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Malignant fibrous histiocytoma of the oral and maxillofacial region: a report of three cases

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ABSTRACT

Malignant fibrous histiocytoma (MFH) is a pleomorphic soft tissue sarcoma. Three cases of MFH were reported in our study. The first case involved in the right infratemporal fossa of a 32-year-old female was presented. MR imaging revealed a 5.0 × 3.3 cm soft tissue mass of inhomogeneous high signal intensity. The second case was found in the right hard palate of a 66-year-old male. CT demonstrated bone destruction and MR imaging showed a 4 × 4 cm sized soft tissue mass of heterogeneous high signal intensity. The final case was found in the left masticator space of a 37-year-old male. The CT image showed a large mass with massive bone destruction of the left mandibular ramus, while the MRI displayed a soft tissue mass, 8 cm diameter. Our cases exhibited the general features of MFH. MRI is essential in the imaging of MFH, namely to depict tumor borders and demonstrate relationships with adjacent structures. ***(Korean J Oral Maxillofac Radiol 2003; 33 : 239-44)***

KEY WORDS : Histiocytoma, Fibrous; Magnetic Resonance Imaging; Head and Neck Cancer

Malignant fibrous histiocytoma (MFH) is a pleomorphic soft tissue sarcoma. Previous designated MFH are lesions which have been diagnosed as malignant fibrous xanthoma,[1] malignant giant cell tumors of the soft tissues,[2] fibroxanthoma,[3] fibroxanthosarcoma,[4] xanthogranuloma, xanthosarcoma,[5] inflammatory fibrous histiocytoma[6,7] and malignant histiocytoma.[8] Patients with MFH ranged in age from 5 to 93 years. The tumor had a peak incidence in the seventh decade. This condition is twice as common in men as in women.[9] It is usually occurred in the extremities, particularly the thigh, arising in deep fascia or skeletal muscle. MFH was initially described in 1964 by O'Brien and Stout, who considered it to have a histiocytic origin. However, it has remained a controversial entity because of its uncertain histiogenesis.[1] It is now more convenient, for the purposes of description and comparison of behavior, to divide MFH into several subgroups proposed by Weiss,[9] including storiform-pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid types.

Although MFH is the most common malignant tumor of the soft tissue in adults,[9] it is extremely rare in the oral and maxillofacial region. The purpose of this article is to present three cases of MFH in the oral and maxillofacial region and to discuss the clinical and radiological features with emphasis on the MR images.

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Case reports

Case 1

A 32-year-old female had right preauricular swelling and pain for 6 months and visited our institute in February 1998. On the extraoral examination, 5 cm diameter fluctuating mass with induration on the right preauricular area and swelling from the right ascending ramus of the mandible to the right mandibular angle area with pain was found. Intraoral examination revealed a white ulcerative lesion on the right buccal mucosa.

Conventional radiographs showed an extensively destructive lesion on the lateral wall of the right maxillary sinus and the posterior alveolar portion. Right maxillary 1st and 2nd molars were displaced. On computed tomograph (CT), there was a soft tissue mass that was in touch with the middle cranial fossa upwards in the infratemporal fossa. Thinning of the cranial base and slight enlargement of the oval foramen were observed (Fig. 1A). In the lower portion of the mass, thinning or absence of adjacent bone was observed near the ascending ramus of the mandible (Fig. 1B). Displacement or

