pleomorphic sarcomas of controversial origin that usually presents in middle aged adults [2,6]. Although Jin-Kyung et al. [6] reported a case of MFH in a 25 year old, a younger age compared with the ages in the existing literature, this case report appears to be the first to be discovered at the age of 16 years.

MFH was first described by O'Brien and Stout in 1964 as tumors of histiocytic origin [7]. These tumours were documented to be composed of varying amount of fibroblast like and histiocytic like cells in varying proportion [8]. Although, some authors believed this tumor is the end of the spectrum of soft tissue tumors as they become undifferentiated. [2,8,9]. MFHs were however, recognized as an entity and named undifferentiated pleomorphic sarcoma (UPS) in 2002 by the World Health Organization [3]. Using the WHO nomenclature, Guo et al. [8] noted that MFH/UPS always express Vimentin and that some of these tumors showed positivity for myogenic antigen and CD68.

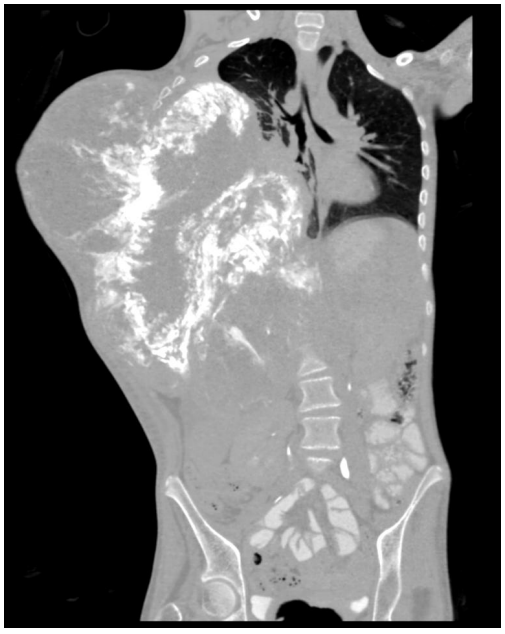

Fig. 4. Coronal MPR images of HRCT of the chest showing the extensive heterogeneous right hemithorax and chest wall mass with heavy coarse calcification surrounding most of the pleura. There is collapsed right lower and middle lobes

MFHs have been classified into subtypes according to their aggressiveness and the cell type [10]. Documented subtypes of MFH in the literature includes the storiform/pleomorphic type, the most frequent histological type accounting for about 50% to 60% of all MFHs. The myxoid type is the second most common but the least aggressive of the MFHs. Others are the giant cell and the inflammatory types [10].

MFH commonly arise from the deep fascia and the skeletal muscles [6,10] of the extremities and the retroperitoneum [6,10,11]. MFHs of the lower extremities account for 59% of all reported cases. Other sites of affectation reported in the literatures are the lung, the liver, the kidney, the bladder, the scrotum and the vas deferens. MFH of the heart, aorta, stomach, small intestine, orbit, the central nervous system, para spinal region, dura matter, facial sinuses, nasal cavity,

oral cavity, nasopharynx, and soft tissues of the neck have all been documented [11]. The pleura and chest wall as reported in this case, is however an unusual and rare site. MFH may develop as a primary tumor or secondary to old pleurisy, pyothorax, radiotherapy and post thoracotomy [12]. This case report appears to be of the primary type as there were no previous clinical history to suggest a secondary MFH in this patient.

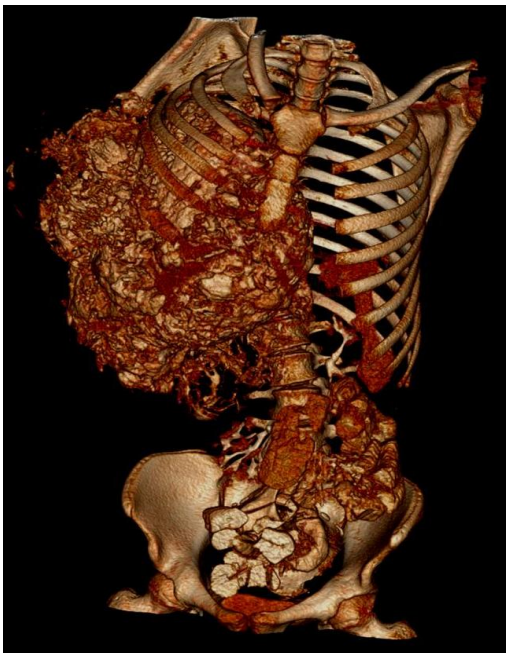

Fig. 5. Volume rendered image shows the extensive right hemithorax mass and overlying 6th–8th rib anterolateral destruction

Clinically, these tumors presents with painless mass that is slow growing in nature over weeks to months. Advanced disease may present with fatigue and weight loss, while large retroperitoneal tumors could present with anorexia and increased abdominal pressure [8], fever and malaise [13]. Other rare clinical presentations were cases of gluteal abscess, episodic hypoglycemia and rapidly enlarging tumour in pregnancy [14].

