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Radiological Characteristics and Management of Intramuscular Myxoma of the Temporal Muscle: Case Report

Tetsuhiro HIGASHIDA[1]

[1]Department of Neurosurgery, Odawara Municipal Hospital, Odawara, Kanagawa

Abstract

The patient was a 51-year-old male with a 3-year history of a slow-growing, asymptomatic, subcutaneous mass in the left temporal region. Magnetic resonance imaging revealed a well-defined extracranial lesion with heterogeneous enhancement and without invasion of the skull. A variety of soft tissue tumors were included in the differential diagnosis. The patient underwent total resection of the tumor, and a diagnosis of intramuscular myxoma was confirmed histologically. There was no evidence of recurrence at 6-month follow-up. The present case is the first characterization of the radiological appearance of intramuscular myxoma in the temporal muscle. I emphasize that increased awareness of this rare lesion and a careful clinical and radiological preoperative assessment are crucial in determining an appropriate treatment strategy for patients with a soft tissue tumor of the head.

Key words: intramuscular myxoma, temporal muscle, magnetic resonance imaging, angiography

Introduction

Myxoma is a benign, mesenchymal tumor originating from fibroblasts, which produces abundant mucopolysaccharides but is incapable of producing mature collagen.[1)] Soft tissue myxomas are uncommon, accounting for only 0.12% of all soft tissue tumors.[2)] Approximately 17% soft tissue myxomas are intramuscular, with an incidence in the general population of only one per million. Intramuscular myxoma is mostly seen in large skeletal muscles and rare in the head and neck.[1,3–5)] This tumor sometimes gets associated with fibrous dysplasia in the same anatomical region, which is known as Mazabraud's syndrome.[6)] Intramuscular myxoma typically presents as a slow-growing painless mass, but symptoms might occur from compression of the surrounding structures.[3,7)] Because of its low prevalence and nonspecific clinical and radiological features, intramuscular myxoma is difficult to diagnose before surgery and is easily confused with other soft tissue tumors.[8)] However, awareness of this tumor type is particularly important because intramuscular myxoma does not metastasize and does not recur after local excision.[4,9)] To my knowledge, the case presented here is only the third report on intramuscular myxoma originating in the temporal muscle.[10,11)] This report could be helpful for the evaluation of such tumors and for determining an appropriate treatment strategy.

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Case Report

A normal healthy 51-year-old male presented with a 3-year history of a slow-growing asymptomatic mass in the left temporal region. Physical examination revealed a palpable, subcutaneous soft tissue mass in the left temporal area, which had a maximum diameter of > 5 cm. The mass was soft, mobile, and fluctuant, and no tenderness to palpation was evident. The overlying skin was intact. Neurological examination demonstrated no obvious abnormality. The movements of the facial muscles and mandible were symmetrical, and there was no limitation of mouth opening. There was no sensory disturbance observed. Computed tomography (CT) of the head revealed a well-defined, extracranial, subcutaneous mass with a smooth and clear border that separated it from the surrounding tissues (Fig. 1A). No bony destruction was visible. The lesion consisted of a homogeneous low-attenuating component that was heterogeneously enhanced with contrast (Fig. 1B). On magnetic resonance imaging (MRI), the mass appeared homogeneously hypointense on T_1-weighted images (Fig. 1E) and hyperintense on T_2-weighted images (Fig. 1F), and revealed heterogeneous enhancement after gadolinium injection (Fig. 1H).

The tumor signal was not suppressed on fat-suppression images (short T_1 inversion recovery: STIR; Fig. 1G), indicating that the tumor did not have fat-rich components. Because the tumor showed enhancement, angiography was performed to evaluate the vascularity of the tumor, which revealed a poorly vascularized mass without any tumor stain (Fig. 1C, D).