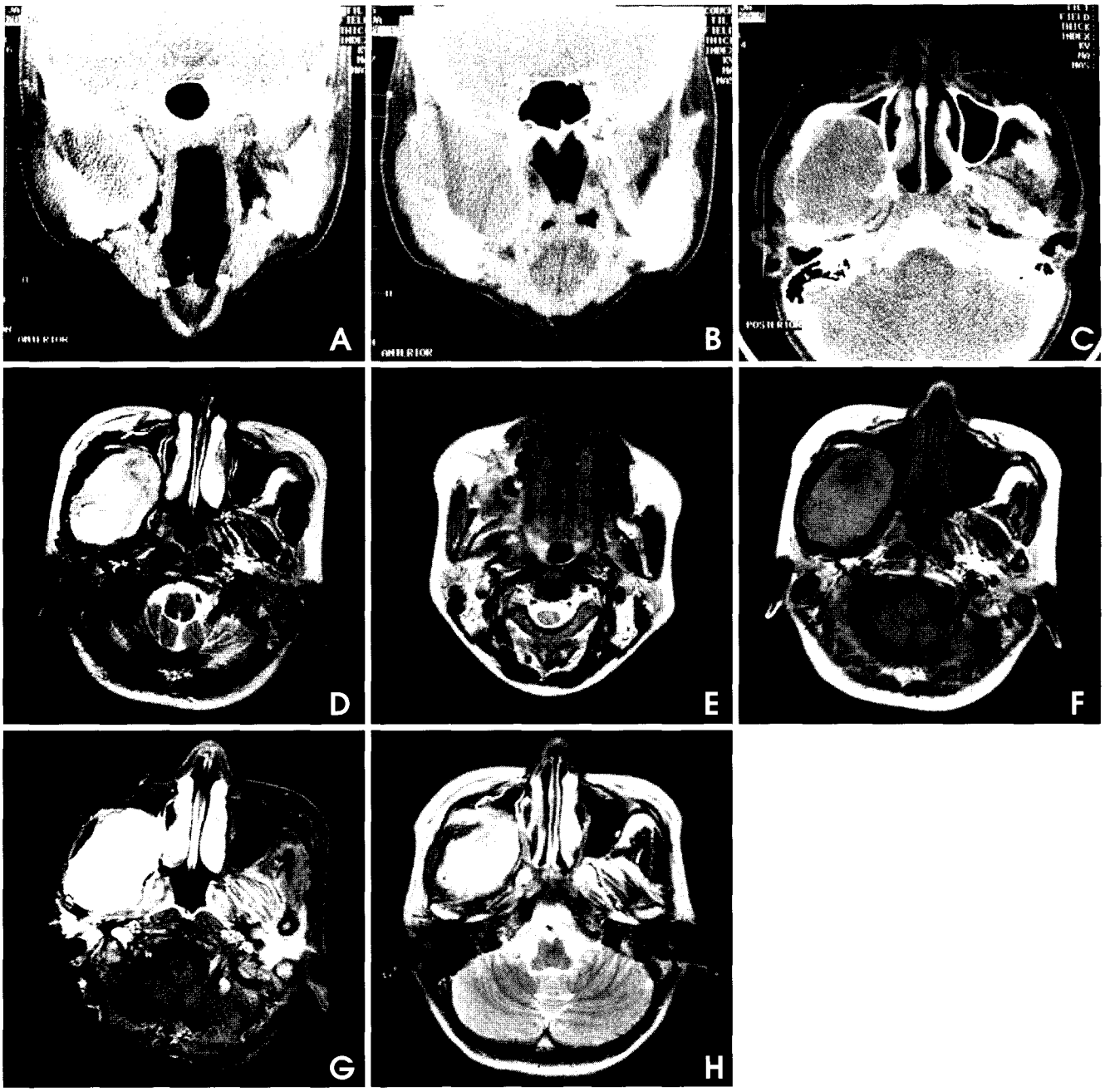

Fig. 1. shows the images from case 1. A, The coronal CT shows a round soft tissue mass in the infratemporal fossa. Thinning of the cranial base and slight enlargement of the oval foramen are observed. B, The coronal CT shows the thinning of the ascending ramus of mandible in the lower portion of the mass. C, The axial CT shows the displacement of the posterior wall of right maxillary sinus. Thinning of the right pterygoid plate is found. D, T2-weighted image shows 5.0 × 3.3 cm soft tissue mass. Multiple areas of increased signal intensity are present posteriorly and those of low attenuation are present anteriorly. E, T2-weighted image shows indistinct border of the lower portion of the lesion. F, T1-weighted image shows inhomogeneous intermediate to low signal intensity within the mass. G, Contrast enhanced T1 weighted image shows intensely enhanced mass. H, The tumor mass is not decreased in size after the chemotherapy.

