Diagn Interv Radiol 2014; 20:310-315

MRI characterization of inflammatory myofibroblastic tumors in the maxillofacial region

Hui Li*, De-Ling Wang*, Xue-Wen Liu, Zhi-Jun Geng, Chuan-Miao Xie

PURPOSE
We aimed to investigate the magnetic resonance imaging (MRI) appearance of inflammatory myofibroblastic tumors (IMTs) in the maxillofacial region in order to improve diagnostic quality and resection efficacy.

MATERIALS AND METHODS
Ten cases of pathologically identified IMTs were analyzed by MRI. The MRI features were examined, including tumor location, tumor shape, tumor margins, and involvement of the surrounding tissues.

RESULTS
Of ten masses investigated in this study, eight masses were irregular neoplasms with unclear margins and two masses, in the parotid gland, were regular neoplasms with clear margins. Precontrast T1-weighted images of all ten masses exhibited isointense signals compared to the adjacent tissue, while contrast-enhanced T1-weighted images showed strong enhancement. Six masses were hypointense and four masses were slightly hyperintense in T2-weighted images. Involvement of the adjacent structures was observed in eight of ten cases. Meanwhile, two patients experienced intracranial involvement.

CONCLUSION
IMTs are rare tumors in the maxillofacial region, displaying a number of distinct MRI characteristics. Most importantly, they display low T2 signal intensity and strong enhancement, and they frequently invade surrounding structures. Thus, MRI can improve the accuracy of IMT diagnoses and provide critical information for surgical planning.

From the State Key Laboratory of Oncology (H.L., D.L.W., X.W.L., Z.J.G., C.M.X. ✉ *xiechm0792@126.com*), Guangzhou, China; the Medical Imaging and Minimally Invasive Interventional Center (H.L., D.L.W., X.W.L., Z.J.G., C.M.X.), Cancer Center, Sun Yat-sen University, Guangzhou, China.

*These authors contributed equally to this work.

Received 28 August 2013; revision requested 6 November 2013; revision received 30 November 2013; accepted 5 December 2013.

Published online 29 April 2014.
DOI 10.5152/dir.2014.13348

Inflammatory myofibroblastic tumors (IMTs) are rare neoplasms composed of myofibroblasts accompanied by prominent small lymphocytes and plasma cells (1). They have previously been categorized in the inflammatory pseudotumor group, but they are now defined as a separate entity based on electron microscope and immunohistochemical findings. Recent studies have shown that cytogenetic clonal abnormalities and anaplastic lymphoma kinase expression are the best definitive markers for diagnosing IMTs (2, 3). IMTs most commonly involve the lung, although they have been described in almost all sites in the body, in both sexes, and at all ages (4). IMTs in the maxillofacial region are exceptionally rare, and the clinical presentation depends on the location of the tumor. Patients can present with fever, pain, swelling, otorrhea, and cranial nerve palsy (5). IMTs clinically mimic malignant lesions, and the clinical significance lies in the difficulty encountered in excluding malignancy, preoperatively. There have been only a few studies that reported substantial numbers of IMTs in the maxillofacial region. Recently, Yuan et al. (6) reported imaging findings for eight IMT cases in the maxillary sinus; however, only three of the patients underwent MRI analysis, and the researchers could not summarize the MRI characteristics. Therefore, we have retrospectively analyzed the MRI findings of ten cases with maxillofacial IMT and summarized the imaging features in order to improve the diagnosis of this tumor.

Material and methods

Patients

A total of ten patients with pathologically confirmed IMTs in the maxillofacial region were reviewed retrospectively. These patients were treated in our hospital between February 2007 and June 2013. The clinical records of the patients were obtained for review, including age, sex, clinical symptoms, laboratory examinations, treatments, and follow-up information. This review was conducted according to the requirements of our institutional review board.

Imaging protocol

MRI examinations were performed using a 1.5 Tesla unit (GE Signa EXCITEII 1.5; GE Healthcare, Milwaukee, Wisconsin, USA) with a combined head and neck coil. The following sequences were obtained for each patient: unenhanced T1-weighted images; fast spin-echo imaging in the axial, coronal, and sagittal planes; unenhanced T2-weighted images in the axial plane; contrast-enhanced T1-weighted images in the axial and sagittal planes; and contrast-enhanced, fat-suppressed T1-weighted images in the coronal plane. A bolus injection of gadopentate dimeglu-