Surgical tumor resection was deemed appropriate in order to obtain a pathological diagnosis and resolve his cosmetic concerns. Under general anesthesia, the skin overlying the tumor was incised. After opening the fascia over the temporal muscle, the tumor was visible as a gray-white mass protruding through the fascia. The pathological findings of an intraoperative frozen section were consistent with those of a benign myxomatous lesion. The tumor was encapsulated, lobulated, well defined, and encased within the sparse fibers of the temporal muscle. The muscle was extremely atrophic, probably because of the presence of a slow-growing tumor. The tumor was easily separated from the surrounding tissues with minimum blood loss and was completely removed. The tumor neither perforated the periosteum nor resulted in any invasion of the skull.

The resected tumor, which was soft, gelatinous, and lobulated, measured 6.5 × 4.0 × 3.0 cm and contained encapsulated nodules (Fig. 2A). There was no gross involvement of muscular fibers within the tumor. A diagnosis of intramuscular myxoma was confirmed on the basis of the histological examination (Fig. 2B), which revealed a monotonous, hypovascular, myxoid stroma with randomly scattered spindle and stellate cells along with striated muscle fibers. There was no nuclear atypism, necrosis, or hemorrhage.

The patient tolerated the procedure well and exhibited an uneventful postoperative course without any complications. There was no evidence of recurrence 6 months after resection.

Discussion

I have comprehensively described the radiological features and characteristic clinical course of a rare case of intramuscular myxoma in the temporal muscle. The reported patients with intramuscular myxoma of the temporal muscle, including the present case, are summarized in Table 1. The previously reported two cases were 5-cm and 3.5-cm in tumor size respectively.[10,11] The present case is the largest intramuscular myxoma in this location that has been reported to date. All reported cases were slow-growing

