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Case report

Malignant haemangiopericytoma of the knee joint: MR findings

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Abstract. The imaging findings of intra-articular haemangiopericytomas have not been described previously. We report on a 35-year-old man with a primary malignant haemangiopericytoma of the left knee joint. MRI demonstrated joint effusion and a heterogeneous mass with haemorrhagic components and multiple peripheral serpentine signal voids, suggesting the presence of vascular channels. Multiple pulmonary metastases were seen on chest CT. These imaging findings, although not pathognomonic, might be useful in suggesting the diagnosis of intra-articular haemangiopericytoma.

Haemangiopericytomas are rare hypervascular tumours arising from the pericytes of Zimmerman [1–3]. These cells surround the capillaries and post-capillary venules [1–3]. Owing to the ubiquity of pericytes in mesenchymal tissues, haemangiopericytomas may occur anywhere in the body although the retroperitoneum, extremities, head and neck are more frequently involved [1, 4, 5]. The clinical course of haemangiopericytomas is unpredictable and an early pre-operative diagnosis is important [1, 2]. Joint haemangiopericytomas are extremely rare [6] and to our knowledge MRI findings of intra-articular haemangiopericytomas have not been described previously.

Case report

A 35-year-old man was admitted to hospital because of pain and swelling in the left knee joint. MRI examination had been performed 10 months previously because of a painless mass of the medial aspect of the left knee joint. After this first examination the patient had refused biopsy. During the present hospitalization, CT of the knee was performed and revealed a solid non-calcified mass at the medial aspect of the joint. Further MRI was performed and depicted an intra-articular mass associated with a large joint effusion. The mass exhibited heterogeneous high signal intensity on T_2 weighted images and intermediate signal intensity on T_1 weighted images (Figure 1). Following iv gadolinium-DTPA, the lesion demonstrated inhomogeneous enhancement (Figure 2). An area of high signal intensity observed on unenhanced T_1 weighted, T_2 weighted and fat suppressed T_2 weighted images was compatible with sub acute haemorrhage (Figure 1). Multiple serpentine signal voids, more prominent at the periphery of the upper pole of the mass and fewer within the mass, were observed on both T_1 and T_2 weighted images. This finding was better evaluated on T_2 weighted images and was thought to represent fast flow vessels (Figure 1b, c). Chest radiography followed by CT revealed multiple non-calcified pulmonary nodules compatible with metastatic lesions (Figure 3).

The intra-articular mass, while much smaller in size on the first MR examination, presented the same signal characteristics on the second MR examination.

At second admission, biopsy was performed and showed that the lesion was a typical malignant haemangiopericytoma. Histological examination revealed a soft tissue tumour consisting of thin-walled vessels lined by a single layer of spindle-shaped tumour endothelial cells. There was considerable branching of vessels showing a "stag-horn" appearance. The mitotic index was >5 per 10 high power fields, and many mitoses were atypical. A reticulum stain showed a dense reticulum network surrounding vessels and individual tumour cells. Immunohistochemical examination showed a weak positive staining of tumour cells in the vimentin. These cells were negative for smooth muscle actin, desmin, epithelial membrane

