



CT & MRI SCAN FINDINGS OF SOLITARY FIBROUS TUMOR ALIGN WITH THAT OF HISTOLOGICAL FINDINGS: AN EFFECTIVE NON INVASIVE DIAGNOSTIC TOOL

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ABSTRACT

Purpose: To analyze the computed tomography (CT) and magnetic resonance imaging (MRI) features of histologically proven soft tissue solitary fibrous tumors to gain validity of imaging findings on final diagnosis.

Materials and Methods: Twenty eight patients with confirmed SFTs by histopathology were retrospectively analyzed. 21 patients had undergone CT scan while 6 had undergone MRI and 1 patient had both CT and MRI scan. The Imaging characteristics before and after contrast enhancement were analyzed and correlated simultaneously with its patho-morphology. **Results:** Among 28 cases, 11 patients had no symptoms. These tumors were located mostly in the thoracic cavity (42%) and mediastinum (14%). The average size range was 8.2 X 6.8cm.² These tumors in CT were predominantly heterogeneous with an average CT value of solid component being 41.5 HU. These were heterogeneously enhanced by contrast in the arterial phase. Most of these demonstrated a positive vessel sign (14/18). These tumors in MR scan were Iso-intense on T1WI and heterogeneous or hyper/iso-intense on T2WI. 85% of these tumors showed heterogeneous enhancement.

Conclusion: SFTs usually are atypical tumors that are easily missed at diagnosis. The reliable diagnostic features in CT scan are - large well-defined hyper vascular tumor, variable degree of necrosis/cystic changes and enhanced serpentine vessels in arterial phase(CT) with flow void sign. Similarly, MR images that show homogenous iso- or hypo-intense signal on T1WI, and heterogeneous iso- or hypo-intense signal on T2WI with serpentine vessels enhanced in arterial phase and flow voids sign on T2WI are diagnostic of solitary fibrous tumors.

KEYWORDS: Solitary fibrous tumor, Soft tissue tumor, Computed tomography, Magnetic resonance imaging, histopathology.

1. INTRODUCTION

Tumors originating from mesenchymal tissues are rarely encountered in radiology clinics. Among them, Solitary fibrous tumors (SFTs) are the rarest form of spindle cells tumors which were firstly described by E. Wagener during the 1870's. These tumors can be present anywhere in the body where muscle tissue however the first case report was described in pleura by Klemperer and Rabin in 1931.^[1] With time, these SFT's were noticed at different locations like - head and neck regions such as cheek, orbit, oral cavity, laryngeal and para pharyngeal spaces^[2,3,4], retroperitoneum, abdomen and pelvis^[5] and

the soft tissues of the extremities.^[6] These tumors have been classified as intermediate malignancies that rarely metastasize by the World Health Organization in 2002.^[7] The tumor are typically composed of spindle cells surrounded by dense collagen stroma, which usually contain hyper- and hypo-cellular areas.^[Error! Bookmark not defined.] Moreover, studies on pleural samples too have demonstrated its benign nature with only approximately 10% recurring locally or metastasizing.^[8,9,10]

The common and gold standard for diagnosing SFT's so far, has been direct histological observation that exhibits variable pleomorphic spindle cells mixed with collagen arranged randomly and the immunohistochemical staining of tumor markers like- CD34, Vimentin and bcl-2 are always positive.^[11] However, these tumors are silent in nature, smaller in size, variably located and easily mistaken with other commonly occurring neoplasm such as - angiosarcoma, liposarcoma, adenolymphoma, schwannoma, hemangioma etc. With these varieties of differentials, patients with SFT's are initially subjected to imaging modalities before clinician advices for histological studies. Keeping this fact in mind, radiological scholars for the last decade have put in an enormous effort at diagnosing these SFT's by means of imaging techniques. Radiologists have been able to diagnose this tumor by means of CT that can accurately determine its size, morphology, location and topography while MRI, USG, PET have been helpful too.

