RESEARCH ARTICLE

MRI findings of solitary fibrous tumours in the head and neck region

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Objectives: To analyse the MRI findings of solitary fibrous tumours in the head and neck region.

Methods: We retrospectively reviewed MR images in eight patients with solitary fibrous tumours proven on histological examination. All the patients underwent conventional MRI, and four patients also underwent dynamic contrast-enhanced MRI and diffusion-weighted imaging in five cases. Image characteristics were analysed.

Results: All lesions were found as solitary well-defined masses ranging in size from 1.9 to 6.8 cm (mean, 4.1 cm). They were mostly homogeneous and isointense to the muscle on T_1 weighted images and heterogeneous and mildly hyperintense on T_2 weighted images. After gadolinium administration, areas that were mildly hyperintense on T_2 weighted images were strongly enhanced. They were mildly hyperintense on diffusion-weighted imaging. The average tumour-apparent diffusion coefficient values were $0.001\,157 \pm 0.000\,304\,9\,\text{mm s}^{-2}$ compared with the muscle $0.000\,760 \pm 0.000\,265\,0\,\text{mm s}^{-2}$, and there was a statistical difference of $p = 0.002$. The time–intensity curves exhibited a rapidly enhancing and a slow washout pattern on dynamic contrast-enhanced MRI.

Conclusions: Solitary fibrous tumours should be considered in cases of heterogeneous hypervascular tumours in the head and neck region. Areas of mild hyperintense intensity on T_2 weighted images that are strongly enhanced after gadolinium injection are suggestive of this diagnosis. Non-restricted diffusion and rapidly enhancing and slow washout pattern time–intensity curves may be additional valuable features.

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Introduction

Solitary fibrous tumours (SFT), first reported in the pleura by Klemperer and Rabin¹ in 1931, are rare spindle-cell neoplasms originating from mesenchymal tissue. SFT are classified as having intermediate biological potential with a low risk of metastasis under the 2002 WHO Classification.² Over 50% of these tumours

are located in the thoracic cavity, but extrathoracic tumours have been reported in many sites, such as the abdomen, trunk and extremities.^{3,4} Although rare, SFT could also be found in various head and neck regions, such as the cheek, orbit, oral cavity, laryngeal and parapharyngeal spaces as well as the thyroid gland,^{4–7} and the common sites affected in the head and neck region are the orbit and oral cavity.^{7,8}

Classically, SFT are composed of variably pleomorphic spindle cells admixed with collagen and arranged haphazardly. The diagnosis is strongly dependent on the microscopic appearance and characteristic immunohistochemical staining for CD34 and B-cell lymphoma protein 2 (Bcl-2).

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Despite their characteristic histological and immunohistochemical features, SFT of the head and neck remain a diagnostic challenge to both clinicians and radiologists because they are often poorly recognized and confused with other more common neoplasms, such as schwannoma, capillary haemangioma, cavernous haemangioma (venous malformation), varix, haemangiopericytoma, giant-cell angiofibroma and benign fibrous histiocytoma. A review of the literature reveals sporadic case reports on SFT in the head and neck region. To our knowledge, however, there is rather only limited descriptions of the radiologic features of SFT in the literature. During the past 8 years, we have encountered eight cases of SFT in the head and neck region. Here, we conducted a retrospective review of the MRI features of SFT and correlated them with the histopathological features.

Methods and materials

From June 2004 to June 2012, eight patients with SFT, proven by pathological examination, occurring in the head and neck region were reviewed. There were 6 males and 2 females, ranging in age from 22 to 68 years, with a mean age of 42 years. All eight patients underwent MRI. We retrospectively reviewed the findings of MR images obtained in these patients.

All MR images were obtained at a 1.5-T MR unit (Signa; GE Healthcare, Milwaukee, WI) with a neurovascular array coil. Imaging protocols contained at least unenhanced axial T_1 weighted spin-echo sequences, axial T_2 weighted fast spin-echo sequences, coronal T_2 weighted fast spin-echo sequences with fat saturation and contrast-enhanced axial and coronal T_1 weighted sequences. An intravenous dose of $0.1\text{--}0.2\text{ mmol kg}^{-1}$ of contrast agent (gadolinium-diethylenetriaminepentaacetic acid, Magnevist®; Bayer Schering Pharma AG, Berlin, Germany) had been administered to the patients undergoing contrast-enhanced MR scanning. Images were obtained with fields of view of 20–24 cm, three acquisitions, matrixes of 256×256 , section thicknesses of 5 mm and spaces of 1 mm.