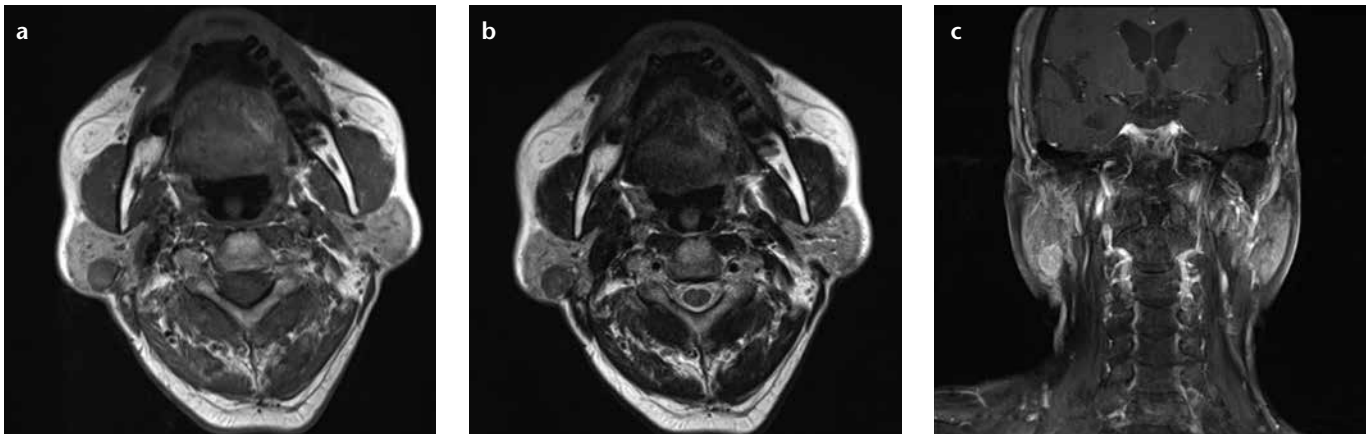

Figure 1. a–c. Inflammatory myofibroblastic tumor (IMT) in the right parotid gland viewed as a regular neoplasm with clear margins. The mass has a homogeneous isointense signal on precontrast T1-weighted image **(a)**, and a hypointense signal on T2-weighted image **(b)**. Postcontrast T1-weighted image **(c)** shows strong enhancement of the mass.

mine (0.1 mmol/kg body weight) was administered for contrast-enhanced sequences. Four patients underwent diffusion-weighted imaging (DWI) before intravenous injection of gadopentate dimeglumine. Diffusion-weighted images were obtained using a spin-echo echo-planar imaging sequence with a chemical-shift selective fat-suppression technique. The sequence was repeated for two motion-probing gradient values (b=0 and 1000s/mm^2).

Image assessment

Two experienced radiologists (H.I. and X.W.L, with seven and eight years of experience in the head and neck tumors, respectively) independently evaluated the magnetic resonance (MR) images. Any disagreements were resolved by consensus. All MR images were viewed on a picture archiving and communication system workstation monitor (Centricity RA1000 Workstation V.3.0; GE Healthcare).

The MR features of the primary tumors included: tumor location, tumor shape (round or lobulated), tumor margins (clear or unclear), signal intensity (classified as hypointense, isointense, or hyperintense compared to adjacent muscles), lesion texture (homogeneous or heterogeneous), and contrast-enhancement patterns (homogeneous or heterogeneous; strong, moderate, or absent). Contrast enhancement was considered to be strong if the degree of enhancement was similar to that of the normal mucosa, moderate if the degree of enhancement was less than that of the normal mucosa, and absent if the lesion exhibited no relative increase in signal. Tumor invasion of regional structures and regional lymphadenopathy were also evaluated.

Results

Of ten patients, six were male and four were female; ages ranged from nine to 70 years, with a median age of 38 years. Four patients were less than 25 years old. On physical examination, seven patients presented with a painless mass in the parotid gland or buccal region, two patients presented with edema in the maxillofacial region, and one patient presented with exophthalmos.

Seven patients underwent complete excision or partial resection of their masses, while three patients underwent extensive excision. None of the patients received chemotherapy, radiotherapy or corticosteroids/nonsteroidal anti-inflammatory drugs. One patient had a recurrence of IMT seven months after surgery. Residual tumors of the three patients, who underwent partial resection, were stable (n=2) or slightly enlarged (n=1) at the last follow-up. All patients were alive at the time of this investigation, and the follow-up time ranged from five to 66 months. No distant metastases were found.

Of ten masses investigated in this study, six arose in the masticator space, two in the parotid gland, one in the maxillary sinus, and one in the orbit. Six masses were on the right side of the maxillofacial region and four were on the left side.