Similarly, SFT being a rare disease, we find limited number of research articles and most of these center around case presentations and case discussions. We, too, are presenting the cases of SFT observed at our center. We have conducted a retrospective study of 28 cases of histologically proven SFT exploring the radiological features observed. By doing so, we validate the radiological diagnosis of these benign tumors based parallel with histology. This we expect would further reinforce the imaging modality for diagnosing SFT, a clinical need at times of early diagnosis and early institution of treatment, where time-clock ticks. We also feel that a larger trial is necessary to validate the findings of this study as it would address other issues, not addressed by a retrospective study.

MATERIALS AND METHODS

Source

The hospital database and medical records were explored, tabulated and tallied. There were 28 patients that had been histologically diagnosed with SFTs from 2012 to 2015. Different variables like the patient's age, sex, physical examination, symptoms, lesion's location, size, number were recorded. Meanwhile the radiological and histological findings were also noted and analyzed.

Scan technique

Out of the total 28 patients, 21 patients were scanned only using a 64-slice MDCT scanner (GE light speed, VCT, American) and 5 of these underwent non-contrast scan and 15 underwent contrast enhanced scan. The scanning parameters were: Tube voltage of 120kVp, tube current of 400mA, the Helical scan with pitch ratio was 1.375:1; collimation thickness was 0.625mm×64 and the slice thickness was 5mm. The contrast-enhanced scan was performed using 100 ml intravenous contrast material (Omnipaque 300; Amersham Health, Princeton, NJ). The scanning time for contrast-enhanced scan was: neck, 25s, 40s was delayed after injection. Chest, 40s was delayed after injection. 30, 60, 120s were delayed

for arterial, portal and venous phase (AP,PP,VP) scanning. The contrast agent was injected at a rate of 3mL/sec.

Similarly, 6 patients out of the total were scanned using a 3.0 T MRI unit (GE, SignaHDxt, America) only. Three of them underwent non-contrast scan and 4 underwent contrast enhanced scan. The scanning parameters were: a T1WI FSE sequence (TR720~1972ms, TE12.0~21.9ms) and FSE sequence T2WI (TR4480~8571ms, TE95.0~119.6ms), a contrast enhanced imaging was obtained with T1WI LAVA (TR 2.6,1.2) or 3D BRAVO (TR 7.8, TE3.0), by the Gd-DTPA were injected at a rate of 2~3mL/s. Moreover, 1 patient underwent both CT scan as well as MR scan, with same parameters.

Image analysis

The CT and MR images were reviewed by 2 radiologists in consensus. For each lesion, the features of location, size, margin, shape, density (CT), signal intensity (MRI), enhancement pattern, vessel sign, infiltration to adjacent tissues were analyzed. The density of the lesion was evaluated as homogeneous or heterogeneous on the plain scan and the exact CT value was measured. The pattern of enhancement was graded as slight, moderate, marked homogeneous or heterogeneous. The CT value of solid component of each phase was measured. The vessel sign of the tumor was reviewed in the arterial phase(CT) and the flow voids sign in the T2WI images(MRI).

RESULTS

Clinical findings

As the tumor's silent nature, most of the cases were incidental diagnosis. It was seen that the development of symptoms occurred at later stages of the disease and these symptoms were of different varieties depending upon the tumor location of tumors with no associated systemic symptoms. Out of the total patients, 40% (11/28) were asymptomatic at presentation, yet a bulk of them, 60% (17/28) did have specific regional symptoms. Among the symptomatic patients, Chest pain and chest tightness was seen in 7%, a palpable mass and cough was observed in 14%. Similarly, only 3.5% of the symptomatic patients demonstrated loss of consciousness with hyperspasmia, neck pain, nasal obstruction and abdominal pain. The specific proportion of distribution of symptoms are depicted in figure 6. Similarly, 42.85% (12/28) of these tumors were located in thoracic cavity, 14.28% (4/28) in the middle mediastinum, 10.71% (3/28) in the retroperitoneum, 7% (2/28) in the meninges and a single case occurred in other organs like - cervical spinal canal, parapharyngeal space, parotid gland, nasal cavity, abdominal cavity, adrenal gland and kidney. As seen in Figure 5, the tumor occurred between the ages of 25 – 78 years with a mean age falling in the 5th decade. The sex distribution of SFTs revealed that the disease occurred in 17 (60%) female while only 11 (40%) were male.