absence of adjacent bone in the posterior wall of maxillary sinus and pterygoid plate was also observed (Fig. 1C). The tumor showed lower or iso-attenuation to surrounding muscles. Magnetic resonance image (MRI) was taken. T2-weighted image showed 5.0 × 3.3 cm soft tissue mass showing inhomogeneous high signal intensity in the infratemporal fossa (Fig. 1D). In the lower portion of the lesion, the signal intensity was decreased relatively and the discrimination with the

adjacent structures was difficult (Fig. 1E). T1-weighted image showed a soft tissue mass of inhomogeneous intermediate to low signal intensity (Fig. 1F). Contrast enhanced T1-weighted image showed intensely enhanced mass except the area showing low signal intensity in T2-weighted image (Fig. 1G).

Malignant fibrous histiocytoma of low grade was diagnosed from incisional biopsy and immunohistochemistry. Chemotherapy was performed and six months later, the tumor mass was not decreased in size on MRI for follow up check (Fig 1H).

Case 2

A 66-year-old male removed the prosthesis of right maxillary molar area at local clinic due to toothache in February 1999. But the sensitivity from cold water and air was increased and the swelling was found on the alveolar bone of the teeth. He was referred to other institute and diagnosed as a malignant peripheral nerve sheath tumor from incisional biopsy. Radiotherapy with 4.5 Gy for 5 weeks was performed and then CT was taken. On CT, a soft tissue mass on the right hard palate with ill defined border destructed right maxilla (Fig. 2A). However, the size of mass was increased progressively. Then he referred to our institute in May 1999.

On clinical examination, there was a 6 × 5 cm sized exophytic dark brown palpable soft tissue mass with induration in the whole right palate including over the midline. Pus discharge and severe foul odor were found. The right maxillary first molar became loosened within the mass.

MRI was taken and T2-weighted image showed a 4 × 4 cm sized soft tissue mass of heterogeneous high signal intensity in the right hard palate and maxillary alveolar process (Fig. 2B). T1-weighted image showed soft tissue mass of low signal intensity (Fig. 2C) and contrast enhanced T1-weighted image showed well enhanced lobulating contoured mass in which had a low signal intensity suggesting necrosis (Fig. 2D). The mass extended into the right maxillary sinus and destructed the inferior and medial wall of the right maxillary sinus. A 3.5 cm sized, round, high enhancing region was found in the right jugulodigastric area suggesting metastatic lymphadenopathy.

Excision of the tumor mass was performed under right radical neck dissection. Right total maxillectomy and left subtotal maxillectomy was performed. The operation site was reconstructed with metal plate with skin graft from left thigh.