The smallest mass in our study was 12×10 mm, while the largest mass was 60×67 mm. Eight masses were lobulated neoplasms with unclear margins, and two masses in the parotid gland, were round neoplasms with clear margins (Fig. 1).

Eight tumors had heterogeneous texture with isointense signals on precontrast T1-weighted image, slightly hypointense (n=4) or slightly hyperintense (n=4) heterogeneous signals on T2-weighted image, and strong heterogeneous signals on contrast-enhanced T1-weighted image. Two tumors had homogeneous texture with isointense signals on precontrast T1-weighted image and slightly hypointense signals on T2-weighted image, and exhibited strong enhancement on contrast-enhanced T1-weighted image (Fig. 2). No evidence of cystic or necrotic change was found in any of the masses. Four patients underwent DWI, where masses were seen as slightly hyperintense (n=3) and slightly hypointense (n=1) (Figs. 2, 3).

Eight patients had encroachment of structures adjacent to the masses, which included: muscles of the maxillofacial region (n=8), bone (n=6), pterygopalatine fossa (n=2), sphenoidal sinus (n=1), and optic nerve (n=1). Meanwhile, two patients had intracranial involvement via invasion through both the foramen rotundum and foramen ovale resulting in thickened cranial nerves (Fig. 4). One patient had

segmental dural thickening (Fig. 5). Clinical and MRI characteristics of the IMTs are summarized in the Table.

Discussion

The maxillofacial region is host to both benign and malignant tumors characterized by the proliferation of spindle-shaped cells. In this region, IMTs are very rare and their characteristics are controversial. IMT was first observed in the lungs by Brunn in 1939 and named by Umiker in 1954, according to its clinical and radiological behavior, which mimics a malignant process (7). Pathologically, IMTs are composed of lymphocytes, plasma cells, histiocytes, fibroblasts, and myofibroblasts, in variable proportions (8). As a result, IMTs are difficult to diagnose preoperatively through either fine-needle aspiration or biopsy. The lungs, liver, and gastrointestinal tract are the most common sites for IMTs. In the maxillofacial region, IMTs have been reported in the orbits, parapharyngeal spaces, maxillary sinuses, submandibular region, and the oral cavity (6, 9, 10).

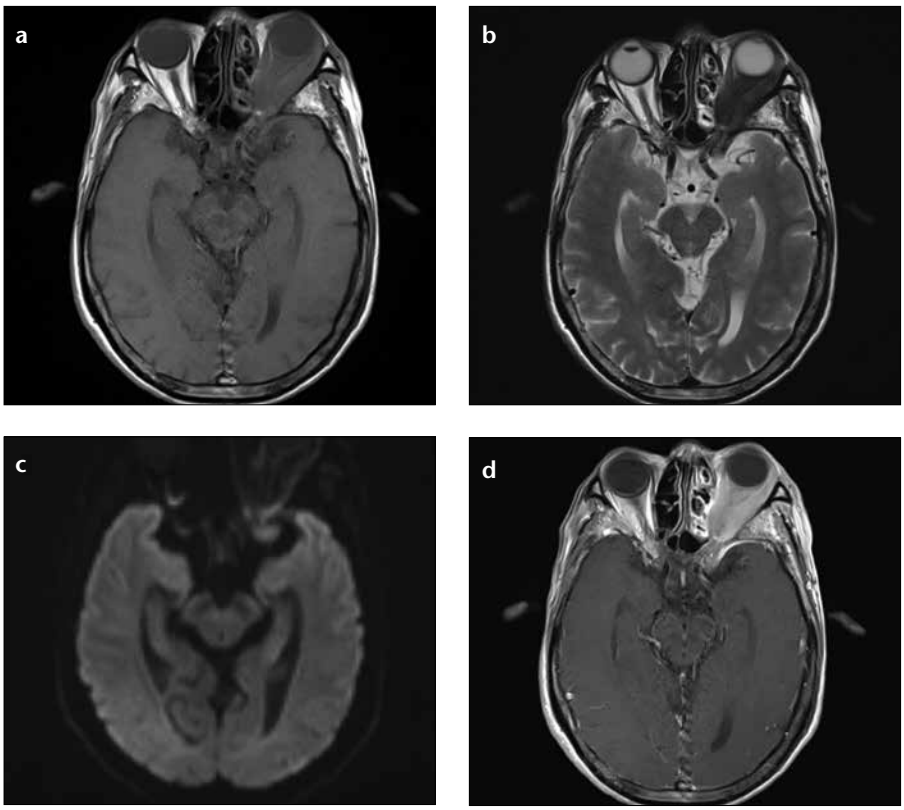

Figure 2. a–d. IMT in the left orbit, viewed as an irregular neoplasm with unclear margins. The mass has an attenuated heterogeneous isointense signal on precontrast T1-weighted image **(a)**, and a hypointense signal on T2-weighted image **(b)**. Invasion of the extraocular muscles, optic nerve, and orbit wall can also be detected. Diffusion-weighed image **(c)** shows heterogeneous hypointensity, while contrast-enhanced T1-weighted image **(d)** shows strong enhancement of the mass.

