The first report of an intercostal hemangioma coexisting with multiple hepatic hemangiomas: a case report and review of the literature

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Case Report

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Abstract

Background

Intercostal hemangioma (IH) is an extremely rare disease, with only 17 cases reported over the past 30 years, and the case of IH coexisting with multiple hepatic hemangiomas (MHHs) has not been reported up to now. IHs appear to be more aggressive than hepatic hemangiomas and are difficult to distinguish from other intercostal tumors.

Case presentation:

we report the first case of an intercostal venous hemangioma (IVH) coexisting with MHHs. The patient was a 58-year-old male who came to our hospital due to long-term and repeated epigastric distention. Plain chest computed tomography (CT) scans revealed two adjacent masses protruding from the left chest wall into the thorax. The nature of the masses remained unclear after enhanced CT scans. We performed video-assisted thoracoscopic surgery to remove the masses. These masses were finally diagnosed as IVHs, based on surgical records, imaging, and pathological findings. In addition, plain abdominal CT scans also indicated the presence of MHHs, this diagnosis was consistent with the patient's two doppler ultrasound findings over the last two years. There was no significant change in the hepatic masses when examined at 32 months follow-up. Unfortunately, there was a recurrence of the left IH, and the 5th rib was slightly eroded and destroyed. This may have been related to the fact that we did not distinguish the nature of the masses before surgery and the incomplete removal of masses during surgery.

Conclusions

Early clinical intervention for IHs of the chest wall can prevent tumor growth and damage to adjacent structures. Preoperative imaging diagnosis is important. Hemangiomas have a high recurrence rate, Complete surgical resection is the common treatment, and chest wall reconstruction can be performed if necessary. The authors hope that our case analysis and literature review will help clinicians identify IVH as highly unusual.

Background

Intercostal hemangioma (IH) is an extremely rare form of benign tumor which can easily be forgotten in the differential diagnosis of intercostal tumors. Patients often come to hospital with a painless mass in their chest. However, IH usually needs to be confirmed by pathology after surgical resection. Herein, we report the first case of an intercostal venous hemangioma (IVH) coexisting with multiple hepatic hemangiomas (MHHs). Our patient had two IVHs. Computed tomography (CT) scans failed to provide a correct diagnosis. After masses were removed by video-assisted thoracoscopic surgery (VATS), the IVHs were diagnosed based on surgical records, imaging, and pathological results. The multiple hepatic masses were diagnosed as MHHs by the results of two doppler ultrasound examinations and two CT scans. Unfortunately, IH recurred at 32 months follow-up. To the best of our knowledge, this is the first case of its type to be reported. In addition, we reviewed 17 cases of IH reported in the English literature over the past 30 years, hoping to provide help for clinical research and diagnosis of IH.

Case Presentation

The patient was a 58-year-old male who had smoked (40 cigarettes/day) and drunk (250g/ day) heavily for more than 30 years. He came to our hospital on the 10th April 2019 for treatment because of repeated upper abdominal fullness and discomfort. He had suffered from pulmonary tuberculosis 30 years ago and the symptoms disappeared after treatment. On the 20th October 2017, doppler ultrasound examination suggested multiple hepatic masses; MHHs were considered. The patient had no symptoms of chest discomfort. His mental state was good, and there had been no significant loss of weight. There was no history of chest trauma. His granddaughter had a congenital hemangioma on her forehead.

Gastroscopy revealed esophagitis, erosive gastritis and duodenitis. Abdominal doppler ultrasound revealed multiple hepatic masses again; these were considered to be MHHs again. Plain chest CT scan images showed two adjacent masses on the left chest wall, with slightly uneven density and sizes of 1.9 x 1.3cm and 1.1 x 0.9cm respectively. CT values ranged from − 10HU to 10HU, with an average of 1HU. The masses protruded into the thorax and were surrounded by a "capsule". These were laterally connected to the chest wall with a broad base and reached the 5th intercostal region. The normal intercostal muscle structure was interrupted and had disappeared. These were thought to be chest wall cysts or localized encapsulated effusions(Fig. 1A). There was an old pulmonary tuberculosis in the upper lobe of the right lung. In addition, scattered small nodules in both lungs and bilateral pleural thickening were considered as inflammatory changes. The patient also had bronchitis and emphysema. Abdominal plain CT revealed multiple slightly low-density nodules and masses in the liver with clear boundaries; the largest of these was about 3.5cm x 2.8cm.