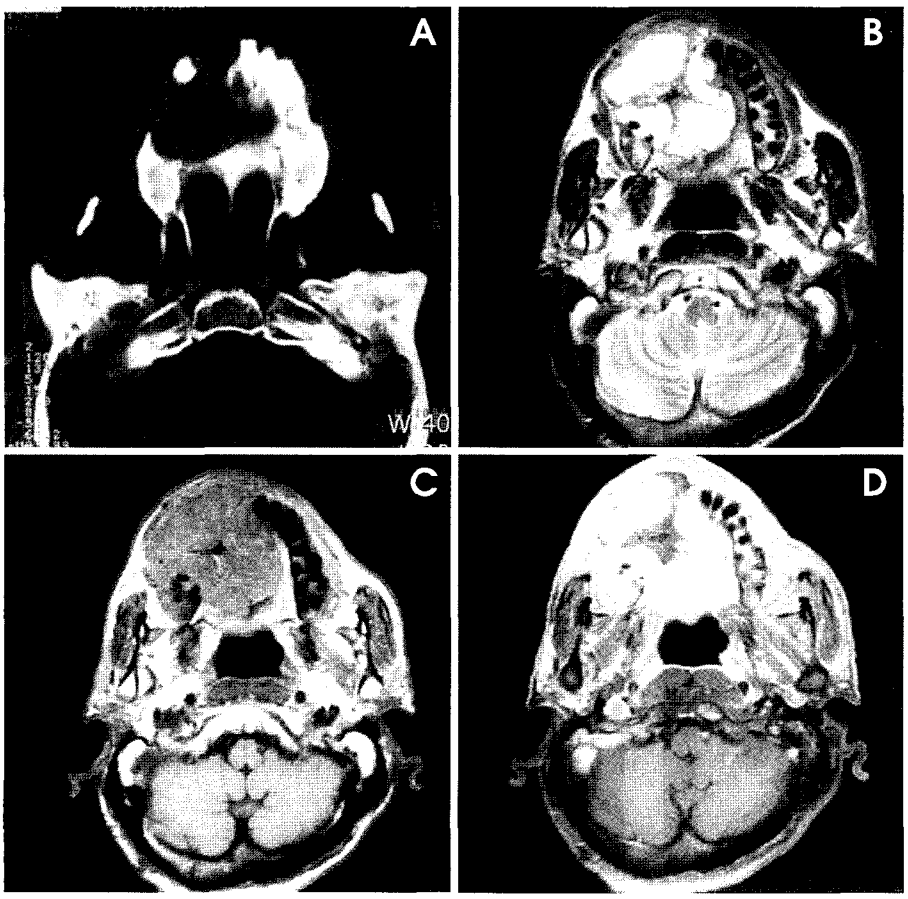

Fig. 2. shows the images from case 3. A, The axial CT shows a bone destruction in the right hard palate and maxillary alveolar process. B, T2-weighted image shows 4 × 4 cm sized lobulated soft tissue mass with heterogeneous high signal intensity. Centrally, low signal intensity separated the mass into several pieces. C, T1-weighted image shows low signal intensity. D, The contrast enhanced T1 weighted image shows well enhanced lobulating contoured mass in which a low signal intensity suggests a necrosis.