Clinically, IMT in the maxillofacial region may present as an incidentally discovered painless and indurate mass, developing within a relatively short duration; although occasionally it may present with specific symptoms related to the site of origin (5, 11). The majority of IMTs are not associated with systemic signs and symptoms like fever, weight loss, or malaise. In our study, a painless mass in the parotid gland or buccal region and swelling in the maxillofacial region were the most obvious clinical features of IMTs, and this was consistent with other reports. IMTs in the maxillofacial region have been diagnosed in all age groups, but the affected patients tend to be children and young adults (1). In this study, out of the ten patients studied, four were less than 25 years old while the youngest patient was only nine years old, in accordance with previous reports.

The mainstay of IMT treatment is complete surgical excision (12). If com-

Table. Summary of the clinical and MRI features of maxillofacial IMT cases

No	Gender/age (years)	Location	Involvement of surrounding structures	Treatment
1	M/70	Masticator space	MP, LaP, M, LeP, TP, SB, sphenoidal sinus, orbit, intracranial	Extensive excision
2	F/40	Masticator space	MP, LaP, M	Partial resection
3	F/16	Maxillary sinus	MP, LaP, MS	Partial resection
4	M/53	Masticator space	MP, LaP, AB	Extensive excision
5	M/24	Masticator space	MP, LaP, MS, PF	Complete excision
6	F/35	Masticator space	MP, LaP, M, LeP, TP, SB, AB, PF, intracranial	Extensive excision
7	M/9	Parotid gland	None	Complete excision
8	F/45	Masticator space	MP, LaP, MS	Complete excision
9	M/22	Parotid gland	None	Complete excision
10	M/52	Orbit	EM, orbit, optic nerve	Partial resection

MP, medial pterygoid; LaP, lateral pterygoid; M, masseter; LeP, levator palati; TP, tensor veli palatine; SB, skull base bone; MS, wall of the maxilla sinus; AB, alveolar bone; PF, pterygopalatine fossa; EM, extraocular muscles.

