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CASE REPORT

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Angiographic features of rapidly involuting congenital hemangioma (RICH)

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Abstract Rapidly involuting congenital hemangioma (RICH) is a recently recognized entity in which the vascular tumor is fully developed at birth and undergoes rapid involution. Angiographic findings in two infants with congenital hemangioma are reported and compared with a more common postnatal infantile hemangioma and a congenital infantile fibrosarcoma. Congenital hemangiomas differed from infantile hemangiomas angiographically by inhomogeneous parenchymal staining, large and irregular feeding arteries in disorganized patterns, arterial aneurysms, direct arteriovenous shunts, and intravascular thrombi. Both infants had clinical evidence of a high-output cardiac failure and intralesional bleeding. This congenital high-flow vascular tumor is difficult to distinguish angiographically from arteriovenous malformation and congenital infantile fibrosarcoma.

Keywords Infantile hemangioma · Congenital hemangioma · Rapidly involuting congenital hemangioma (RICH) · Arteriography · MRI · Congenital fibrosarcoma · Arteriovenous malformation (AVM) · GLUT-1

Introduction

Postnatal infantile hemangiomas are common benign vascular tumors [1]. Some deep hemangiomas may require imaging studies (particularly magnetic resonance imaging) to confirm the diagnosis; otherwise, most postnatal infantile hemangiomas are diagnosed based solely on the clinical presentation. Arteriography may be used for further evaluation in some indeterminate hypervascular lesions, and in some cases where a biopsy cannot be performed because of the high risk of bleeding. Postnatal infantile hemangioma appears angiographically as an organized, glandlike vascular neoplasm with vessels and a staining ''parenchymal'' component [2].

Rapidly involuting congenital hemangioma (RICH) is a rare endothelial cell neoplasm that differs from the common postnatal infantile hemangioma by its full-grown presentation at birth and accelerated involution [1, 3]. RICH is usually a single large tumor associated with lesional ulceration and congestive heart failure and can easily be confused with congenital infantile fibrosarcoma and arteriovenous malformation. Therefore, diagnosis of such a congenital vascular lesion usually requires diagnostic imaging studies including arteriography in order to exclude infantile fibrosarcoma and arteriovenous malformation. Biopsy may be indicated if there is suspicion of the former.

Fig. 1a–c Congenital hemangioma. Distal external carotid digital subtraction angiogram (lateral projection). Arterial phase (**a**) showing supply to the lesion from the superficial temporal and occipital arteries, as well as a small, anterior, deep temporal branch. The distal segments of the feeding arteries contain aneurysms (*arrows*) and there are direct arteriovenous fistulae. There are smoothly dilated early draining veins (typical for hemangioma), but a very poor parenchymal blush. Superselective injection (**b**) into one of the frontal branches of the right superficial temporal artery shows the aneurysm, which gives off some irregular distal arterial branches (*arrows*), as well as direct communication with draining veins (*large arrow*). Left distal external carotid artery injection (**c**) following embolization shows extensive shunting from the superficial temporal artery

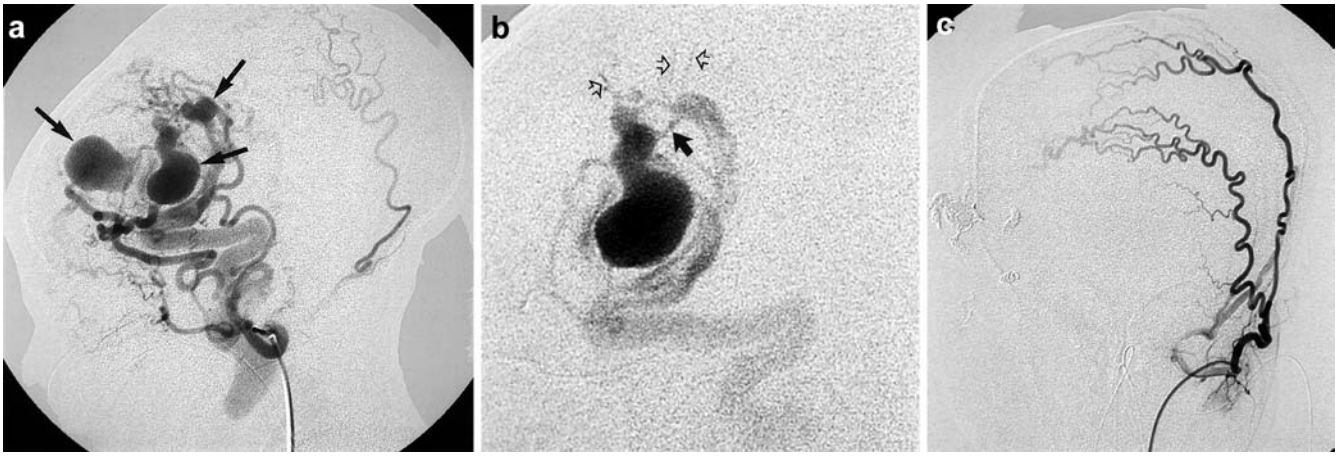
The angiographic features of RICH have not been described previously. The purpose of this study is to demonstrate the differentiating angiographic findings of RICH in comparison to congenital infantile fibrosarcoma and postnatal infantile hemangioma.