Additionally, diffusion-weighted imaging (DWI) was acquired in five patients (single-shot gradient-echo echo-planar pulse sequence). The DWI parameters were as follows: repetition time/echo time, 2500–3000/70 ms for a b factor of 0 and 1000 s mm^{-2} ; thickness/space, 5/0.5 mm; matrix, 128×128 ; fields of view, 24 cm; and 8 acquisitions. Dynamic contrast-enhanced (DCE) MRI (T_1 gradient-echo; 4.8/2.2 ms [repetition time/echo time], flip angle = 90° , i.v. injection of 0.2 mmol kg^{-1} gadolinium-diethylenetriaminepentaacetic acid, Magnevist; Bayer Schering Pharma AG) was performed in four patients. Images were analysed offline using Functool software (GE Medical Systems, Waukesha, MI) on the MR unit.

The MR images were reviewed by two dedicated head and neck radiologists in consensus. The MR images were evaluated by the location, size, shape, margin, internal

architecture and pattern and degree of enhancement of the lesion. The size of the lesion was measured at the greatest diameter. The shape was graded as ovoid or irregular, and the margin was graded as well defined or ill defined. As for the internal architecture, we compared the signal intensity of the lesion on T_1 and T_2 weighted images with that of the neck musculature. On contrast-enhanced T_1 weighted images, the pattern of enhancement of the lesion was categorized as homogeneous or heterogeneous and the degree of enhancement as mild, moderate and marked.

All patients underwent complete excision of the tumour at our institution. Diagnosis of SFT was confirmed by pathologists based on histopathological and immunohistochemical analyses. The histological techniques included routine haematoxylin and eosin staining and immunohistochemical evaluation. Immunohistochemical analysis included staining for CD34, vimentin, CD99, Bcl-2, S-100 protein, smooth muscle antibody and cytokeratin.

Results

The clinical and MRI features of eight patients with SFT in the head and neck region are summarized in Table 1.

Clinical data

The most frequently presenting symptoms of SFT in the head and neck region were indolent palpable mass (3/8: 37.5%) and proptosis (3/8: 37.5%) when occurring in the orbit. One patient presented with oropharynx bleeding and another with facial palsy. After the operation, all patients were in good health with no evidence of recurrent tumour during a follow-up of 1–96 months (mean, 32 months).

MRI findings

Based on imaging findings, three of the eight lesions arose from the orbit, two of the eight lesions from the masticator space, two of the eight lesions from the parapharyngeal space and one of the eight lesions from the parotid gland. Five of the eight lesions localized in the right side and three of the eight lesions in the left side of the head and neck. All eight lesions were found as solitary well-defined masses ranging in size from 1.9 to 6.8 cm (mean, 4.1 cm). They presented as irregular in shape in four of eight patients and ovoid in four of the eight patients. On the MR images, SFT in the head and neck region were mostly homogeneous and isointense or mildly hypointense in signal intensity to the muscle on the T_1 weighted images and heterogeneous and mildly hyperintense in signal intensity on the T_2 weighted images (Figure 1a,b). After gadolinium administration, all cases showed marked heterogeneous enhancement (Figure 1c), and hypointense linear septa were observed in two cases (Figure 2a–c). Erosion of the orbital floor was present in one case (Figure 2d).