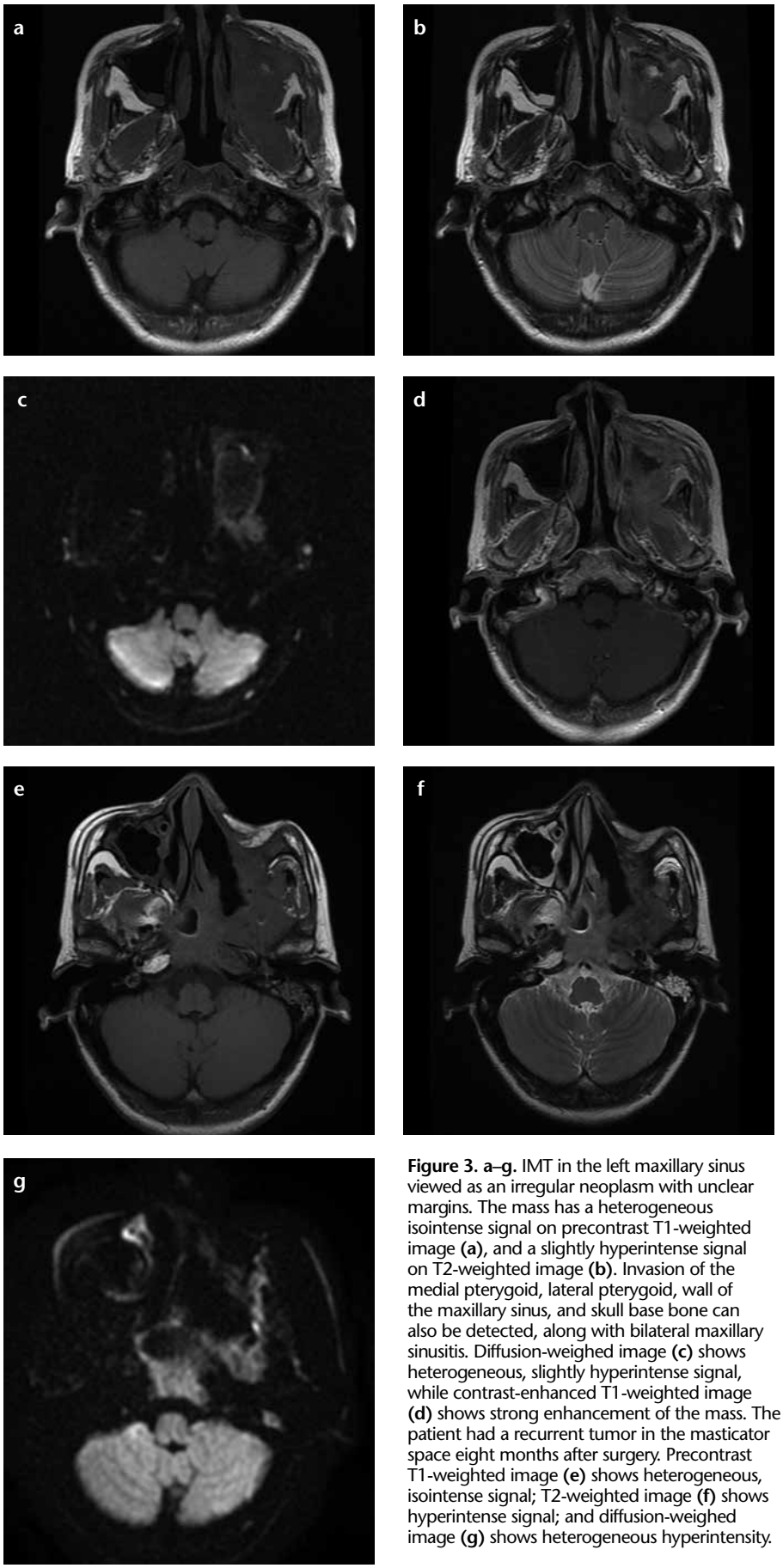

Figure 3. a–g. IMT in the left maxillary sinus viewed as an irregular neoplasm with unclear margins. The mass has a heterogeneous isointense signal on precontrast T1-weighted image **(a)**, and a slightly hyperintense signal on T2-weighted image **(b)**. Invasion of the medial pterygoid, lateral pterygoid, wall of the maxillary sinus, and skull base bone can also be detected, along with bilateral maxillary sinusitis. Diffusion-weighed image **(c)** shows heterogeneous, slightly hyperintense signal, while contrast-enhanced T1-weighted image **(d)** shows strong enhancement of the mass. The patient had a recurrent tumor in the masticator space eight months after surgery. Precontrast T1-weighted image **(e)** shows heterogeneous, isointense signal; T2-weighted image **(f)** shows hyperintense signal; and diffusion-weighed image **(g)** shows heterogeneous hyperintensity.

plete surgical excision is impossible or unsuccessful in case of aggressive IMTs, high-dose systemic corticosteroids, radiotherapy, or chemotherapy can be employed (13). In our study, seven patients underwent complete excision or partial resection of the masses and three patients underwent extensive excision. One patient had a recurrence, while no evidence of recurrence was observed in the other nine patients.

Because IMTs can clinically and radiographically mimic a malignant process, correctly recognizing the lesion is necessary not only for selecting the appropriate treatment, but also for guiding the surgical resection. Based on the MR findings of IMTs in the maxillofacial region in our study, we compiled the MR features of these tumors as follows. First, IMTs are solid masses with unclear margins and lobulated shapes. In our study, all masses were solid and no evidence of cystic or necrotic change was found. Despite their apparently benign morphological nature, IMTs have been reported to have locally aggressive growth behavior (10, 14). In our study, 80% (8/10) of the masses had aggressive, lobulated appearances, with unclear margins on MRI. Second, the tumors generally have hypointense or slightly hyperintense T2 signals, and they exhibit strong enhancement in the contrast-enhanced T1-weighted image. Unlike most tumors or tumor-like diseases, IMTs were reported to have hypointense T2 signals, which may be explained by the relative lack of mobile protons within fibrotic IMT lesions (15). In our study, T2 signal was hypointense or slightly hyperintense compared to adjacent muscles in 80% (8/10) of the masses, and this was consistent with previous studies. Moreover, IMTs were described as hypervascular tumors (7), and all the masses in our study exhibited strong heterogeneous enhancement after contrast administration, consistent with hypervascularization. Third, tumors commonly exhibit local invasion. IMTs have infiltrative features recognizable on MRI, mimicking more common aggressive malignant neoplasms (16). Yuan et al. (6), reported eight cases of IMTs of the maxillary sinus, with all cases exhibiting bone destruction or infiltration into adjacent tissues.