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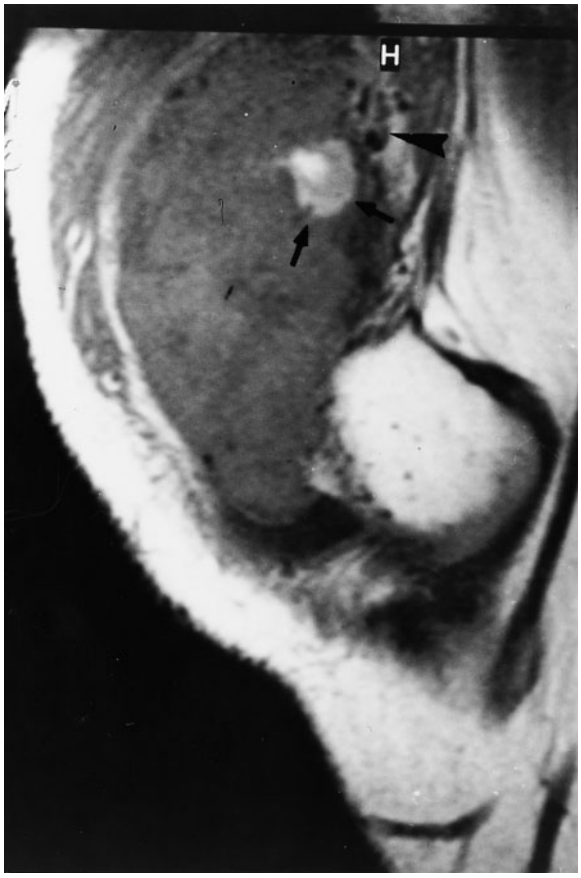
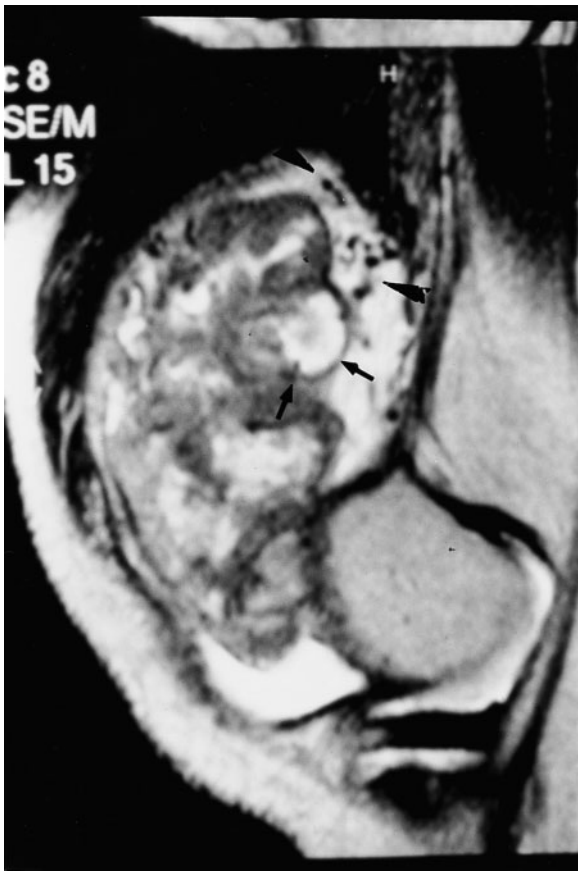


(a)



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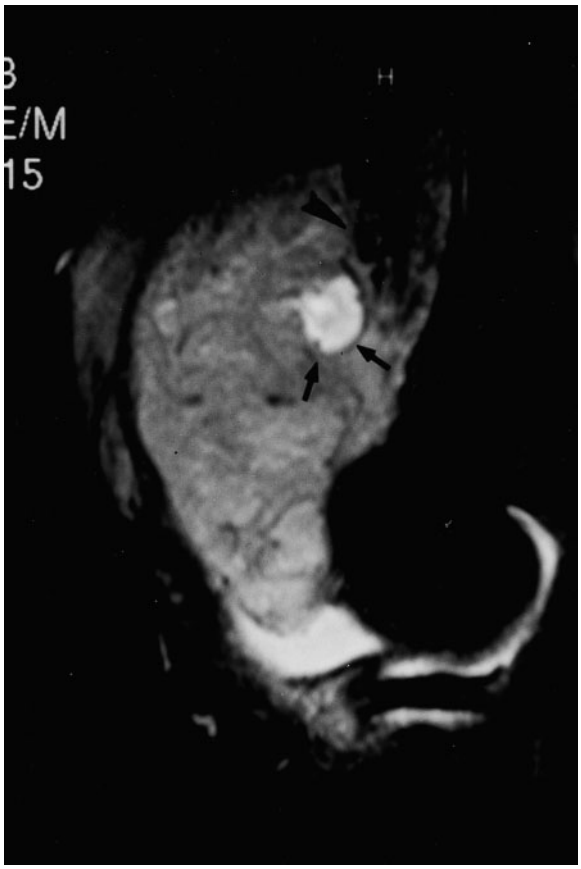

(c)

Figure 1. (a) T_1 weighted, (b) T_2 weighted and (c) fat suppressed T_2 weighted sagittal images of the left knee, demonstrating joint effusion and a large intra-articular heterogeneous mass with multiple peripheral serpentine signal voids (arrowheads). An area of high signal intensity on T_1, T_2 and fat suppressed T_2 weighted images represents sub acute haemorrhage (arrows).