All of these patients with histological diagnosis of SFT had undergone surgical excision of the tumor. However, 3 patients with the tumor located in the thoracic cavity and mediastinum had recurrence after surgery which was noted during a 36 months follow up period.

Imaging findings

All the CT findings are presented in table 1. In total 22 lesions were found and located in different parts such as in thoracic cavity(9), mediastinum(4), retroperitoneum(3), parotid gland(1), adrenal gland(1), kidney(1), parapharyngeal space(1), abdominal cavity(1), nasal cavity(1).

It was observed that the size of these tumors were between $1.5 \times 1.0\text{cm}^2$ and $23.0 \times 16.5\text{cm}^2$, with an average size of $8.2 \times 6.8\text{cm}^2$. We found two cases with small size ($2.0 \times 1.7\text{cm}^2$ and $1.5 \times 1.0\text{cm}^2$) occurred in the nasal cavity and the cervical spinal canal respectively due to their early symptoms. 59% (13/22) of these lesions had an oval shape and well defined margins. These tumors, on ct appeared homogeneous in 45% (10/22) of the cases while they were heterogeneous in 54.5% (12/22) cases with an average CT of the solid component being 41.5 HU. Among the 18 cases that received radio-contrast, 12 of them showed slight enhancement during the arterial phase while 6 showed heterogeneous marked enhancement. Similarly, during dynamic CT scan of 5 cases it was observed that the average CT value in AP, PP, VP were respectively 46.4, 73.3, 86.5HU. Additionally, the CT scan also demonstrated cystic changes within the tumor in 9 cases while 4 lesions had calcification. 77.77 % of SFT demonstrated positive vessel sign (14/18).

Furthermore, 6 patients with SFT had undergone MR scan, the details are outline in table 3. The locations of these lesions were predominantly in the thoracic cavity (3) followed by meninges (2), and a (1) case in the mediastinum and cervical spinal canal each. The morphology observed were homogenously enhanced lesion in one case while heterogeneously enhanced in 6 cases. Similarly these tumors during the T1WI were homogeneous while they were heterogeneous, hyper or isointense on T2WI.

Pathological findings

All of the 28 lesions existed as a solitary mass and were surgically resected completely and were subjected to histopathological analysis. Among them, 21 lesions were benign SFTs, 3 were malignant and 4 were borderline. Some tumors were oval (n=18), well defined margin(n=22), others were irregular(n=11), with ill defined margin(n=7). These tumors were composed of hyper- and hypo cellular spindle cells with proliferation and surrounded by collagen fibers as depicted in figure 2C. The cut sections of these tumor had a grayish white or yellowish white color with a fish-meat like texture and the whole tumor was embedded inside the capsule. Predominantly we found single tumor in all cases.

Besides these features, some of the lesions also demonstrated irregular areas of myxoid degeneration and necrosis with internal hemorrhage. Immunological staining were strongly positive for CD34, Vimentin, Bcl-2, while negative for SMA, CK, the details of which are summarized in table 4.

Table 1: CT findings of 22 patients.