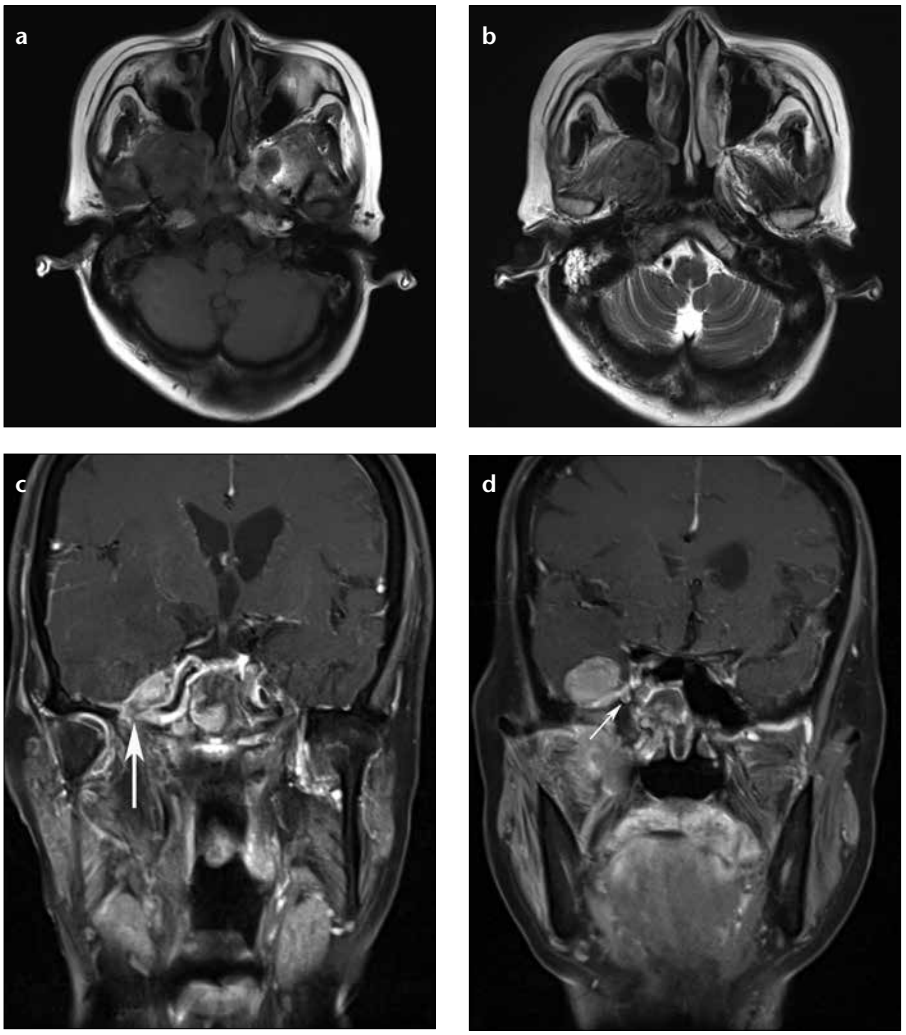

Figure 4. a–d. IMT in the right masticator space viewed as an irregular neoplasm with unclear margins. The mass has a heterogeneous isointense signal on precontrast T1-weighted image **(a)**, and a slightly hyperintense signal on T2-weighted image **(b)**. Right mastoiditis can also be seen. The same mass exhibits strong enhancement on contrast-enhanced T1-weighted image **(c, d)**. There is intracranial involvement via invasion through both the foramen ovale **(c,** *thick arrow)* and the foramen rotundum **(d,** *thin arrow)*. Invasion of the medial pterygoid, lateral pterygoid, levator palati, tensor veli palatini, skull base bone, and sphenoidal sinus can also be seen **(a–d)**.

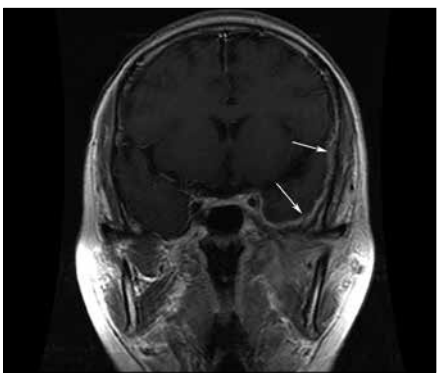

Figure 5. IMT in the left masticator space viewed an irregular neoplasm with unclear margins. Contrast-enhanced T1-weighted image shows strong enhancement and dural thickening *(arrows)*. Invasion of the medial pterygoid and the lateral pterygoid can also be seen.