Table 1 Summary of clinical and imaging findings in eight patients

Case	Age/ sex	Symptom	Location	Size (cm)	Margin/ shape	Preoperative diagnosis	T ₁ WI	T ₂ WI	MR enhancement	DWI	DCE
1	30/M	Proptosis	R; orbit	2.4	Well/ ovoid	SFT	Iso	Mild hyper	Hetero/ marked	/	/
2	27/M	Palpable mass	R; masticator space	6.2	Ill/ irregular	Haemangioma	Iso	Mild hyper	Hetero/ marked	Mild hyper	II
3	68/M	Facial palsy	L; masticator space	5.3	Ill/ irregular	Haemangioma	Mild hypo	Mild hyper	Hetero/ marked	Mild hyper	/
4	55/M	Palpable mass	R; parotid gland	1.9	Well/ ovoid	Adenolymphoma	Iso	Mild hyper	Hetero/ marked	/	/
5	23/F	Oropharynx bleeding	L; parapharyngeal space	3.7	Well/ ovoid	Pleomorphic adenoma	Mild hypo	Mild hyper	Hetero/ marked	Mild hyper	II
6	49/M	Palpable mass	L; parapharyngeal space	6.8	Well/ irregular	Schwannoma	Mild hypo	Mild hyper	Hetero/ marked	Mild hyper	II
7	22/M	Proptosis	R; orbit	2.6	Well/ ovoid	Haemangioma	Iso	Mild hyper	Hetero/ marked	Mild hyper	II
8	59/F	Proptosis	R; orbit	3.6	Well/ irregular	Haemangioma	Iso	Mild hyper	Hetero/ marked	/	/

II, the time-intensity curves exhibited a rapidly enhancing and slow washout pattern; DCE, dynamic contrast enhanced; DWI, diffusion-weighted imaging; F, female; hetero, heterogeneous; homo, homogeneous; hyper, hyperintense; hypo, hypointense; ill, ill-defined; iso, isointense; L, left; M, male; R, right; SFT, solitary fibrous tumours; T₁ WI, T₁ weighted image; T₂ W, T₂ weighted image; well, well-defined; /, undo. Size was denoted as the greatest diameter. Signal intensity of the tumour was compared with those of the neck musculature.

Five patients underwent DWI. A mild hyperintense wad was seen on DWI (Figure 1d). The average tumour apparent diffusion coefficient (ADC) values of five patients were $0.001\,157 \pm 0.000\,304\,9\,\text{mm s}^{-2}$ compared with the muscle $0.000\,760 \pm 0.000\,265\,0\,\text{mm s}^{-2}$, and there is a statistical difference with $p = 0.002$, and DCE-MRI was also performed in four patients. The time-intensity curves (TICs) of four patients exhibited a rapidly enhancing and slow washout pattern on DCE-MRI (Figure 1e).

Microscopy and immunohistochemistry

The microscopic and immunohistochemical findings are summarized in Table 2. Microscopic examination revealed benign SFT for seven patients and borderline tumour for one patient. Histological examination showed that the tumours consisted of spindle cells with collagenous tissues (Figure 2e). Immunohistochemical examination revealed that all the eight tumours were positive for CD34 (Figure 2f) and vimentin (Figure 2g); five were positive or partially positive for CD99; six were positive or partially positive for Bcl-2 and all of the tumours were negative for S-100, cytokeratin and smooth muscle antibody. The presence of rounded or linear low-intensity foci on both T₁ and T₂ weighted images was attributable to the collagen content, low cellularity and associated reduced proton mobility, and the SFT also are vascular tumours that are vigorously enhancing.

Discussion

SFT are rare spindle-cell neoplasms originating from mesenchymal tissue, which are classified as having intermediate biological potential (rarely metastasizing) under the 2002 WHO classification.^{1,2} Although mostly occurring in the pleura, this tumour has also been

recently reported in many sites, including abdomen, trunk, extremities and the head and neck region.³⁻⁸ In our study, eight SFT in the head and neck region were located in the orbit, masticator space, the parapharyngeal space and the parotid gland.

Microscopically, SFT are well-circumscribed non-encapsulated tumours, which show a patternless pattern with alternating hypocellular and hypercellular areas separated by thick bands of hyalinized collagen and branching haemangiopericytoma-like vessels.⁹ Immunohistochemistry is a significant contributor in distinguishing SFT from other soft-tissue tumours in the head and neck region. SFT strongly stain for CD34 and vimentin. Less commonly, they will express Bcl-2 and CD99, but they are usually negative for epithelial, vascular, neural and muscle markers.¹⁰ Our findings were generally consistent with previous results. Table 2 presents a summary of the immunohistochemistry results of our cases. In our study, SFT were immunoreactive for CD34, vimentin, partial CD99 and Bcl-2 but negative for S-100 protein, cytokeratin and smooth muscle antibody. Typically, there is little cellular atypia and no mitotic activity. Possible malignant variants are rare and distinguished by having 4 mitoses per 10 high-power microscopic fields. Out of the eight cases, we were able to identify only one case with atypical or malignant features.