Case	Size (cm ²)	Shape/margin	Density/ Cystic changes	Enhancement pattern	Vessel sign	Infiltrative manifestation	Previous diagnosis	Nat-ure	Rec.
1	8.2x 9.0	Oval/Well-defined	Homo /-	Slight, Hetero	+	-	Mesothelioma	B	-
2	7.2x 10.0	Irreg/Well-defined	Homo /-	/	/	-	Mesothelioma	B	-
3	17.2x14.5	Irreg/well-defined	Hetero /+	Slight, Hetero	+	Pericardial and Plural effusion	Sarcoma	B	-
4	4.8x 6.7	Irreg/Ill-defined	Hetero /-	/	/	+	Sarcoma	M	+
5	10.3x8.0	Irreg/Well-defined	Homo /-	Slight, Hetero	+	-	SFT	B	-
6	6.3x 6.0	Oval/Well-defined	Hetero /+	Slight, Hetero	+	-	Mesothelioma	B	-
7	12.7x11.0	Oval/Well-defined	Homo /-	Slight, Hetero	+	Pleural effusion	Sarcoma	B	-
8	7.1x 8.0	Irreg/Ill-defined	Homo /+	Marked, Hetero	-	+	Thymoma	B	-
9	2.4x 11.0	Oval/Well-defined	Hetero /+	/	-	Pleural effusion	Leiomyosarcoma	B	-
10	4.6x 6.5	Irreg/Ill-defined	Hetero /+	Marked, Hetero	-	-	Mediastinum-type lung Ca	B/M	-
11	7.0x 8.5	Oval/Well-defined	Hetero /+	Slight, Hetero	-	Pericardial effusion	Schwan-noma	M	+
12	16.5x23.0	Oval/Well-defined	Hetero /+	Slight, Hetero	+	Pericardial effusion, Heart displacement	SFT	B/M	-
13	5.4x 5.5	Oval/Well-defined	Homo /-	Marked,Hetero	+	-	Lymphoma	M	+
14	1.7x 2.0	Oval/Well -defined	Homo /-	/	/	-	Fibroma	B	-
15	4.8x 3.0	Irreg/Ill-defined	Homo /-	Marked,Hetero	+	+	Nasopharyngeal angiofibroma	M	-
16	5.8x 5.0	Irreg/Ill defined	Hetero /-	Marked,Hetero	+	+	Nasopharyngeal Ca	B	-
17	7.0x 8.5	Oval/Well-defined	Homo /-	Slight, Hetero	-	-	Adrenal adenoma	B	-
18	6.8x 5.0	Oval/Well-defined	Homo /-	Marked,Hetero	+	-	Castleman disease	B/M	-
19	12.7x15.0	Oval/Well-defined	Hetero /+	Slight, Hetero	+	-	Renal Ca	B	-
20	10.3x11.0	Oval/Ill-defined	Hetero /+	Slight, Hetero	+	-	Sarcoma	B	-
21	19.8x17.5	Irreg/Well-defined	Hetero /-	Slight, Hetero	+	Embrace to kidney	Liposarcoma	B/M	-
22	10.4x11.0	Oval/Well-defined	Hetero /-	Marked,Hetero	+	-	Hemangiopericytoma	B	-

Table 2: MRI findings of 7 patients.

Case	Size(cm ²)	Shape	Flow voids sign	T1WI	T2WI	Enhancement pattern	Infiltrative manifestation	Previous diagnosis	Nature	Rec.
1	15.6 x 13.0	Oval/Well-defined	+	Hypo	Hyper	Slight, Hetero	Pleural effusion	Sarcoma	B/M	
2	3.5 x 3.2	Oval/Well-defined	+	Iso	Mild-hyper	Marked, Homo	-	Mesothelioma	B	
3	11.6 x10.0	Irreg/Well-defined	+	Iso	Mild-hyper	/	-	Mesothelioma	B	
4	5.5 x 6.0	Oval/Well-defined	+	Hypo and iso	Hyper and iso	Slight, Hetero	-	Schwannoma	B/M	
5	3.4 x 4.4	Irreg/Well-defined	+	Iso	Iso	Marked, Homo	-	Meningioma	B/M	
6	6.7 x 7.7	Oval/Well-defined	+	Hyper	Hyper	/	-	Schwannoma	B/M	
7	1.5 x 1.0	Oval/Well-defined	-	Iso	Mild hyper	/	-	Fibroma	B	

Abbreviations: Table 1, 2 : Homo- homogeneous, hetero- heterogeneous, iso- isointense, hypo- hypointense, hyper- hyperintense, B- benign, M- malignant, B/M- borderline malignant, '+' positive, '-' negative, Ca- carcinoma, CT- computed

tomography, MRI- magnetic resonance imaging, T1WI- T1 weighted imaging, T2WI- T2 weighted imaging, irreg- irregular, '/' not done, Rec- recurrence.