According to the chest contrast-enhanced CT (CECE) scan images, in the arterial phase (Fig. 1B,1D), the two adjacent masses presented with nodular/tubular enhancement (CT values ranged from 0HU to 34HU, with an average of 16HU), and in the delayed phase (Fig. 1C,1E), the contrast agent presented further enhancement of filling (CT values ranged from 0HU to 40HU, with an average of 29HU). The intercostal artery ran along the lower margin of the "capsule" and the lateral edge of the two masses. The CECT diagnosis suggested that these may be neurogenic tumors or hamartomas, even though they exhibited the typical "fast in, slow out" and progressive filling enhancement. Hemangiomas are rare in our region. According to the abdominal CECT scan images, the edges of the hepatic masses were significantly enhanced in the arterial phase (Fig. 2G). The portal vein phase (Fig. 2H) and the delayed phase (Fig. 2I) gradually filled and strengthened to the center. This was typical of the enhancement associated with hepatic hemangioma; thus, these were

considered as MHHs. Physical examination revealed slightly coarse breath sounds in both lungs. Laboratory examinations showed no evidence of thrombocytopenia or abnormal coagulation.

The surgeon performed VATS exploration on the patient, and found a “tumor” of approximately 3 x 2 cm in size with a clear boundary and a complete capsule. The intercostal nerve traversed the capsule, and two tumors were visible inside when the capsule was opened; the tumors were then completely removed. In addition, multiple pleura adhesions were found in the left thoracic cavity and were severed. The intraoperative diagnosis was “left posterior mediastinal tumors/neurogenic tumors?”. Pathological findings suggested hemangiomas (Fig. 3J,3K). We invited two chief physicians from the Department of Pathology to classify the tumors independently, and for both masses, they gave the diagnosis of venous hemangioma (VH). However, the origin of the tumors had not been confirmed by pathological examination; the surgically removed tissues had been discarded and could not be examined.

We strongly suspected that the clinician had made a mistake in his description of the location of the tumors; these are not associated with the posterior mediastinum according to relevant literature [1]. The surgeon told us that the surgical record was written by a doctor who was undergoing standardized training. At this time, it had been too long since the surgery, and the surgeon could not accurately describe the location of the tumors. Pathologists had referred to the location of the tumors described by the clinicians and did not make an independent judgment. Therefore, two chief radiologists in our team (both of these have been engaged in imaging diagnosis for more than 20 years) judged the location of the tumors from the CT images independently. The radiologists strongly agreed that the tumors originated from the chest wall intercostal space. Normally, the intercostal artery passes behind the corresponding intercostal space between the intrathoracic fascia and the intercostal intima. The intercostal nerve runs underneath the corresponding intercostal artery. A review of CT scan images showed that the intercostal artery ran along the lower edge of the “capsule” and the lateral edge of the two masses; the normal intercostal muscle structure was interrupted and disappeared. According to the surgical records, the "capsule" was intact, and the intercostal nerve traversed the "capsule" (hemangiomas are known not to have capsules). Therefore, the "capsule" contained parietal pleura and intercostal intima (at least) and these two VHs represented tumors of the intercostal area. According to the literature, these should have originated in the intercostal muscles.

The patient presented with multiple hepatic masses without any symptoms of hepatic discomfort. The patient's two doppler ultrasound findings within two years and the CT scan results prior to VATS were considered to reveal MHHs. Therefore, surgical resection of the hepatic masses was not performed; 32 months later, CT showed no significant changes in the hepatic masses.

Literature review

We searched the English literature relating to IH from January 1990 to January 2022 and found 17 case reports [2–18]; the present study represents the 18th case report (Table 1). By screening these case repor ts we identified the involvement of 10 males and 8 females. Seven patients were under 30 years old, one patient [7] was found to have hemangioma in infancy (2-year-old); nine patients were affected on both the left and right side. Three patients had VH, five patients had cavernous hemangioma (CA), two patients had arteriovenous hemangioma (AVH); three patients had mixed hemangioma (MH); there was one case of intramuscular hemangioma of small-vessel type (SVIH) and one case of intramuscular hemangioma of large-vessel type (LVIH). The subtypes were not specified in three cases. Hemangioma was successfully diagnosed by magnetic resonance imaging (MRI) in four cases and by CT in two cases. Phlebolith was detected in two hemangiomas [5, 8]. Eleven patients were without chest symptoms; 2 patients were not specified. Two patients had a history of trauma, two patients had other vascular lesions and five patients had rib destruction.