Clinically, the symptoms and signs of SFT depend on the location of the tumour and the involved organs. The earlier presentation in the head and neck region is owing to the visibility of a mass or the local symptoms. Most patients present with asymptomatic mass, but some patients have various oral or facial dysfunctions such as bleeding and numbness. Patients with orbital SFT usually present with slowly progressive unilateral exophthalmos. Patients with SFT occurring in superficial soft tissues like the parotid gland often present with an asymptomatic palpable mass. SFT occur with equal frequency in both

Figure 1 Images of a 23-year-old woman presenting with left parapharyngeal mass (Case 5). (a) The T_1 weighted axial MR image shows a well-defined ovoid mass (arrow) that is isointense to the muscle with inner hypointense. (b) The T_2 weighted axial MR image shows the heterogeneous high-signal intensity of the mass (arrow). (c) The contrast-enhanced T_1 weighted axial MR image reveals the heterogeneously strong enhancement of the mass (arrow). (d) The diffusion-weighted axial MR image reveals mild hyperintense signal intensity in the mass (arrow). (e) The dynamic contrast-enhanced MRI reveals that the time-intensity curves exhibit a rapidly enhancing and slow washout pattern.

sexes, and they usually occur in adults, whose ages range from 30 to 64 years.¹¹ In our study, the age range was from 22 to 68 years, but the incidence tended to be higher in males than in females, with a sex ratio of 6:2. This demographic discrepancy seemed to be caused by a selection bias for the limited number of cases.

The diagnosis of SFT by clinical presentation alone is impossible. There are certain radiological appearances suggestive of the diagnosis.^{12,13} Regarding MRI features, the most characteristic feature within our series was that of a well-defined enhancing mass. The signal characteristics as compared with the surrounding neck musculature may also aid in diagnosis. In our series, SFT in the head and neck region were mostly homogeneous and isointense or mildly hypointense in signal intensity to the muscle on the T_1 weighted images and heterogeneous and mildly hyperintense in signal intensity

on the T_2 weighted images. After gadolinium administration, all cases showed marked heterogeneous enhancement with hypointense areas. Khandelwal et al¹⁴ reported that the intratumoural hypointense areas on T_1 weighted images correlated with myxoid or cystic degeneration. Kim et al¹⁵ reported that isointense to hypointense signal intensity on T_2 weighted images reflected fibrous tissue with high collagen content, and hyperintense signal intensities were related to internal haemorrhage, cystic degeneration or relatively fresh fibrosis. And Kim et al¹⁶ reported that the signal intensity of SFT on T_2 weighted MR images decreased as the collagen content increased on microscopic examination. We also believe that this variable signal intensity depends mainly on the differences in the main components of the tumour, namely the amount of cellular components, collagen and fibroblasts, and also

Figure 2 Images of a 59-year-old woman presenting with right orbital mass (Case 8). (a) The T_1 weighted axial MR image shows a well-defined irregular mass (star) that is isointense to the muscle with an inner hypointense linear septa (arrow). (b) The T_2 weighted axial MR image shows the heterogeneous hyperintense signal intensity in the mass (star) with the inner hypointense linear septa (arrow). (c) The contrast-enhanced T_1 weighted axial MR image reveals the heterogeneously strong enhancement of the mass (star). The linear septa were less enhanced than in the other portion of the tumour (arrow). (d) The contrast-enhanced T_1 weighted coronal MR image reveals erosion of the orbital floor (arrow). (e) Photomicrograph shows that the tumour is composed of a haphazard proliferation of spindle cells separated from the hyalinized collagen tissue (haematoxylin-eosin stained, magnification $\times 200$). (f) The inset shows that the tumour cells and the capillary endothelial cells have immunohistochemically positive CD34 results (original magnification $\times 200$). (g) Immunohistochemistry of the tumour cells showing positive staining for vimentin (original magnification $\times 200$).

on the presence of degeneration. Intense enhancement of SFT is generally attributed to high vascularity because of the prominent vascular channels within the tumour.¹⁷ We also observed the linear septa within the tumours in two cases. Those lines corresponded to the hypocellular collagenous stroma intervening in the hypercellular area. The most common radiographic osseous finding in SFT is regressive remodeling of adjacent bone due to the long-standing lesions. Erosion of the orbital floor was present in one case of our study, which was proven as borderline neoplasm by histochemical examination.