In our study, eight patients showed involvement of adjacent structures, which included muscles of the maxillofacial region, bone, pterygopalatine fossa, paranasal sinus, and optic nerve. Notably, two patients in this study had intracranial involvement by means of invasion through the foramen rotundum and the foramen ovale. However, clinical nerve palsy was not found in these patients. Tumor invasion has also been reported by McKinney et al. (17). They hypothesized that this kind of invasion may be related to inflammatory perineural edema or ischemic neuropathy. Finally, no distant metastases are found in the IMTs. Although the clinical data and imaging features of IMTs may mimic malignancy, there is no evidence of malignant transformations, metastases, or deaths. Consistent with previous studies, no patients had lymphogenous or hematogenous metastases in this study.

Radiologically, IMTs in the maxillofacial region need to be differentiated from a variety of malignant tumors, such as rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma, lymphoma, and adenoid cystic carcinoma. Rhabdomyosarcomas are some of the most frequently occurring soft tissue sarcomas, and they are most common in children. Rhabdomyosarcoma masses are often iso- or hyperintense in T2-weighted images. Leiomyosarcomas are typically present in older patients, mostly on the skin and in the soft tissue of the head and neck (18, 19). As IMTs tend to be in children and young adults, and are usually hypointense or slightly hyperintense on T2-weighted image, they are easy to differentiate from sarcomas. However, MRI findings and the clinical presentation of fibrosarcoma in the maxillofacial region are often difficult to distinguish from IMT (20). Low T2 signal of fibrous tissue can be found in both types of tumors. There were two patients in this study that had intracranial involvement via invasion through the foramen rotundum and foramen ovale. This kind of invasion may be characteristic in IMTs and can be used to differentiate IMTs from fibrosarcomas. Lymphomas exhibit homogeneous soft-tissue signals with limited hemorrhage, calcification or necrosis, and the masses have mild to moderate homogeneous enhancement after contrast administration. Diffuse and bilateral cervical lymphadenopathy is also commonly found. IMTs can be differentiated from lymphomas by the heterogeneous lesion texture and the absence of lymphadenopathy.

The parotid gland IMTs in this study needed to be differentiated from pleomorphic adenoma, Warthin tumor, and carcinoma of the parotid gland. The high T2 signal of pleomorphic adenomas and their strong enhancement after contrast administration are well-known specific MRI findings. It is believed that the T2 hypointensity of a parotid tumor and postcontrast ill-de-

fined margins are useful indicators for malignancy. However, the two IMTs in the parotid gland in this study were round neoplasms with clear margins, low T2 signal and strong enhancement; these features maybe helpful in differential diagnosis. For Warthin tumors, the typical findings include the heterogeneity of the tumor, moderate T1 and T2 signal, absence of strong enhancement, and a presence in the parotid tail. IMTs may be differentiated from Warthin tumor by their strong enhancement.

A relatively limited number of patients was one of the main limitations of this paper. This limitation was due to the low incidence of IMTs in the maxillofacial region. A follow-up study with increased number of cases would be beneficial for a more detailed MRI characterization of maxillofacial IMTs. Moreover, several articles have highlighted that dynamic enhanced-MRI and DWI with ADC evaluation could improve the performance of MRI in distinguishing between benign and malignant head and neck tumors, and characterize the different histological types of different tumors. These novel imaging techniques may also be useful in differential diagnosis of IMTs and other tumors. In this study, only four patients underwent DWI, and they presented as slightly hyperintense (n=3) and slightly hypointense (n=1). If confirmed in larger series, this slight signal may also be a distinguishing feature for IMTs, as malignant tumors are supposed to be hyperintense on DWI.

In conclusion, on MRI analysis IMTs tend to present as solid masses with irregular margins, lobulated shapes, low T2 signal, strong enhancement and frequent invasion of surrounding structures. MRI analysis can help distinguish IMTs from malignant tumors, determine the extent of the lesion and its relationship with adjacent tissues. Thus, MRI can be a valuable tool for differential diagnosis of IMTs, as well as providing guidance in treatment and surgical resection.

Conflict of interest disclosure

The authors declared no conflicts of interest.