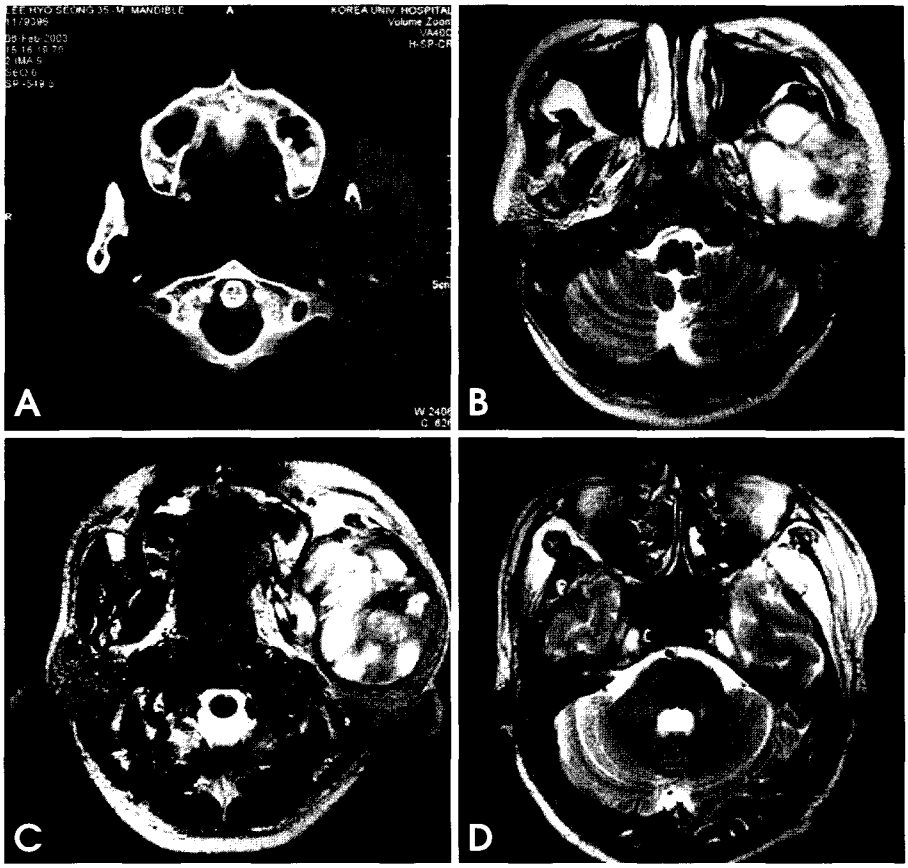

Fig. 3. shows the images from case 3. A, The axial CT shows a massive bone destruction of the left mandibular ramus. B, T2-weighted image shows a lobulated soft tissue mass in left masticator space. C, The central portion of the lesion shows heterogeneous texture due to necrosis in T2-weighted image. D, The skull infiltration along the temporalis muscle is found in T2-weighted image.

Excisional biopsy was performed and the lesion was confirmed as malignant fibrous histiocytoma with histologic grade III.

Case 3

A 37-year-old male had suffered from paresthesia and pain on left mentum area. He visited Korea University hospital for mouth opening limitation in January 2003. Then CT was taken. On CT, a large mass in the left masticator space with massive bone destruction of the left mandibular ramus was found (Fig. 3A). The impression was performed as ameloblastoma and then he was transferred to our institute.

On clinical examination, mouth opening was restricted and maximum mouth opening was 10 mm. Left facial swelling was found although there was no tenderness. Intraorally, an ulcerative lesion of 3 cm diameter was extended from left buccal cheek to left retromolar area with foul odor.

MRI was taken and T2-weighted image showed a soft tissue mass of 8 cm diameter in left masticator space. The center of the lesion was located in left mandibular condyle (Fig. 3B). The central portion of the lesion showed heterogeneous texture supposed to be due to necrosis (Fig. 3C). The upper extent of the tumor mass was infiltrated into skull along the temporalis muscle although the border of the tumor mass was well defined partially (Fig. 3D). In the lower extent, the mass destructed mandible and the enlargement of level II lymph node was shown.

Spindle cell sarcoma, most likely malignant fibrous histiocytoma was diagnosed from punch biopsy and immunohistochemistry. He is under observation.

Discussion

Although MFH is the most common soft tissue sarcoma in adult life and it can occurs in bone and other organs, it is a rare tumor in the oral and maxillofacial region. MFH in bone accounted for 5% of all malignant bone tumors.[10] Three-quarters of the cases were occurred in the extremities, with nearly 50% within the lower limbs. Other involved sites included the retroperitoneum, head and neck, and the abdomen and pelvis.[11] Three percent to 10% of all MFHs were occurred in the head and neck, and in this area, the sinonasal tract was the most common location accounting for 30% of all cases. Other common sites in the head and neck region included the craniofacial bones (15-25%), larynx (10-

15%), soft tissue of the neck (10-15%), major salivary glands (5-15%), and oral cavity (5-15%).[12] Our cases were located in the infratemporal fossa, hard palate, and masticator space.

Conventional radiographs usually showed an aggressive lesion with cortical destruction. Periosteal reaction was infrequent and matrix calcification was rare.

CT can provide critically useful information. In many cases, MFH was intensely enhancing although the lesion was extremely variable.[13] These tumors tended to produce a lobulated mass clearly defined from the surrounding tissues. The masses often had diminished attenuation centrally due to necrosis, hemorrhage, or mucoid material.13 Adjacent bone might show cortical erosion, which was a highly suggestive sign of MFH although this could occasionally be seen in synovial sarcoma.[13] Many of these tumors were iso-attenuating to surrounding muscle and were poorly delineated. The alterations in fat planes and muscle texture were the important clues to the presence of the tumor.