Table 1
Characteristics of patients in published English literature

Author	Age, sex	Size (cm)	Intercostal space	Symptoms of chest	imaging diagnosis		Rib destruction	Pathology	Follow up	medical history
					CT	MRI				
Wincheste [2]	39y, F	5	Right (5)	Discomfort, fullness	Unclear	-	N	CA	-	-
Robinson [3]	58y M	3	Right (3)	No symptoms	Pleural-based discrete mass	-	N	CA	-	-
Hashimoto [4]	32y, M	1.2	Left (6)	No symptoms	Neurogenic tumour	Hemangioma	N	VH	N (24m)	-
Dzian [5]	36y, M	9.5X9x3	Left (7 ~ 8)	Pain	Unclear	Hemangioma	Unclear	VH	N (10m)	-
Dantis [6]	18y, M	6x4.5x4	Left (7)	Swelling, pain	Unclear	-	Y	CA	N (12m)	-
Ulku [7]	11y, F	8.5x7.5x5.5	Right (9 ~ 11)	Unclear	Unclear	Unclear	N	CA	N (6m)	-
Saldanha [8]	34y, F	3x2	Left (3)	Unclear	Hemangioma	-	Unclear	CA	N (12m)	-
Elbawab [9]	14y, M	6x4x3.5	Right (5)	No symptoms	Unclear	Unclear	Y	Hemangioma	N (6m)	-
Agarwal [10]	44y, F	unclear	Right (3)	No symptoms	Unclear	Hemangioma	N	Hemangioma	N (18m)	-
Mei [11]	14y, F	7x6x5	Right (4)	Pain, hemoptysis	Pulmonary metastasis	-	Y	Hemangioma	N (10m)	Trauma
Aguilo [12]	23y, F	4x5	Left (7)	No symptoms	-	-	N	AVH	-	Subdural AVM
Kara [13]	46y, M	4	Right (2)	No symptoms	Hemangioma	-	N	AVH	N (48m)	-
Yuan [14]	44y, F	unclear	Left (2)	No symptoms	Vasculopathy	Unclear	N	CA + CH	N (36m)	-
Ono [15]	33y, M	9x9.5	Right (3 ~ 5)	No symptoms	-	Unclear	N	SVIH + LVIH	N (36m)	-
Ali [16]	22y, F	7x7x6	Left (6 ~ 9)	No symptoms	Unclear	Unclear	Unclear	CA + CH	N (6m)	-
Yonehara [17]	33y, M	5x5	Left (6)	No symptoms	Tumors of bone/cartilage	-	Y	SVIH	N (60m)	Trauma
Kubo [18]	27y, M	5.5x3.5	Right (7)	Pain, exertional dyspnea	Unclear	Hemangioma	N	LVIH	N (6m)	-
Current case	58y, M	3x2	Left (5)	No symptoms	Neurogenic tumor/ Hamartoma	-	Y (after 32m)	VH	Y (32m)	MHHs

y, years; m, months; M, male; F, female; Y, yes; N, no; AVM, arteriovenous malformation.

Discussion And Conclusions

Hemangiomas are very rare in the chest wall, especially in the intercostal region. Most IHs originate from the intercostal muscles but can also originate from intercostal connective tissue outside the pleura [10, 14]. Hemangiomas may occur inborn or due to trauma, and two cases of hemangiomas in our included literature were detected after trauma. Our patient had no history of chest trauma but did have MHHs. The definition of hemangioma has always been controversial although people generally believe that a hemangioma is a benign tumor. According to the size of the vascular lumen, the dominant type of vascular structure, and the shape and color of the tumor, these can be divided into a variety of different subtypes. The most commonly used classification is based on the type of vascular structure, including CH, CA, VH, AVH and MH. However, these classification methods do not take into account the patient's age, clinical manifestations, and pathological behavior. The International Society for the Study of Vascular Abnormalities (ISSVA) [19] recommends distinguishing vascular malformations (e.g., capillary, lymphatic, venous, and arteriovenous malformations) from vascular tumors (e.g., infantile hemangioma) rather than calling them all "hemangiomas". A vascular tumor is a proliferation of vascular endothelial cells and has three different stages of development: proliferation, stasis and degeneration. Vascular malformations do not show signs of degeneration [20]; endothelial cells are normal but can be

enlarged by hypertrophy [21]. This classification method is helpful for the diagnosis, treatment, and prognosis of vascular diseases. However, whether intramuscular hemangiomas represent vascular tumors or malformation has not been determined; we still refer to these as "hemangiomas".

