Angiosarcoma: Review of CT and MR Imaging features

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Learning objectives

To review 1) the clinical features of angiosarcoma, 2) its imaging findings (on magnetic resonance imaging (MRI), computed Tomography (CT), and fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET)), and 3) its differential diagnosis. This knowledge will be helpful in prompt and accurate diagnosis.

Background

Angiosarcoma is a rare mesenchymal malignant neoplasm of the vascular or lymphatic endothelium, accounting for 2% of all soft-tissue sarcomas [1-4]. They can affect any organ but tend to occur in the skin in the head and neck regions [3, 4]. Although primary hepatic angiosarcoma is rare and accounts for only 2% of primary hepatic tumors, it is the most common malignant mesenchymal tumor of the liver [5]. Here, we present a few educational cases of angiosarcoma and corresponding CT, MRI and FDG-PET images (Table 1, Figure 1~7). The radiologic characteristics of angiosarcoma are demonstrated in the images.

Findings and procedure details

Hepatic angiosarcoma

Primary hepatic angiosarcoma is a very rare and aggressive malignancy of mesenchymal origin. It accounts for less than 2% of all primary liver neoplasms [6]. The tumor most commonly affects men between 50 and 70 years of age. Its association with exposure to several environmental carcinogens, such as thorotrast, arsenic, and vinyl chloride is well known. However, exposure to these agents is now rare, and the etiology of most hepatic angiosarcomas remains unknown [7, 8]. At the time of initial diagnosis, the majority of patients present with multifocal growth within the liver as well as with extrahepatic metastatic spread, which make for a poor prognosis [8, 9]. Abdominal CT scans show heterogenous lesions, more frequently hypodense, with peripheral enhancement, and two growth patterns, multinodular and solitary mass. T1-weighted magnetic resonance imaging shows low intensity lesions with focal areas of high intensity which suggest hemorrhage [5]. FDG uptake is moderate or high [10]. The reason FDG accumulates in hepatic angiosarcoma is not known. However, based on the findings of experimental and clinical studies, the degree of FDG accumulation in various tumors is said to generally depend on the activity of cell membrane glucose transporters (especially GLUT-1), and intracellular hexokinases.
Cardiac angiosarcoma

Primary cardiac tumors are extremely rare, with an incidence estimated at 0.0017% to 0.05% [11]. Two-thirds are benign, 50% of which are myxomas [12]. The remaining 25% are malignant, and the majority of these are angiosarcomas [13]. When compared to echocardiography, CT and MRI reveal more detail in the cardiac soft tissues and can define extracardiac involvement and metastasis. Calcifications associated with tumors can be visualized with CT, which may be an advantage over MRI. Additionally, CT does not have the imaging limitations associated with MRI and may also be used for transthoracic biopsies. Primary cardiac tumors are frequently heterogenous and isointense on T1-weighted images and hyperintense on T2-weighted images. Tumors are sometimes heterogenous on T2-weighted images. This may be due to infiltration of the pericardial space with hemorrhagic and necrotic material [14].

Breast angiosarcoma

Primary angiosarcoma of the breast is very rare, accounting for 0.04% of malignant breast neoplasms, and is most frequent in women between the ages of 30 to 40. Several studies have reported that clinical signs are usually non-specific and that MRI can detect and characterize the lesion better than mammography and ultrasonography [15,16]. MRI features of breast angiosarcoma are a mass with heterogeneously low-signal intensity on T1-weighted images and high signal intensity on heavily T2-weighted images [17]. Angiosarcoma of breast is an endothelial malignant tumor with poor prognosis because of the frequency of metastasis and recurrence [18].

Skin

Angiosarcomas account for less than 0.1% of all head and neck cancers [19,20]. The imaging features in the presented case are consistent with the few reports describing cutaneous angiosarcoma in the setting of lymphedema [21,22,23]. MRI has an especially important role in the evaluation of cutaneous angiosarcoma. The extent of tumor is often larger than apparent on physical exam, leading to clinical underestimation of disease. But in our knowledge, there are no reports in the English literature describing the MRI features of idiopathic extremity angiosarcoma, that is, angiosarcoma outside the setting of chronic lymphedema and post therapy.
Images for this section:

Table 1. Patient List

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<td>10.7</td>
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**Note.** - SUVmax = maximum standardized uptake values

Table 1
**Fig. 1:** Images show hepatic angiosarcoma in a 64-year-old man. (A, D) Axial CT scan shows multiple hypoattenuating liver lesions (arrow). (B, E) On the axial contrast-enhanced CT images, some of the lesions show ring-shaped peripheral enhancement. (C, F) FDG-PET/CT shows ring-shaped uptakes and cold centers most likely representing necrosis or hemorrhage.

