Cross-sectional imaging of primary osseous hemangiopericytoma

Abstract The aim of this study was to assess cross-sectional imaging features and the value of CT and MRI in primary hemangiopericytoma of bone. In five patients with histologically proven primary osseous hemangiopericytoma CT and MR scans were evaluated retrospectively. Both CT and MRI were available in four patients each. In three patients both imaging techniques were available. On CT primary hemangiopericytoma of bone presents as an expansive lytic lesion with bone destruction and inhomogeneous contrast enhancement. Magnetic resonance imaging depicts osseous hemangiopericytoma as hyperintense lesion on T2-weighted images with intermediate signal intensity on T1-weighted images. Curvilinear tubular structures of signal void in the tumor matrix on T1-weighted images and corresponding hyperintense structures on T2-weighted and on fat-suppressed short tau inversion recovery images were present in three patients. Although cross-sectional imaging findings are non-specific, they add to the diagnosis and provide valuable information about the extent of bone destruction and local tumor spread in patients with primary osseous hemangiopericytoma. While CT demonstrates the extent of bone destruction best, MRI better visualizes medullary and soft tissue extension of the tumor. Curvilinear signal abnormalities support the diagnosis of hemangiopericytoma of bone. This imaging pattern is best visualized on fat-suppressed or contrast enhanced T1-weighted MR images.

Keywords Hemangiopericytoma · Bone · MRI · CT

Introduction

As described by Stout and Murray in 1942, hemangiopericytoma is a rare benign or malignant tumor derived from the pericytes of Zimmermann [1]. The tumor may originate from the soft tissue as well as the skeletal system with a strong propensity to the soft tissue of the thigh, the pelvis, and the retroperitoneum. An intracranial origin of this entity, arising from the meninges, is also known. Osseous hemangiopericytoma most commonly derives from the pelvis or the femur, but any bone can be affected. Of 62 cases of primary osseous hemangiopericytoma reported in the literature, description of CT and MRI is limited to a small number of case reports. The imaging features described are usually nonspecific. The purpose of this retrospective study was to work out typical cross-sectional imaging features of primary osseous hemangiopericytoma.

Materials and methods

A retrospective analysis of five patients with primary hemangiopericytoma of bone examined in our hospital between 1988 and 2000 was performed. The diagnosis was based on operation (n = 3) or biopsy (n = 2). All specimens were reevaluated by two pathologists to confirm the diagnosis. There were four women and one man between 27 and 72 years of age. Both MRI and CT were available in four patients each. Three patients underwent both, CT and MR imaging. All MR examinations were performed with a 0.5-T Gyroscan T3 NT-scanner (Philips, Best, The Netherlands). The examination protocol included T1-weighted spin-echo sequences (TR 620 ms/TE 20 ms) and T2-weighted spin-echo (TR 3000 ms/TE...
120 ms) sequences. In two patients fat-suppressed short tau inversion recovery (STIR) sequences (TR 4400 ms/TE 40 ms/TI 100 ms) were available. In four patients 0.1 mmol gadolinium/kg body weight (Magnevist, Schering, Berlin, Germany) was administered followed by a post-contrast T1-weighted spin-echo sequence (TR 620 ms/TE 20 ms). The CT and MR scans were retrospectively assessed by two radiologists in consensus regarding CT and MR morphology, tumor extent, as well as MR signal characteristics of the depicted lesions. Reviewers were aware of histopathology.

Results

Tumor locations were the right iliac bone, the sacrum, and the femur. One patient showed a hemangiopericytoma of the pterygopalatine fossa. This patient had undergone surgical resection of the tumor 9 years before she was referred to our institution due to a local recurrence of the tumor. Four patients developed metastasis during follow-up. A history of previous intracranial meningeal tumor was excluded in all patients, as there is a well-known risk of misreading metastatic meningeal hemangiopericytoma as primary hemangiopericytoma of bone [2].