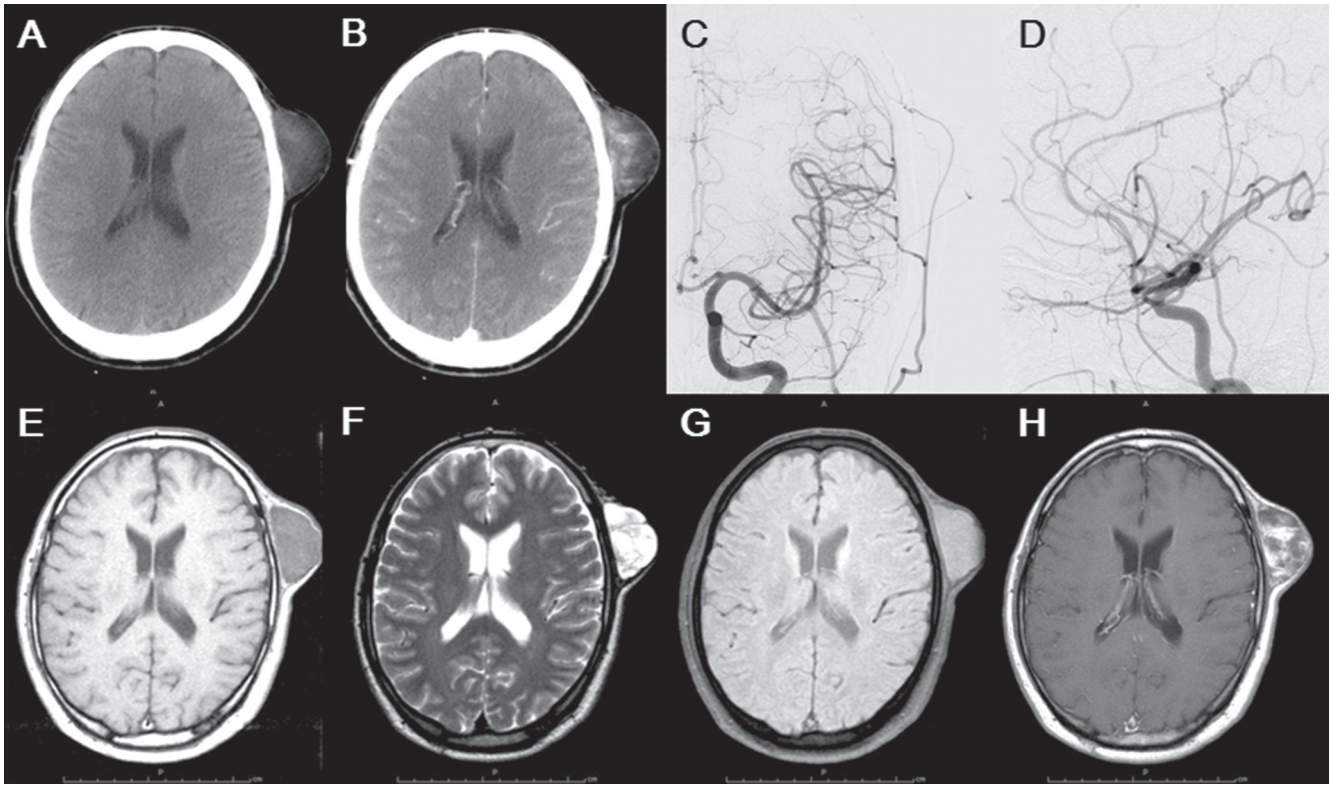

Fig. 1 Axial plain (A) and contrast-enhanced (B) computed tomography images showing an extracranial mass in the left temporal region. No tumor stain was observed on anteroposterior (C) and lateral (D) angiography with contrast injection from left common carotid artery. Axial T_1-weighted (E), T_2-weighted (F), fat-suppression (G), and contrast-enhanced (H) magnetic resonance images showing an extracranial mass in the left temporal region.

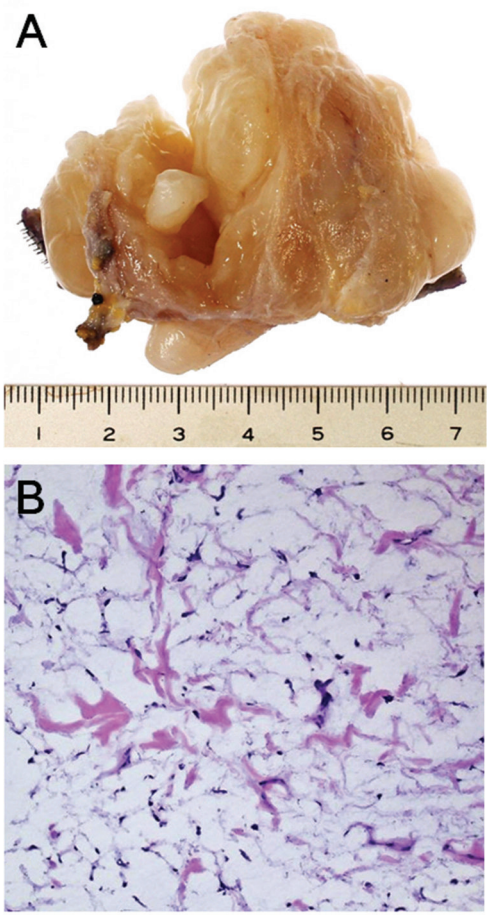

Fig. 2 A: Gross appearance of a surgical specimen. B: Photomicrograph of a surgical specimen showing bland spindle and stellate cells separated by extracellular myxoid matrix. Hematoxylin and eosin stain, original magnification × 100.

and asymptomatic, and did not recur after complete excision. None of them exhibited any signs of Mazabraud's syndrome such as bony deformities. On CT, the previously reported cases revealed well-defined, homogeneous, low-attenuating, and non-enhanced masses; however, the present case displayed heterogeneous enhancement. Neither of the previously reported cases included MRI or angiographic analysis. Because of inconclusive CT findings, preoperative biopsies were performed in the previously reported cases to facilitate appropriate surgical planning. However, in the present case, intraoperative frozen section analysis followed by total tumor excision was deemed feasible because sufficient preoperative clinical and radiological evaluation suggested that the tumor was poorly vascularized and exclusively localized in the extracranial area without invasion of surrounding structures.