Case reports

The angiographic studies of four newborns with a histopathological diagnosis of RICH ($n=2$), congenital infantile fibrosarcoma ($n=1$) and a case of postnatal infantile hemangioma ($n=1$) were reviewed retrospectively. Both newborns RICH presented with a high-output cardiac state. Massive intralesional bleeding occurred in one of the newborns with RICH and in the infant with congenital fibrosarcoma. The histology of the resected specimens was reviewed.

Case 1

A large scalp mass was present at birth in a term male newborn (elective C-section, 38 weeks). The patient was hemodynamically stable at birth; however, moderate cardiomegaly was noted on a chest radiograph. Echocardiogram revealed right ventricular hypertrophy presumably related to high cardiac output. Computed tomography (CT) and magnetic resonance imaging (MRI) of the head showed no intracranial involvement. MRI showed an extracranial mass lesion with intense contrast enhancement and multiple various-sized flow voids. On day 5, spontaneous pulsatile bleeding occurred from the scalp mass, for which a ligation of the bleeding surface arteries was performed. Subsequently, angiography with embolization was performed, which was followed by steroid treatment (Fig. 1). A biopsy of the scalp mass was also performed during the angiographic procedure. This showed small and thin-walled vessels, as well as, large abnormal channels surrounded by fibrous stroma. Foci of hemosiderin were present. Characteristic lobules of capillaries were not observed, but this was attributed to sampling since in the clinical setting, the histopathology was consistent with RICH [4]. Immunostaining for glucose transporter-1 molecule was negative in the endothelium of the vascular channels. The embolization was carried out using microcatheters as a delivery catheter and various embolization materials, including platinum coils, polyvinyl alcohol (PVA) (1100 μm) and Gelfoam. The patient responded to treatment and the mass has significantly decreased in size during the first year. The patient is free of disease with a barely noticeable residual scar in the involved area.



Case 2

A large mass was noted at birth over the right posterior shoulder area in a term female newborn (37 weeks). The patient had mild congestive heart failure (CHF), thrombocytopenia, and elevated prothrombin time. CHF was managed by digoxin, fluid restriction, and oxygen. The initial diagnostic consideration was hemangioma and corticosteroid treatment was started (4 mg/kg). MRI was performed, which showed a large hypervascular heterogeneous mass with large vessels and patchy intense contrast enhancement (Fig. 2). The patient subsequently underwent angiography and particle embolization (Fig. 2), but died on day 13 because of an intracranial bleed. Angiographic findings included abnormal vessels consisting of focal expansion of the medium-sized arteries, followed by a plexus of small vessels, prior to drainage into dilated veins. A histopathologic study of the lesion was performed during the autopsy, which revealed both small and large lobules of capillaries with large centrilobular draining vessels and unusually massive interlobular draining and feeding vascular channels with frequent thrombi. The histopatology was within the spectrum of RICH.

Case 3

At birth, a 9.0×8.0×4.5-cm mass was found over the right scapula in a term male newborn following an uneventful vaginal delivery. There were ectatic veins over the surface of this mass. On the day after birth the patient's hematologic profile was consistent with consumptive coagulopathy. The tumor was presumed to be "hemangioma" with a Kasabach-Merritt phenomenon. Corticosteroid was started and continued for 2 weeks, but tapered since the mass did not shrink. Ultrasonography (US) and CT showed a hypervascular heterogenous mass with large-caliber vessels with some cystic changes centrally. Angiography and embolotherapy were performed because of severe bleeding (Fig. 3). Because the imaging studies were inconsistent with hemangioma, the lesion was subsequently resected surgically with partial removal of the latissimus dorsi muscle. On pathologic examination the tumor was found to be *congenital fibrosarcoma* with all of the margins free of tumor with the exception of 1 of 17 slides, which showed a microscopically positive resection margin. Metastatic workup was negative. About 10 days later the surgical wound was re-excised. The patient was free of disease 1 year later.