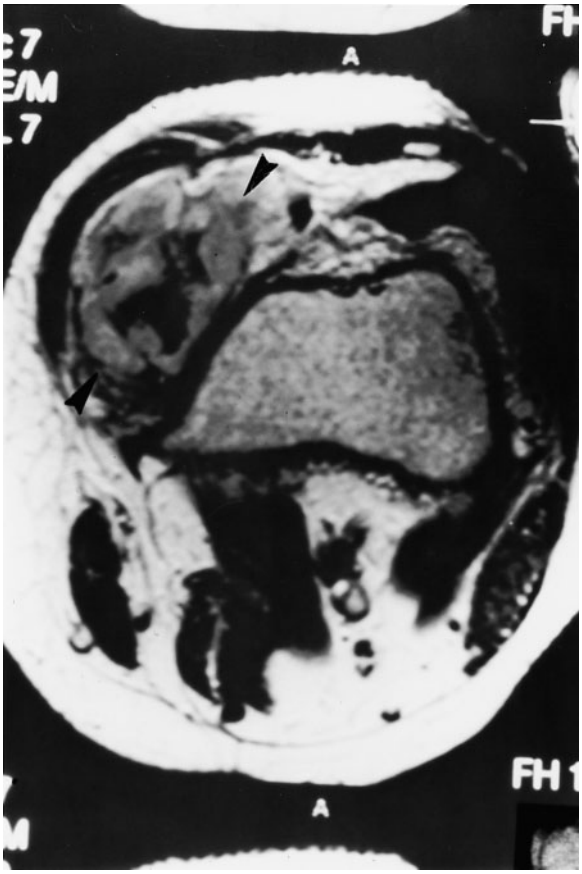

Figure 2. Contrast enhanced T_1 weighted axial image demonstrating heterogeneous enhancement of the tumour (arrowheads).

antigen, CD31, CD34 and factor VIII-related antigen and cytokeratins (Figure 4).

Discussion

Haemangiopericytomas are rare mesenchymal tumours that originate from the pericytes, which are contractile cells surrounding capillaries [1, 2]. Microscopically the tumour shows thin-walled vessels surrounded by tumour cells with indistinct

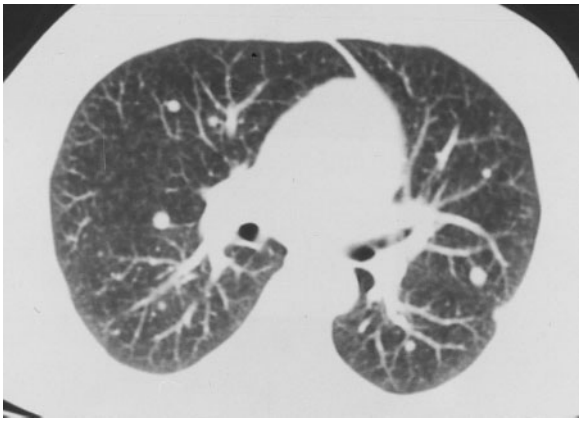

Figure 3. CT scan of the chest demonstrating multiple pulmonary metastatic lesions.

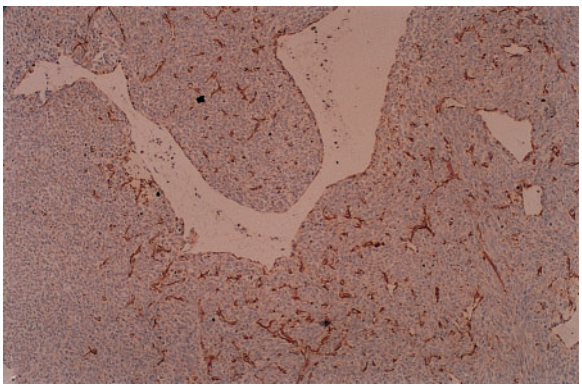

Figure 4. Haematoxylin and eosin histological section of the malignant haemangiopericytoma, showing multiple thin-walled blood vessels surrounded by neoplastic endothelial cells.