References

1. Devaney KO, Lafeir DJ, Triantafyllou A, et al. Inflammatory myofibroblastic tumors of the head and neck: evaluation of clinicopathologic and prognostic features. Eur Arch Otorhinolaryngol 2012; 269:2461–2465. [CrossRef]
2. Park SB, Cho KS, Kim JK, et al. Inflammatory pseudotumor (myoblastic tumor) of the genitourinary tract. AJR Am J Roentgenol 2008; 191:1255–1262. [CrossRef]
3. Park SB, Lee JH, Weon YC. Imaging findings of head and neck inflammatory pseudotumor. AJR Am J Roentgenol 2009; 193:1180–1186. [CrossRef]
4. Gallego L, Santamarta TR, Blanco V, Garcia-Consuegra L, Cutilli T, Junquera L. Inflammatory myofibroblastic tumor of the lung and the maxillary region: a benign lesion with aggressive behavior. Case Rep Dent 2013; 2013:879792.
5. Maire JP, Eimer S, San GF, et al. Inflammatory myofibroblastic tumour of the skull base. Case Rep Otolaryngol 2013; 2013: 103646.
6. Yuan XP, Li CX, Cao Y, Singh S, Zhong R. Inflammatory myofibroblastic tumour of the maxillary sinus: CT and MRI findings. Clin Radiol 2012; 67:e53–57. [CrossRef]
7. Kim SJ, Kim WS, Cheon JE, et al. Inflammatory myofibroblastic tumors of the abdomen as mimickers of malignancy: imaging features in nine children. AJR Am J Roentgenol 2009; 193:1419–1424. [CrossRef]
8. Palaskar S, Koshti S, Maralingannavar M, Bartake A. Inflammatory myofibroblastic tumor. Contemp Clin Dent 2011; 2:274–277. [CrossRef]
9. Amin M, Ali R, Kennedy S, Timon C. Inflammatory myofibroblastic tumor of the nose and paranasal sinuses masquerading as a malignancy. Ear Nose Throat J 2012; 91:E1–3.
10. Lourenco SV, Boggio P, Simonsen NM. Inflammatory myofibroblastic tumor of the tongue: Report of an unusual case in a teenage patient. Dermatol Online J 2012; 18:6.
11. Chen YF, Zhang WD, Wu MW, Ou-Yang D, Zhang Q. Inflammatory myofibroblastic tumor of the head and neck. Med Oncol 2011; 28 (Suppl 1):S349–353. [CrossRef]
12. Thistlethwaite PA, Renner J, Duhamel D, et al. Surgical management of endobronchial inflammatory myofibroblastic tumors. Ann Thorac Surg 2011; 91:367–372. [CrossRef]
13. Diop B, Konate I, Ka S, et al. Mesenteric myofibroblastic tumor: NSAID therapy after incomplete resection. J Visc Surg 2011; 148:e311–314. [CrossRef]
14. Luo YZ, Dai LB, Zhou SH, Luo XM, Fan J, Ruan LX. Etiology analysis and computed tomography imaging of a tonsillar inflammatory myofibroblastic tumor: report of an immunocompetent patient and brief review. Head Neck Onco 2012; 4:4. [CrossRef]
15. Masciocchi C, Lanni G, Conti L, et al. Soft-tissue inflammatory myofibroblastic tumors (IMTs) of the limbs: potential and limits of diagnostic imaging. Skeletal Radiol 2012; 41:643–649. [CrossRef]
16. Singhal M, Ramanathan S, Das A, Singh G, Bagga R, Khandelwal N. Omental inflammatory myofibroblastic tumour mimicking peritoneal carcinomatosis. Cancer Imaging 2011; 11:19–22.
17. McKinney AM, Short J, Lucato L, Santa-Cruz K, McKinney Z, Kim Y. Inflammatory myofibroblastic tumor of the orbit with associated enhancement of the meninges and multiple cranial nerves. AJNR Am J Neuroradiol 2006; 27:2217–2220.
18. Eppsteiner RW, DeYoung BR, Milhem MM, Pagedar NA. Leiomyosarcoma of the head and neck: a population-based analysis. Arch Otolaryngol Head Neck Surg 2011;137:921–924. [CrossRef]
19. Tanaka H, Westesson PL, Wilbur DC. Leiomyosarcoma of the maxillary sinus: CT and MRI findings. Br J Radiol 1998; 71:221–224.
20. Dalley RW. Fibrous histiocytoma and fibrous tissue tumors of the orbit. Radiol Clin North Am 1999; 37:185–194. [CrossRef]