Table 3: The immunohistochemical results of 28 patients.

Case	CD-34	Vimentin	CD 99	Bcl-2	S-100	SMA	CK	Desmin	CD117	KI-67	EMA	Nature
1	+	+	+	+	-	-	-	-	-	-	-	B
2	+	+	+	+	-	+	-	-	-	+	-	B
3	+/-	+	+	+	-	+	-	-	-	+	+	M
4	+	-	+	+	-	-	-	-	-	+	-	B
5	+	+	+	+	+	-	-	-	-	+	-	B
6	-	+	+	+	+	-	-	-	-	+	-	B/M
7	+	+	-	+	-	-	-	-	-	10%	-	B
8	+	+	+	+	-	+	-	-	+/-	5%	-	B/M
9	+	+	+	+	+	-	-	-	-	8%	-	M
10	+	-	+	+	-	-	-	-	+	30%	+	B
11	+	+	+	+	-	-	-	-	-	1%+	-	B
12	-	+	+	+	-	+	-	-	-	+	-	B
13	-	+	+	+	-	-	-	-	-	<1%	-	B/M
14	+	-	+	+	-	-	-	-	-	1%+	-	B
15	+	+	+	+	-	-	-	-	-	+	-	M
16	+	+	+	+	+	-	-	-	-	20%+	-	B/M
17	+	+	+	+/-	-	-	-	+	-	10%+	-	B
18	+	+	+	+	-	-	-	-	-	4%+	-	B
19	+	+	+	+	-	-	-	-	-	2%+	-	B
20	+	+	-	+	-	+	-	-	-	3%+	+/-	B

21	+	+	+	+	-	-	-	-	-	8%+	-	B
22	+	+	+	+	-	-	-	-	-	3%+	-	B
23	+	+	+	+	-	-	-	-	-	3%+	-	B
24	+	+	+	+	-	+	-	+	-	8%+	-	B
25	+	+	+	+	-	-	-	-	-	1%+	-	B
26	+	+	+	+	-	-	-	-	-	4%+	-	B
27	+	+	+	+	-	-	-	-	-	7%+	-	B
28	+	+	+	+	-	-	-	-	-	4%+	-	B

Abbreviations : Table 3: '+'positive, '-'negative, '+-'partial positive, 'B'benign, 'M'malignant, 'B/M' borderline malignant.

DISCUSSION

SFT's being rarer tumors, with thorax being the predominant location, our findings too supported this fact (1) although minimal occurrence were noticed in extra thoracic locations too. These tumors were have been commonly misdiagnosed with other benign slow growing tumors. Mostly the tumor, in our study were seen to be asymptomatic, with the growth of the tumor, symptoms were seen to occur according to locations like chest pain or distress. This disease typically affected old patients in their 50s with female sex being predominant. We were of the opinion that a female patient in their 50s presenting with regional symptoms can give a clue to the diagnosis of SFT. These are also commonly misdiagnosed in the thoracic cavity as diagnosed as sarcomas and mesotheliomas, schwannoma and lung cancer in the mediastinum and meningioma and angiofibroma in the head and neck region while hemangioma, hemangiopericytoma and stromal tumor in the abdominal cavity.