There are some radiological characteristics of intramuscular myxoma described in literatures. On MRI, intramuscular myxoma is typically a well-defined mass with homogeneous hypointensity on T_1-weighted images and markedly hyperintensity on T_2-weighted or fat-suppression (STIR) images. Most lesions have focal areas of hypercellularity that are visible as areas of heterogeneous internal enhancement with gadolinium, which might cause further confusion with malignant tumors.[5,8,12,13] In addition, a fat rim separating the tumor from adjacent muscle might be evident on MRI. Angiographic examination reveals a poorly vascularized soft tissue mass.[8,14] These radiological features of intramuscular myxoma are consistent with the findings in the present case, with the exception that no clear fat rim was observed, possibly because the tumor was much larger than the atrophied temporal muscle.

Differentiating between intramuscular myxoma and soft

Table 1 Reported cases of intramuscular myxoma of the temporal muscle

Case no.	Author (yr)	Age (yrs)/sex	Maximum diameter of tumor	Duration of tumor growth	Mazabraud's syndrome	CT findings	MRI findings	Angiography findings	Timing of biopsy/findings	Surgery/ outcomes
1.	Serrat et al. (1998)[11]	62/M	5 cm	4 yrs	No	Low enhance (–)	N/A	N/A	Preoperative/ Stellate shaped cells in mucoid background without malignancy	Complete resection/No recurrence at 9-month follow-up
2.	Robin et al. (2004)[10]	46/F	3.5 cm	> 1 yr	No	Low enhance (–)	N/A	N/A	Preoperative/ Mesenchymal cells without malignancy or mucoid background	Complete resection/No recurrence at 18-month follow-up
3.	Present case	51/M	6.5 cm	3 yrs	No	Low enhance (+)	T_1WI low T_2WI high Enhance (+)	Vascularity (–)	Intraoperative/ Benign myxomatous lesion	Complete resection/No recurrence at 6-month follow-up

CT: computed tomography, F: female, M: male, MRI: magnetic resonance imaging, N/A: not applicable.

tissue sarcomas is rather clinically important for treatment planning.[2,9] Among various types of soft tissue sarcoma, myxoid sarcomas are particularly challenging to differentiate from intramuscular myxoma because both are strikingly similar in radiological and histological appearance.[4,8,12,14] Nevertheless, soft tissue sarcomas are generally highly proliferative and vascularized, and frequently contain focal areas of specific elements such as fat, chondroid matrix, hemorrhage, and necrosis, whereas these are absent in intramuscular myxoma.[8,12] Therefore, soft tissue sarcomas tend to grow more rapidly and reveal heterogeneous variable signal intensity, ill-defined margins along with evidence of invasion of adjacent structures, and intense diffuse enhancement on MRI images.[5] Furthermore, rich vascular network on angiography is the most conclusive difference from intramuscular myxoma.[14]

Differential diagnosis is further complicated in case of intramuscular myxoma with unusual location and uncharacteristically large size, as was the present case. Although clinical findings and extensive radiological evaluation are very helpful in preoperative assessment, definitive diagnosis is only possible with histological assessment.[8] Because intramuscular myxoma has benign prognosis, the goal of surgery should be complete tumor removal, and any recurrence implies insufficient resection or inaccurate diagnosis.[13] Intramuscular myxoma should be included in the differential diagnosis of soft tissue tumors of the head. The thorough analysis presented here should raise awareness of this rare lesion. I emphasize the role of surgeons, radiologists, and pathologists in careful differential diagnosis, in order to avoid an inappropriate treatment.

Conflicts of Interest Disclosure

The author has no personal, financial, or institutional interest in any of the drugs, materials, or devices in the article. The author, who is a member of The Japan Neurosurgical Society (JNS), has registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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Address reprint requests to: Tetsuhiro Higashida, MD, PhD, Department of Neurosurgery, Odawara Municipal Hospital, 46 Kuno, Odawara, Kanagawa 250-8558, Japan.
e-mail: tets018@gmail.com