Case 4

A 14-month-old male, reportedly a product of a full term pregnancy, developed a large scalp mass lesion in the frontal region in the newborn period. After initial rapid growth, the lesion stabilized. He had a history of febrile seizures. No other medical conditions were reported. Arteriography was performed in order to clarify the diagnosis, demonstrate blood supply, and to perform preoperative embolization (Fig. 4). The angiographic features of the lesion were consistent with a classic hemangioma and no embolotherapy was carried out. The mass was subsequently excised and was shown to be a postnatal infantile hemangioma.

Discussion

Rapidly involuting congenital hemangioma (RICH) is considered to be a variant of the more common postnatal infantile hemangioma. It is fully developed at birth and rapidly involutes [1, 3, 4]. However, because of significant intratumoral arteriovenous shunting, newborns with RICH commonly present with high-output cardiac failure. Although imaging features of postnatal infantile

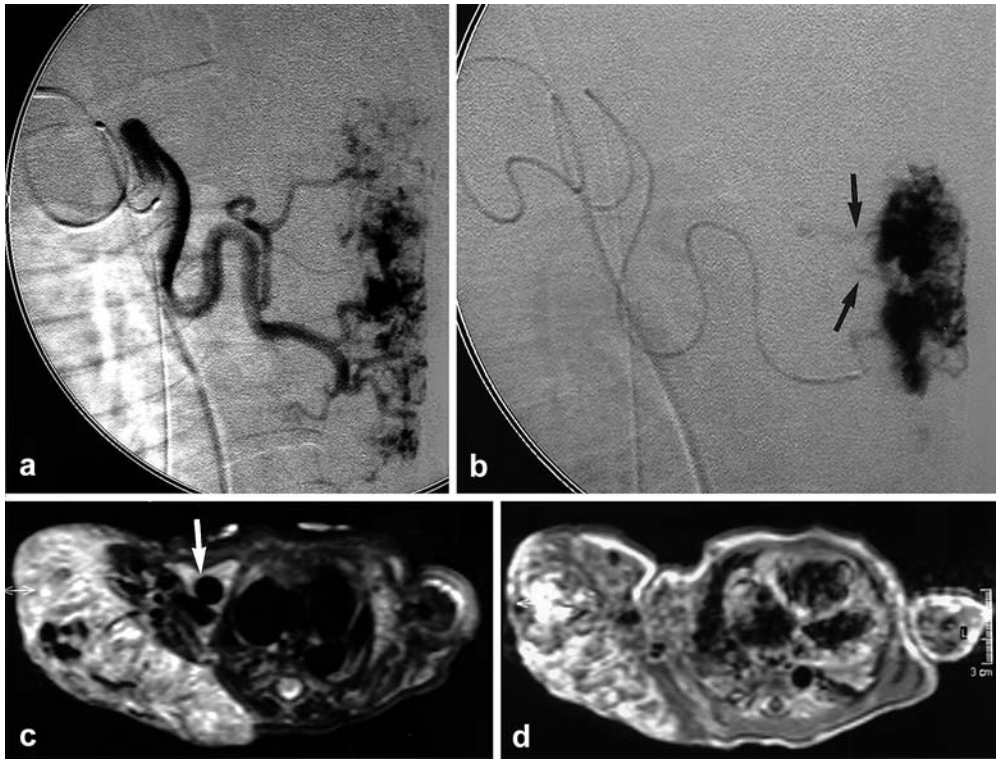


Fig. 2a–d Congenital hemangioma. Right subscapular artery digital subtraction arteriogram, lateral projection (**a**, **b**). The feeding arteries to the lesion show small, irregularly shaped aneurysms and multiple smaller feeding branches, but minimal parenchymal blush (**a**). Superselective injection into a branch of the subscapular artery (**b**) showing arterial aneurysms, distal feeding branches and early draining veins (*arrows*). T2 weighted axial image (**c**) showing a hyperintense lesion with large arterial and venous flow voids. Inhomogeneity of the mass may be related to the intratumoral vessels, which were found to contain some thrombi by histologic study. Note the markedly enlarged right axillary artery and vein (*arrow*). Post-contrast T1 weighted image (**d**) showing somewhat inhomogeneous enhancement of the tumor