Imaging examinations play an important role in the diagnosis of hemangiomas, the selection of appropriate clinical treatment plans, and the follow-up strategy. CT scans can reveal the location, shape, density, extent of involvement, and mode of enhancement of the mass, which is also sensitive to phlebolith. Phlebiolith is a unique sign of hemangiomas [22]. Magnetic resonance imaging (MRI) can scan a mass in multiple directions and sequences and provides high resolution images of the soft tissue. Qualitative analysis of masses can be carried out according to signal intensity and enhancement mode can be displayed for different sequences. By reviewing the reported literature relating to plain CT images, IH is a soft tissue mass with a clear boundary that features a uniform or slightly uneven density that may include lobulation or Phleboliths. IH generally has medium or slightly lower signals than adjacent muscle tissue on T1-weighted MRI, and high signals on T2-weighted MRI, and can retain high signals even after fat suppression. Contrast-enhanced CT and MRI scans can show a similar pattern of enhancement to the HH, with typical "fast in, slow out" or gradual "centripetal filling" enhancement. Nodular or tubular enhancement can be recognized in some IHs. However, some intercostal hemangiomas can also have no obvious characteristics. The imaging diagnosis should be differentiated from neurogenic tumors, fibromas, hamartomas, pleural tumors, and bone tumors (when the bone is damaged).

Early clinical intervention for IHs of the chest wall can prevent tumor growth and damage to the adjacent structures, such as the bone. We do not recommend preoperative biopsy of a mass with a suspected hemangioma on imaging because of the risk of massive bleeding and the possibility that biopsy results may remain inconclusive. The management of intercostal hemangioma is related to the patient's age, location, size, and depth of invasion of the mass. According to our literature review, periodic observation was performed in only one case. Surgical resection is a common method for the treatment of IH. Preoperative imaging diagnosis has important guiding significance for surgery. If there are large blood vessels supplying the mass, then vascular embolization or ligation is recommended before resection. In addition, we recommend extensive resection of a tumor until no tumor remains. Chest wall reconstruction should be performed if necessary; this can effectively avoid the recurrence of hemangioma.

Abbreviations

IVH
Intercostal venous hemangioma
VH
Venous hemangiomaMHHs:Multiple hepatic hemangiomas
MH
Mixed hemangioma
SVIH
Intramuscular hemangioma of small-vessel type
LVIH
Intramuscular hemangioma of large-vessel type
MRI
Magnetic resonance imagingCT:Chest computed tomography
VATS
Video-assisted thoracoscopic surgery
CECE
Chest contrast-enhanced CT

Declarations

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

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Disclosure

Ethics approval and consent to participate: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Consent for publication：All authors have approved the manuscript for publication in this journal.

Competing interests: Authors report no conflict of interest.
Authors' contributions: X.G.L. and L.Y. collected case data, reviewed relevant literatures and wrote this manuscript. Y.H.Z. and G.S. analyzed this case and

reviewed relevant literatures. X.W. reviewed the relevant literatures and critically reviewed the manuscript for its intellectual content. All authors read and approved the final manuscript.