On CT hemangiopericytoma presented as a destructive, lytic lesion with cortical destruction in three patients. The growth pattern was more expansive than infiltrating. After i.v. administration of contrast agent the tumor was well delineated from the surrounding tissue and demonstrated a locally inhomogeneous contrast enhancement (Fig. 1). No calcifications were found within the tumor matrix. The intra- and extracerebral tumor extent was depicted on pre-contrast CT scans, but soft tissue extension was much better defined after administration of contrast agent.

On fat-suppressed STIR images, available in two patients, the tumor was depicted as a markedly hyperintense lesion (Fig. 1). All tumors were slightly hyperintense to muscle on T1-weighted images. In three tumors irregularly distributed hypointense areas were present on T1-weighted images. These hypointense areas included curvilinear structures of signal void on pre-contrast images with distinct contrast enhancement after intravenous administration of gadolinium-DTPA (Fig. 2). On T2-weighted images the tumors were hypointense with small hypointense regions (Fig. 3). Three examinations showed curved tubular structures of increased hyperintense signal intensity, correlating to the hypointense lines seen on T1-weighted images. After i.v. administration of contrast agent all tumors showed a distinct increase of signal intensity. In all patients the tumor margin was clearly delineated on MRI. Only minor perifocal edema was present. In three patients cortical breakthrough with soft tissue infiltration was depicted. In these patients the extraosseous tumor margin was better demarcated on MRI than CT. Imaging features of all five cases are summarized in Table 1.

Discussion

Hemangiopericytoma is an uncommon tumor arising from the pericytes of Zimmermann [1]. Due to its vascular origin, hemangiopericytoma can arise in any region of the body. Hemangiopericytoma usually occurs in the soft tissue but has also been described intracranially. Primary hemangiopericytoma of bone is very rare; more often soft tissue hemangiopericytoma invades bone. Osseous metastases of primary intracranial hemangiopericytoma have been described [2]. According to the Mayo Clinic series the incidence of primary osseous hemangiopericytoma is 0.1% of malignant primary bone tumors and 11% of malignant vascular bone tumors [3]. The patient age ranges from 12 to 90 years culminating in the fourth and fifth decade with a male-to-female ratio of 1:8:1 [4]. Histological features of osseous and soft tissue hemangiopericytoma are identical. Tumor cells typically cluster around numerous capillaries. They usually contain round to oval nuclei, generally lacking from anaplasia [4]. The tumor cells have reticulin sheaths and indistinct cytoplasmic borders. Small areas of necrosis, hemorrhage, or cystic degeneration have been described. Immunohistochemical staining is helpful in the differentiation against other malignancies [5, 6]. Differential diagnosis includes hemangioendothelioma, angiosarcoma, and atypical meningioma. Surgical resection is recommended as primary therapy of osseous hemangiopericytoma. The postoperative treatment options include radiation and chemotherapy [5]. As late recurrences are described a long-term follow-up is required.

Radiographic features are nonspecific, including lytic bone destruction (100%) and cortical destruction with soft tissue extension (72%). The margin can be well defined (40%) or ill defined (60%). In some cases a sclerotic rim is described (16%). Periosteal new bone formation is not uncommon (32%). Pathologic fractures are rare (8%) [4]. On CT cystic spaces, lobulations and speckled calcifications within the tumor were described with varying frequency. Contrast enhancement of solid areas and septations have been observed [7, 8]. Specific features of this highly vascularized tumor are only known for angiography. They include commonly displaced feeding arteries branching into small and large radially arranged vessels with a so-called spider-shape appearance. The capillary phase shows a well-defined, dense round or oval tumor stain. Draining veins are sometimes depicted in the late arterial phase, indicating arteriovenous shunts [9].

In our patients the extent of bone destruction, including cortical breakthrough, was demonstrated more clearly on CT scans than on radiographs or MR images. As the prediction of the clinical behavior of osseous hemangiopericytoma is unclear, this feature is important. Its high incidence underlines the assumption of