![Images of hepatic angiosarcoma](image1.png)

**Fig. 2:** Cardiac angiosarcoma in a 37-year-old man. (A, C) Contrast-enhanced chest CT shows a solid mass (arrow) arising from the lateral wall of the right atrium, measuring 55 x 45 mm. Most of the mass is located inside the right atrium. (B, D) FDG-
PET/CT demonstrates increased uptake of FDG in the right atrium (arrow). The maximum standardized uptake value (SUVmax) was 9.9.

**Fig. 3:** Figure 3: Breast angiosarcoma in a 35-year-old woman. (A) Axial contrast-enhanced CT scan shows a heterogeneous enhanced mass of the left breast, measuring 91 x 68 mm. (B, C) Axial magnetic resonance image (MRI) shows that the breast lesion is hypointense on T1-weighted image (T1WI) (B), and hyperintense on T2-weighted image (T2WI) (C). (D, E) Axial MRI (prone) is hyperintense on diffusion-weighted image (DWI) (D) and heterogeneous on the apparent diffusion coefficient (ADC) map (E). (F, G, H) Dynamic sagittal 3D Vibrant plane- (F), early- (G) and late- (H) phase images show persistent heterogeneous enhancement of the mass.

**Fig. 4:** Figure 4: Images show cutaneous angiosarcoma of right forehead in a 81-year-old man. (A) Axial contrast-enhanced CT scan shows homogeneously enhanced thickened skin of the right forehead (arrow). (B) FDG-PET/CT demonstrates increased uptake in
the thickened skin (arrow). (C, D, E, F, G) Axial MRI shows that the forehead lesion is hypointense on T1WI (C), slightly high-intensity on T2WI (D), hyperintense on DWI (E). There is heterogeneous signal intensity on ADC map (F) and homogeneous enhanced (G).

**Fig. 5:** Images show cutaneous angiosarcoma of right occiput in a 67-year-old man. (A) Axial CT scan shows iso-dense thickened skin of the right occiput (arrow). (B) Axial contrast-enhanced CT scan shows ring-shaped enhancement (arrow). (C, D, E, F, G) Axial MRI shows that the occipital lesion is low-intensity on T1WI (C), slightly high-intensity on T2WI (D), high-intensity on DWI (E), heterogeneous signal intensity on ADC map (F) and shows ring-shaped enhancement (G).

**Fig. 6:** Images show cutaneous angiosarcoma of left hip in a 66-year-old man. (A) Axial CT scan shows iso-dense lesions of left hip (arrows). (B) Axial contrast-enhanced CT scan shows heterogeneous enhancement (arrows). (C, D, E, F) Axial MRI shows that the left hip lesions are slightly hypointense on T1WI (C), hypointense on T2WI (D), hyperintense on DWI (E) and heterogeneously enhanced (F). (G) FDG-PET/CT demonstrates increased uptake in the lesions (arrows)
**Fig. 7:** Images show cutaneous angiosarcoma of left leg in a 66-year-old man (same patient in Figure 6). (A) Axial CT scan shows a hypodense lesion of left hip (arrow). (B) Axial contrast-enhanced CT scan shows heterogeneous enhancement (arrow). (C, D, E, F) Axial MRI shows that the left hip lesions are iso-intense on T1WI (C), iso-intense on T2WI (D), hyperintense on DWI (E) and heterogeneously enhanced (F). (G) FDG-PET/CT demonstrates increased uptake in the lesions (arrow).
Conclusion

We herein described the CT, MRI and FDG-PET images of angiosarcoma in various lesions. Studies have shown that better prognosis is associated with smaller tumors and absence of metastasis [24, 25]. This emphasizes the importance of early diagnosis. Radiologists should learn clinical features, CT, MRI and FDG-PET findings, and differential diagnose angiosarcoma and differentiate it from other tumors. The radiologic characteristics of angiosarcoma are heterogeneous signal intensity on the ADC map and strong enhancement in hyperintense area on diffusion-weighted image (DWI) and T2WI (other tumors in the differential diagnosis tend to enhance in the hypointense areas on the ADC map). FDG uptake in the same area is also a valuable sign in differentiating angiosarcoma.

Personal information

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References