Similarly, the CT images of our study was found to be a powerful diagnostic tool for SFT. Even the plain CT was able to localize a well defined oval shaped tumor with variable size (smaller in the subcutaneous or superficial organs and larger in deep tissues). The most consistent size to look for in plain CT scan would be between 2.0 X 1.7 cm² and 1.5 X 1.0 cm². Similarly the smaller lesions had a homogeneous isodense solid lesions while larger lesions were homogeneous with low density. The average CT value of solid component observed in our study was 41.5 HU. It was further found that the lesions with areas with myxoid degeneration or necrosis that present as low density or cystic changes can be SFT that was in lieu with previous studies that had demonstrated areas of hyalinization, necrosis and myxoid degeneration.^[6] As known, the images in contrast CT depends on the enhancement pattern of the tumor components and proportion of spindle cells and collagen fibers. Marked enhancement was observed in hypercellular lesions and blood vessel-rich area while slight enhancement was observed in hypo-cellularity lesions with rich fiber stromal area. Our study with CT enhancement too demonstrated homogenous or heterogeneous enhancement on AP and persistent enhancement on delayed phase (type 1), signifying the hyper-cellular and vascular-rich characteristics of SFT. Moreover the slight enhancement on AP with progressive enhancement on delayed phase (type 2), also verified the hypo-cellular and rich collagenous component within the tumor. All of these enhanced lesions in the abdominal region demonstrated a slight enhancement in the arterial phase (AP) and gradually enhanced in portal phase (PP) and venous phase (VP) (type 2) with a value of 46.4, 73.3, 86.5 HU respectively. These findings does allow us to use contrast CT as a tool for diagnosing tumors with varieties of fibrous, vascular and cellular components. Moreover, these findings, correspond with previous studies^[5] too. (and sft of the liver 2013). Additionally, another striking feature observed with SFT were the serpentine

vessels that appeared during the arterial phase^[16], also known as the 'vessel sign', which can give an aid in giving a clue to the diagnosis of SFT although its nature cannot be based on this feature alone.

Magnetic resonance imaging can be very helpful in distinguishing the fibrous and collagenous component of SFT, which are either homogeneously iso/hypo-intense on T1WI while heterogeneously iso/hyper-intense signal on T2WI. Moreover, hyperintense signals additionally allows us to know whether there are necrotic/ cystic changes or myxomatous degeneration within the lesion.^[5] One case occurred in frontal lobe meninge, present as a mixed signal mass with crescent hyper-intense area, which is called a "black-and-white" mixed pattern or "yin-yang" pattern in reported literature.^{[3],[12]} Similarly, enhancement with gadolinium (contrast agent) further can visualize smaller SFTs that are seen as homogeneous lesions while larger lesions can be seen as heterogeneous images. Similarly, SFTs being rich in fibers and collagenous stroma, this property can be exploited by enhancing from the arterial phase to the delayed phase on dynamic contrast enhanced images.^[7] This gadolinium enhancement can also demonstrate flow void sign on T2WI which is an important diagnostic clue towards SFT. **Error! Bookmark not defined.** (sft of the liver 2013 and imaging findings of solitary fibrous tumor in the abdomen and pelvis 2014 and sft in abdomen and pelvis: imaging characteristics and radiologic-pathologic correlation), which can reveal hyper-vascularity of SFT. The lesions in our study too demonstrated all of these aforementioned features.

Furthermore, the usefulness of these imaging modalities in differentiating benign from malignant lesions, as observed in other studies were quite inconsistent to our findings. The key reason could be the small number of malignant case encountered and its rarity (approximately 10%~15% of tumors naturally are malignant) could have been the variables. However, apart from the morphology, the radiology was effective in demonstrating ill-defined margin with invasion to adjacent tissues and neighboring infiltrations. Similarly, the malignant lesion occurred in 50% in the mediastinum while another 50% of the tumor was borderline. The distributions of tumors in our study that revealed one of the lesions recurred locally one year after surgery which has prompted us to state that tumors occurring in the mediastinum have more chances of being malignant with higher recurrence rate.

CONCLUSION

From this retrospective study, we can say that the imaging studies may be sufficient enough to diagnose SFT although, as with any other tumor, tissue diagnosis remains to be the gold standard. A large well-defined hyper-vascular tumor with variable degrees of necrosis and cystic change and marked, persistent enhancement or slight and progressive enhancement in CT scans and homogenous, iso- or hypo-intense signal on T1WI, with heterogeneous iso- or hypo-intense signal, serpentine

vessels and positive flow voids sign on T2WI are important features of the tumor diagnosis in the MR scans. These features in any small, hidden mass with regional symptom should direct a clinician to consider SFT as a differential and guide for histological examinations for specific cell markers which would cut down the extra burden of cost, time, energy and misdiagnoses.

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