MRI has dramatically improved our ability to evaluate the soft tissue tumors. On T1-weighted images, MFH showed typically of intermediate to low signal intensity, often similar to the surrounding muscle. On T2-weighted images, it tended to be of high signal intensity, although often quite inhomogeneous centrally.[14] MFH showed contrast enhancement that was usually peripheral and nodular. Heterogeneous signal intensities on T2-weighted and contrast enhanced images were also a frequent finding.[15] If bleeding was present within the tumor, it might be demonstrated as areas of high signal intensity on T1-weighted images.[16] Calcification, which could occur in from 5 to 20% of these tumors, was usually best seen on CT but could occasionally be seen on MRI as the areas showing low signal intensity on all sequences.[11] In our cases, conventional radiograph and CT showed a soft tissue mass thinning, displacing and destructing adjacent bone. On MRI, T2-weighted images showed heterogeneous high signal intensity, but the heterogenicity of the signal intensity was varied by the case. T1-weighted images showed inhomogeneous low signal intensity in our cases. Also, MFHs in our cases were enhanced very well. One case showed intensely enhanced mass and others were well enhanced. Within the mass, all three cases showed a low signal intensity that suggested a necrosis. The MRI findings of our cases were similar to that of the cases occurring in other locations. In our patients, tumor encapsulation was present on the MRI scan, apparently on T1-weighted images. Two of three cases had a lobulating contoured border and one case had a round border.

In younger patients osteosarcoma is the most important differential diagnosis, but osteosarcoma frequently shows sclerotic and osteolytic lesions as well as periosteal reactions, which are rare findings in MFH. However, in the patients over 40 years, osteosarcoma more frequently shows a predominantly or purely osteolytic pattern, which has been reported as high as 68%, the differential diagnosis is difficult. Fibrosarcoma may show an identical morphology on the radiographs in the same age groups as MFH, and is also the principal histopathologic differential diagnosis. Osteomyelitis and dedifferential chondrosarcoma can rarely present in a similar fashion. Differential diagnosis on MRI may be more complex and imaging findings even less specific: signal intensities on T1- and T2-weighted images as well as contrast enhancement are similar to those of most other malignant bone tumors. Extraosseous tumor spread and peripheral, nodular contrast enhancement, although frequent findings, can not be a clue for differential diagnosis of MFH from other malignant bone tumors. Nevertheless MRI is an essential imaging modality for diagnosis of MFH to depict tumor borders and demonstrate relationships with adjacent structures, as required for preoperative staging.[15]

The mainstay of treatment was surgical excision whenever possible.[13] It was often combined with radiotherapy, either before or after surgery. The surgical excision was at times difficult to achieve when tumor was located adjacent to vital structures. Chemotherapy might be employed prior to surgical resection of these lesions.[17,18] The prognosis of MFH was not favorable and the recurrence of the tumor was common. The prognosis depended on the site of the primary lesion. The larger and more deeply situated lesions carried a worse prognosis, with recurrence rates of 51% even with radical excision. The recurrences were often multiple.[14] Metastatic disease occurred in 40% of the cases. The recurred sites included the lung, liver, lymph nodes, and bone.[11]

Previous radiotherapy was known to predispose to development of MFH. The occurrence of MFH as a late consequence of irradiation was a well-documented phenomenon. Because there were many vital structures in the head and neck region, wide resection of sarcomas in the jaws and oral cavity was usually restricted or impossible. Consequently, incomplete excision of the sarcoma carried a significantly high risk of local recurrence. The prognosis of post-radiation sarcomas was generally poor.[19,20]

Conclusion

Our cases exhibited general features of MFH, namely sligh-

tly heterogeneous intermediate signal intensity on T1-weighted image and high signal intensity on T2-weighted image. Differential diagnosis on MRI may be more complex and the MR findings are even less specific. Signal intensities on T1- and T2-weighted image as well as contrast enhancement can not differentiate MFH from other malignant bone tumors. Nevertheless MRI is essential in the imaging of MFH to depict tumor borders and demonstrate relationships with adjacent structures.

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