cytoplasm. Typical cases show "stag-horn" appearance of the dilated vascular channels. A reticular meshwork surrounds blood vessels and tumour cells [1, 2]. According to the biological activity, this tumour may be classified as benign, borderline malignant or malignant [2]. The incidence of malignancy differs between reports [1–8]. The diagnosis of malignancy rests on the presence of a high mitotic rate (>4 per 10 high power field), the greater degree of cellularity, pleomorphism of the tumour cells and the areas of haemorrhage and necrosis [1, 2]. Haemangiopericytomas most commonly occur in the fifth decade and present as slow growing, painless masses [3, 7]. Both sexes are equally involved [3, 7]. This tumour has been diagnosed in multiple areas of the body although the majority of reported cases were located in the retroperitoneum, extremities, head and neck [1–8]. There is one report describing only the histological findings of a synovial haemangiopericytoma arising in the knee joint [6]. The reported tumour presented benign histological behaviour while in our case the lesion was typical of a malignant haemangiopericytoma.

Malignant haemangiopericytomas present as mass lesions that are frequently complicated by haemorrhage and necrosis [1]. On MRI the lesion appears heterogeneous with high signal on T_2 weighted sequences and low to intermediate signal on T_1 weighted sequences, while diffuse and heterogeneous tumour enhancement occurs following contrast medium administration [4, 5, 8]. Areas of haemorrhage with high signal intensity on T_1 and T_2 weighted images have also been reported [5]. Another important MRI finding of haemangiopericytomas is the presence of serpentine signal void channels, more prominent at the periphery of the mass [4, 5, 8]. Haemangiopericytomas are hypervascular tumours and present specific angiographic features [3]. A few

feeder arteries enter the tumour from the periphery and branch into radially arranged vessels [3]. Serpentine signal voids on MRI represent these fast flow feeder vessels. In our case the mass was heterogeneous with a central area of sub acute haemorrhage and serpentine signal voids, more prominent at the periphery of the lesion.

Distant metastases of haemangiopericytoma have been described, mainly involving liver, lung and bone [5, 8]. In the present case pulmonary metastases were seen on chest radiography and CT. Speckled calcifications, previously described on CT of primary and metastatic malignant haemangiopericytomas [7], were not seen in our case.

Mass-like lesions and tumours affecting the joint should be included in the differential diagnosis of an intra-articular haemangiopericytoma of the knee. Intra-articular mass-like lesions may be seen in rheumatoid arthritis (RA) and pigmented villonodular synovitis (PVNS) [9, 10]. On MRI, areas of low signal intensity may be seen in both conditions and are explained by the presence of haemosiderin within the PVNS tissue and the pannus tissue of RA [9, 10]. However, such low signal intensity areas do not have a serpentine form and are mainly seen on gradient echo T_2 weighted images. Synovial thickening and intra-articular masses, of low signal intensity on both T_1 and T_2 weighted images, may also be seen in amyloid arthropathy [11]. However, these intra-articular masses are contiguous with subchondral bone lesions.

Synovial chondromatosis and synovial chondrosarcoma may present as intra-articular masses. Round areas of signal void may be seen within the masses and represent calcification and haemosiderin deposits [9, 12].

Synovial haemangiomas are rare intra-articular benign tumours almost always affecting the knee joint [9, 13]. Serpentine signal voids, located in the central and peripheral part of the lesion, are seen in 70% of cases [9]. Areas of low signal intensity, owing to haemosiderin laden tissue, and punctuate signal voids, owing to the presence of phleboliths, may be helpful in the differential diagnosis with haemangiopericytomas [9, 13].

Synovial sarcomas are malignant mesenchymal tumours. Only 10% develop within the joint cavity and the knee is the most common site [9]. On MRI the lesion appears as a heterogeneous mass containing areas of haemorrhage [9]. Fluid–fluid levels have been described in 10–25% of reported cases [9, 14].

In conclusion, haemangiopericytoma should be included in the differential diagnosis of intra-articular masses arising in the knee joint. MRI demonstrating a mass with peripherally located serpentine signal voids may be suggestive of the diagnosis.

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