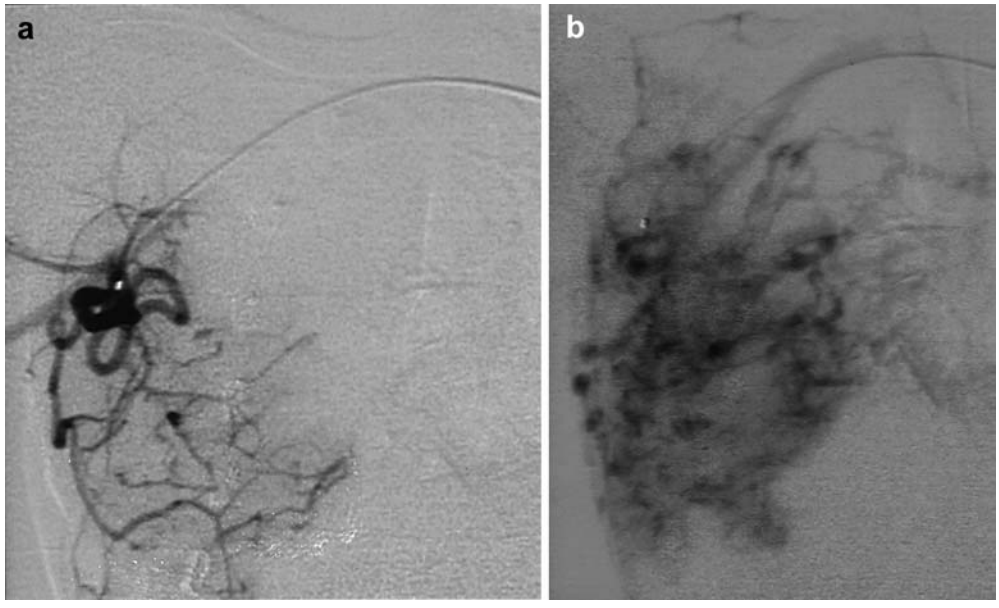


Fig. 3a, b Congenital fibrosarcoma. Selective angiography in the lateral thoracic branch of the right subclavian artery. Early arterial phase (**a**) showing the irregularly arranged feeding arteries with considerable tortuosity. The irregularity and tortuosity are consistent with a malignant tumor rather than a hemangioma. The venous phase (**b**) showing inhomogeneous parenchymal blush with drainage into multiple small irregular veins (Used with permission, W.B. Saunders Company)

hemangioma, including angiographic findings, have been well documented [2], this is not true for RICH. RICH has histologic differences from the typical infantile hemangioma of postnatal onset. RICH is characterized by relatively small lobules of capillaries surrounded by fibrous tissue. The central aspect of the lesion shows ''involution'' with connective tissue and few or no lobules. Thin-walled draining channels and large abnormal veins are prominent, particularly in the center of the lesion. Vascular thrombi, calcification, hemosiderin, and ''cyst'' formation may be encountered. The ''cysts'' are suspected to have been vascular aneurysms. Endothelial cells are typically negative for glucose transporter-1 protein (GLUT-1), whereas infantile hemangioma of postnatal onset is typically positive [4, 5].

A highly vascular mass in a newborn is suggestive of RICH. MRI and, if necessary, biopsy usually provides a definitive diagnosis. Ultrasonography with color or pulsed Doppler and MRI/MRA typically demonstrate fast-flow in the tumor vessels of RICH, which is a finding similar to that observed in postnatal infantile hemangioma [6, 7]. However, because of large intralesional cystic vascular abnormalities, RICH shows inhomogeneous contrast enhancement, whereas typical homogeneous enhancement is seen in postnatal infantile hemangioma. The radiological differentiation between postnatal infantile hemangioma and congenital infantile fibrosarcoma has been described in the literature. Postnatal infantile hemangioma typically shows homogeneous contrast enhancement, whereas congenital fibrosarcoma demonstrates inhomogeneous enhancement with a cystic architecture [8]. The differentiating imaging features of RICH from congenital fibrosarcoma have not yet been reported. However, both cases of