Associations with non-Hodgkin's and Hodgkin's lymphoma, multiple myeloma and malignant histiocytosis have been documented [12,15]. In this case the patient had slowly enlarging right hemithorax from the progressive tumor growth. There was associated weight loss and intermittent low grade fever. All consistent with the documented features in the literatures [8,13]. The associated chest pain in this case was probably due to pressure effect of the mass on contiguous structures. The grade 2 finger clubbing, though not commonly documented in literatures, could be due to marked reduction in blood oxygen from the extensive lung collapse from the right hemithorax MFH.

3.1 Role of Imaging

Radiological imaging is crucial for detailed description of the tumor including the extent and involvement of adjacent tissues as well as distant metastases, which will be invaluable in surgical planning. Imaging is also essential in the differential diagnosis, follow up and prognostication. Imaging techniques employed in the evaluation of MFH includes plain X-ray, Ultrasonography, Computed Tomography, Magnetic resonance imaging, Nuclear imaging Angiography and Positron emission tomography (PET). Plain films though non-specific may show local soft tissue mass with or without calcifications. Adjacent bone erosion or destruction, a feature that distinguishes it from soft tissue sarcomas except in synovial sarcomas, may be a pointer to the diagnosis [15]. Ultrasonography (USS) has the advantage of being cheap, readily available and ionizing radiation free. USS may show a heterogeneous well-defined, lobulated solid intrathoracic mass which may be isoechoic to muscle with hypoechoic areas of necrosis or hemorrhage and calcific high reflective areas. Computed Tomography is very useful in the description of the MFH soft tissue density mass, usually similar to muscle attenuation, with areas of nodular or peripheral enhancement post contras [15] as well as central hypodense areas due to hemorrhage, necrosis or myxoid area. Osseous involvements like bony erosion, bone destruction or fracture are well visualized on CT. CT show no fat density in MFH, a distinguishing feature from liposarcomas. In this case, CT features were consistent with those documented in the literatures [15]. No fat density was seen on CT images, thereby excluding liposarcomas as a possible differential.

MRI is an excellent imaging modality, particularly for peripheral soft tissue tumors [15]. It is ionizing radiation free unlike most other modalities and has excellent soft tissue contrast in multiple planes. MRI will show the extent of the soft tissue mass by clearly differentiating the tumor from the muscles [15]. MFH is usually heterogeneous in intensity on all sequences due to different

components or from hemorrhage, fibrous tissue and calcifications.

Although MFH signal intensities are non-specific, Myxoid MFH may show low signal intensity on T1-weighted images and prominent high signal intensity on T2-weighted images due to the high water content of these lesions [7,15]. Calcific foci and sub-acute hemorrhage may be seen as low and high signal on both T1-weighted and T2-weighted sequences respectively. While necrotic areas may show a fluid signal pattern. Post Gadolinium scans, typically reveal nodular and peripheral enhancement of MFH solid components. And a relatively well defined tumor margin of low signal intensity presenting the tumor pseudo capsule may be observed [15]. Angiography is nonspecific, but MFH usually shows a hypervascular mass with early venous return [16] while some MFH show hypovascular angiograms [17]. Sen et al. [4] reported the use of Positron emission tomography/computed tomography in defining primary MFH and secondaries. There was an enlarged cervical node, indicating secondary deposit in this case. Definitive diagnosis is however made by histology. Appropriate MRI, PET and Angiography were not available and were not done in this patient.

3.2 Treatment

Management is usually multidisciplinary. Surgical resection of the primary mass is the mainstay of treatment [2,6]. Adjuvant treatment with Radiotherapy and Chemotherapy are usually instituted to prevent local recurrence or secondaries to other organs particularly in patients with high probability of recurrence especially in patients with retroperitoneum and head and neck sarcomas. Although, chemotherapy results have not been shown to be effective and in general, adjuvant therapy has not been proven to improve survival in MFH [18]. Recent studies however, has indicated that Doxorubicin and Ifosfamide are the most active drugs against MFH with documented response rate of 10% and 36% [19]. This patient however had Vincristine, Actinimoycin, Cyclophosphamide and prednisolone combination (VAC-P) possibly due to the initial working diagnosis of intrathoracic tumour before the histopathology result and subjective assessment of some improvement.

MFH prognosis depends on the tumor grade, depth, size, metastatic status, patient's age, and histologic subtype. However, certain features like the tumour location (MFH of the abdominal cavity, the retroperitoneum or the head and neck), a tumor size of 5 cm or more, deep tumor location and high grade tumor generally have poor prognosis [20]. It is worthy of note that most MFH are of the high grade histologic type [21] and usually present with distant metastasis at the time of diagnosis [12]. Literature have also documented that most cases do recur locally despite the institution of aggressive treatment while the overall 5 year survival is documented to be between 25-70% [21].

In this case, the extensive nature of this tumor, bony destruction, compressive features, and the cervical node secondary as well as the relatively young patient's age were pointers to the poor prognosis. And surgery was not contemplated in this case due to the poor prognostic factors.