In addition to morphological MR data, we present in this work the results of multimodal MRI of SFT, including DWI and DCE-MRI. The DWI findings of SFT in the head and neck region have been rarely reported. In our cases, the lesions showed mild hyperintense signal intensity on DWI. Through the software, we achieved quantitative ADC values, and the ADC values of the tumours were much higher than the muscles. So, to some extent, the higher ADC values proved that this tumour might be benign. Non-restricted diffusion may be a feature that favours a pre-operative diagnosis of SFT in the head and neck region. On DCE-MRI, the TICs of

tumours exhibited a rapidly enhancing and slow washout pattern in our cases, which may be an additional clue to make the correct diagnosis. DCE-MRI findings of SFT in the head and neck region have been rarely reported. Only Kim *et al*¹⁵ also reported that the TICs of orbital SFT on dual-phase CT demonstrated rapid intense enhancement at early phase scanning, followed by significant washout of contrast material at late-phase scanning. The difference of TICs between MRI and CT might be owing to the difference of contrast-enhancement theory.

The radiological appearance of SFT in the head and neck region is not specific to a diagnosis of SFT. They are often misdiagnosed as other common tumours on initial images, such as cavernous haemangioma (venous malformation), schwannoma, pleomorphic adenoma and adenolymphoma in our cases. On the basis of the aforementioned imaging characteristics of SFT seen on MR images, schwannoma and cavernous haemangioma should be the main lesions included in differential diagnosis. Schwannomas are apparently derived from Schwann cells, which may arise from any myelinated nerve fibre. They are usually solitary, fusiform, smoothly margined and encapsulated lesions. Extracranial

Table 2 Summary of the histological features of orbital solitary fibrous tumours in the present series

Case	Age/sex	Benign/malignant	CD34	Vimentin	CD99	Bcl-2	S-100	SMA	CK
1	30/M	Benign	+	+	—	±	—	—	—
2	27/M	Benign	+	+	±	±	—	—	—
3	68/M	Benign	+	+	+	/	—	—	—
4	55/M	Benign	+	+	+	+	—	—	—
5	23/F	Benign	+	+	—	±	—	—	—
6	49/M	Benign	+	+	+	±	—	—	—
7	22/M	Benign	+	+	±	+	—	—	—
8	59/F	Borderline	+	+	—	—	—	—	—

+, positive; —, negative; ±, partial positivity; /, undo; Bcl-2, B-cell lymphoma protein 2; CK, cytokeratin; SMA, smooth muscle antibody.

schwannomas have a distinct tendency to originate in the head and neck region. On MRI scans, most tumours appeared hypointense or isointense relative to muscle on T_1 weighted images and variable hyperintense on T_2 weighted images, depending on the structure of the individual tumour.¹⁸ Unlike SFT, schwannomas have more cystic degeneration and the different enhancing characteristics showing lower enhancement degree. Cavernous haemangiomas are believed to be haemodynamically isolated and encapsulated venous malformations.¹⁸ They can be differentiated from SFT by more hyperintensity on T_2 weighted MR images with fat suppression, and the different enhancing characteristics showing patchy enhancement and delayed pooling of contrast material on dynamic studies.

Surgery is generally regarded as the treatment of choice for SFT. Although local excision is usually curative, SFT have been reported to recur and metastasize. The complete surgical excision appears to be the preferred treatment of SFT. The prognosis of SFT might be highly related to the situation of its initial treatment, and total resection is associated with longer progression-free survival.¹² Total resection is favoured

to prevent leaving microscopic nests of cells of SFT behind as a nidus for recurrence. In our series, all the patients experienced total resection of the tumours. None of our study patients experienced local recurrence or distant metastasis. After the operation, all patients made uneventful recoveries, and they are still clinically and radiologically free from tumour recurrence during the follow-up period.

In conclusion, SFT are uncommon neoplasms occurring in the head and neck region. According to our results, the diagnosis of SFT may be suggested in cases of well-defined smooth or irregular contoured mass, exhibiting mild hyperintense T_2 areas that are strongly enhanced after gadolinium administration. Non-restricted diffusion and rapidly enhancing and slow washout pattern TICs may be additional valuable features. Although these findings remain non-specific, SFT should be considered in cases of heterogeneous hypervascular tumours in the head and neck region.

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