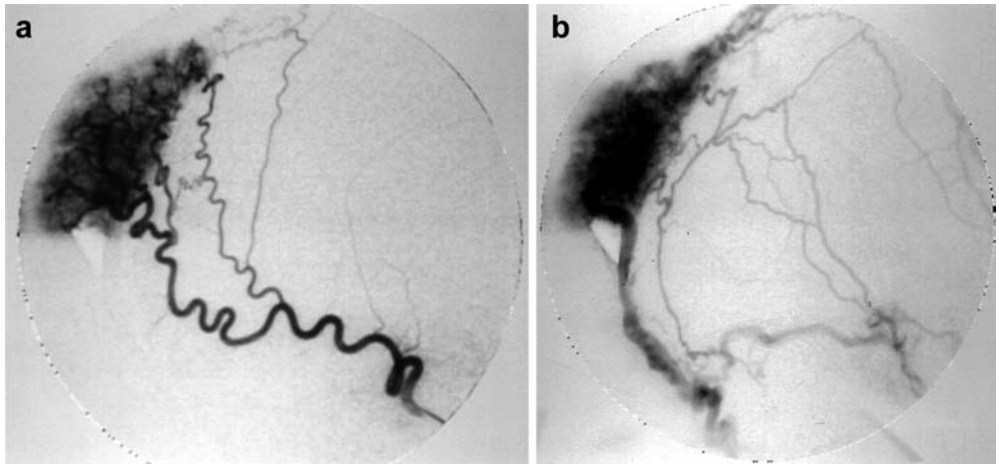


Fig. 4a, b Infantile hemangioma. Contrast injection in the distal external carotid artery (**a**) (lateral projection) showing prominent and tortuous feeding arteries with persistent parenchymal staining in a lobular pattern in the venous phase (**b**). A large draining vein is seen in the forehead

RICH reported herein showed intense but patchy contrast enhancement on MRI examination that was felt to be related to large space-occupying vasculature with intraluminal thrombi. A similar enhancement pattern is also expected in malignancies (e.g., fibrosarcoma).

Arteriography may be used in further evaluation of indeterminate hypervascular congenital masses. This can also be used in some cases where a biopsy cannot be performed due to the high risk of bleeding (either spontaneously or during the procedure) or if arteriovenous malformation is suspected. Embolization is usually indicated if there is associated cardiac failure, significant bleeding from the mass, or a need to minimize bleeding prior to surgery or biopsy. The angiographic findings of RICH, as well as the differentiation from congenital fibrosarcoma, also have not been reported in the literature. Although the differentiation of postnatal infantile hemangioma from congenital infantile fibrosarcoma is readily apparent on angiography, RICH may not be angiographically differentiated from congenital infantile fibrosarcoma or arteriovenous malformation (AVM). Congenital fibrosarcoma is fed by the arteries that resemble the classic tumor vessels of irregular caliber in a disorganized branching pattern [7] (Fig. 3). The capillary phase in a fibrosarcoma is seen as a dense but inhomogeneous tumor blush, while the venous phase exhibits a number of tortuous and mildly enlarged veins (Fig. 3b). Therefore, postnatal infantile hemangiomas can be easily differentiated by their persistent parenchymal staining in a lobular pattern and also by the smooth, enlarged regular branches of normal adjacent arteries around the tumor [2] (Fig. 4). RICH has unique angiographic findings, including inhomogeneous parenchymal staining, large and irregular feeding arteries in a disorganized pattern, multiple various sized aneurysms, direct arteriovenous shunts and intravascular thrombi (Figs. 1, 2). Some of these features can be found in malignant tumors, and others are also seen in AVM (Fig. 5). However, in the context of a mass in a newborn, the finding of partially thrombosed aneurysms and direct arteriovenous shunts strongly suggests RICH.

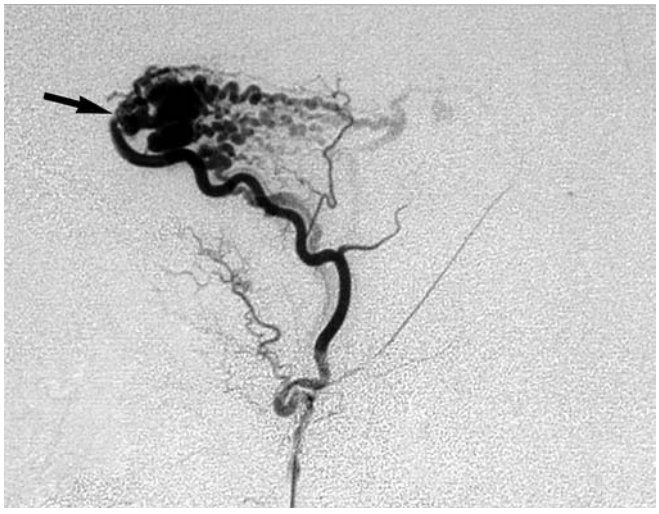


Fig. 5 Arteriovenous malformation (AVM) in the left temporal region in a 17-year-old male. Selective superficial temporal arteriogram (lateral projection) showing a somewhat prominent feeding artery (superficial temporal artery) with an arteriovenous fistula (*arrow*), small venous aneurysms and tortous draining veins. This vascular lesion was successfully treated with ethanol injections following coil embolization of the fistula

The following conclusion can be made: RICH has unique angiographic features that are distinct from those of postnatal infantile hemangioma but have some similarities to arteriovenous malformation or a malignant tumor. Biopsy may be necessary for definite diagnosis.

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