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Figures

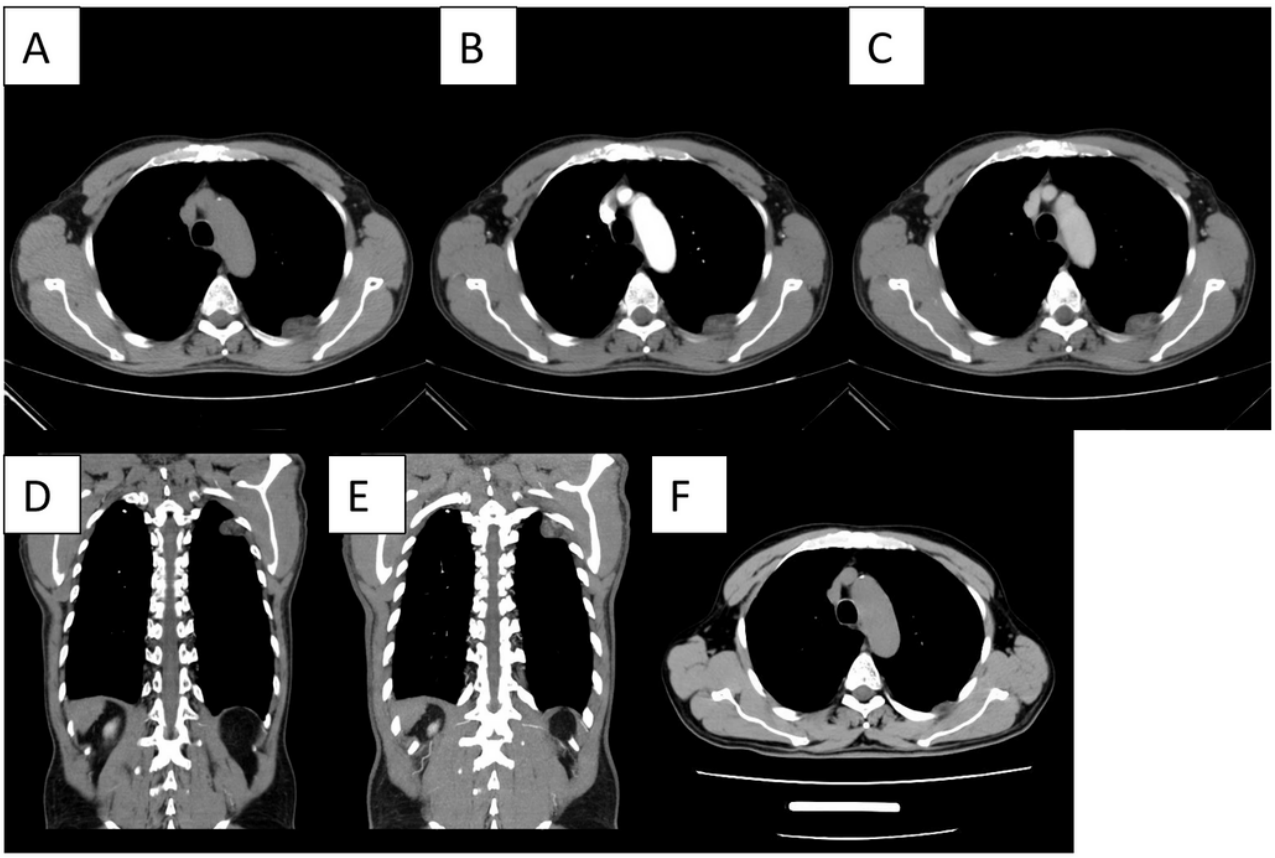

Figure 1

Plain chest CT (A) showed two adjacent low-density masses on the chest wall, the normal intercostal muscle structure was interrupted and disappeared; Nodular/tubular enhancement was seen in the masses at the arterial stage (B, D) ; the contrast agent was further enhanced by filling in the delay period (C, E); 32 months later, plain CT revealed recurrence of hemangioma (F).

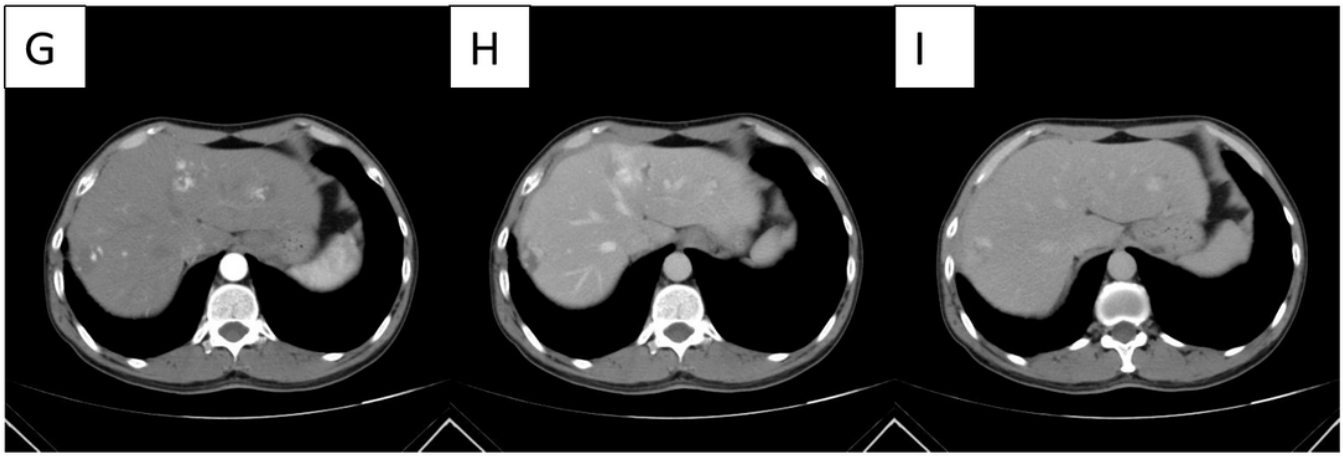

Figure 2

On enhancement, multiple hepatic masses showed typical "fast in, slow out" and "centripetal filling" enhancement.

Figure 3

(H&E staining) The walls of blood vessels were made of smooth muscle of varying thickness; the lumens varied in size, they were separated by fibrous connective tissue, and adipocytes were seen; There is no proliferation or atypia in flattened endothelial cells, and red blood cells are filled or scattered in some vascular lumens.