4. CONCLUSION

This is a case of an extensive malignant fibrous histiocytoma of the pleural and the chest wall occurring at an unusual young age and presenting late to the tertiary health facility. Treatment with radiotherapy and chemotherapy yields no or very little result. This case report emphasize the need for further evaluation and the use of better diagnostic imaging modalities in evaluation of an opaque hemithorax on plain film.

CONSENT

All authors declare that written informed Institutional departmental consent was obtained from the patient.

ETHICAL APPROVAL

It is not applicable.

ACKNOWLEDGEMENTS

We thank all the staff of the Computed tomography unit of the Department of radiology University College Hospital, Ibadan, Nigeria for their support.

COMPETING INTERESTS

Authors declared that no competing interests exist.

REFERENCES

1. Meziane M. Localized fibrous tumor of the pleura [on line]. Cleveland Clinic Foundation.

Available:http://wwwemedicinecom/radio/topic448htm (August 2, 2007)

2. Sajad AS. Malignant fibrous histiocytoma of chest wall. Journal of Pakistan Association of Dermatologists. 2012;22(3): 283-286.
3. WHO. IARC publication.
   Available:http://www.iarc.fr/en/publications/pdfsonline/pat-gen/bb5/bb5-classifsofttissue.pdf.
4. Christophoros NF, Karayannopoulou G, Kostopoulos I, Papakonstantinou C. Giant Malignant Myxoid Fibrous Histiocytoma of the Chest Wall. J Thorac Oncol. 2010;5(6):907. DOI:10.1097/JTO.0b013e3181d873a5.
5. Meyers SP. MRI of bone and soft tissue tumors and tumor like lesions, differential diagnosis and atlas 1/e Meyers. Publisher: Thieme publishing Group; 2008.
6. Jin-Kyung A, Ki Keun O. Malignant fibrous histiocytoma of chest wall. Yonsei Med J. 2005;46:177-179.
7. O'Brien JE, Stout AP. Malignant fibrous xanthomas. Cancer. 1964;17:1445-1455.
8. Guo H, Xiong Y, Nong L, Zhang S, Li T. Reassessment of the pathological diagnosis in 33 cases of malignant fibrous histiocytoma. Beijing Da Xue Bao. 2008;40(4):374-379.
9. Hollowood K, Fletcher CD. Malignant fibrous histiocytoma: Morphologic pattern or pathologic entity? Semin Diagn Pathol. 1995;12:210-220.
10. Kransdorf MJ, Murphey MD. Imaging of Soft Tissue Tumors. WB Saunders Company, Philadelphia; 1997.
11. Sabesan T, Xuexi W, Yongfa Q, Pingzhang T, Ilankovan V. Malignant fibrous histiocytoma: Outcome of tumors in the head and neck compared with those in the trunk and extremities. Br J Oral Maxillofac Surg Epub Jul 18, 05 [Medline]. 2006;44(3):209-212.
12. Nonaka M, Kadokura M, Ohkubo F.Post radiation inflammatory malignant fibrous histiocytoma arising from the chest wall. Ann Thorac Cardiovasc Surg. 2001;7:371-374.
13. Makimoto Y, Yamamoto S, Takano H, Motoori K, Ueda T, Kazama T. Imaging findings of radiation induced sarcoma of the head and neck. Br J Radiol, [Medline]. 2007;80(958):790-797.
14. Falidas E, Rallis E, Ntatsi A. Malignant fibrous histiocytoma masquerading as gluteal abscess. J Cutan Med Surg. 2011;15:345-345.
15. Yip D. Malignant Fibrous Histiocytoma Imaging. E medicine, 10/18/11.
16. Ros PR, Viamonte MJ, Rywlin AM. Malignant fibrous histiocytoma: mesenchymal tumor of ubiquitous origin. AJR Am J Roentgenol. 1984;142:753–759.
17. Burgener FA, Landman S. Angiographic features of malignant fibrous histiocytomas. Radiology. 1976;121:581–583.
18. Liarmakopoulos E, Lampropoulos P, Marinis A, Markakis C, Manoloudaki K, Chatzimarkou A, Rizos S. Soft tissue paraspinal inflammatory malignant fibrous histiocytoma presenting as a lumbar abscess. Case Rep Oncol. 2011;4:343–349.
19. Nakayama Y, Minagawa N, Torique T, Yamaguch K. Malignant fibrous histiocytoma originating from the mesorectum: A case report. World J Surg Oncol. 2011;9:15. (Published online 2011 February 2). DOI: 10.1186/1477-7819-9-15
20. Oda Y, Tamiya S, Oshiro Yea. Reassessment and clinicopathological prognostic factors of malignant fibrous histiocytoma of soft parts. Pathol Int. 2002;52:595-606.
21. Ahlén J, Enberg U, Larsson C, Larrson O, Frisk T, Brosjö O, von Rossen A. Malignant fibrous histiocytoma, aggressive fibromatosis and benign fibrous tumors express mRNA for the metalloproteinase inducer EMMPRIN and the metalloproteinases MMP-2 and MT1-MMP. Sarcoma. 2001;5(3):143-9. DOI:10.1080/13577140120048601.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sciencedomain.org/review-history.php?iid=948&id=12&aid=8551