Case 16442

Eurorad ••

Spindle cell haemangioma of the

foot

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ISSN: 1563-4086 Section: Musculoskeletal system Area of Interest: Musculoskeletal soft tissue Imaging Technique: MR Case Type: Clinical Cases Authors: Mats Van den broecke 1, David Creytens 2, Gwen Sys 3, Koenraad Verstraete 4 1 Universiteit Gent, BELGIUM 2 Professor, Ghent university hospital, department of pathology 3 Professor, Ghent university hospital, department of radiology orthopedic surgery 4 Professor, Ghent university hospital, department of radiology Patient: 35 years, male

Clinical History:

We report a case of a 35-year-old male patient who experienced a painless mass at the dorsal aspect of the second web space of the left forefoot for two years. The patient mentioned only a little discomfort when wearing narrow cycling shoes. Clinical investigation revealed an unspecific soft tissue mass at the dorsal aspect of the proximal phalanx of the second and third toe. There was no numbness and Tinel's sign was negative.

Imaging Findings:

Magnetic resonance imaging revealed a lobulated mass at the dorsum of the forefoot between the proximal phalanges of the second and third toe. [Fig. 1] The lesion contained tangles of tortuous blood vessels, no phleboliths were found. The mass appeared slightly hyperintense to muscle on T1 images and hyperintense on T2 images. Dynamic contrast-enhanced imaging showed strong enhancement during the first pass of contrast medium and a continuous further but slower enhancement in the second phase. [Fig. 2] Based on these imaging findings a vascular tumuor (haemangioma) was suspected and biopsy was performed.

Histological assessment revealed a relatively well-circumscribed vascular lesion composed of dilated thin-walled cavernous type vascular channels, lined by a thin layer of endothelium without cytonuclear atypia or multilayering. A little phlebolith was found in these vessels. More central in the lesion solid spindle cell areas with a slit-like growth pattern were observed.

Discussion:

Spindle cell haemangioma (SCH), also known as spindle cell haemangioendothelioma, is an uncommon vascular tumour that was first described in 1986 by Weiss and Enzinger. SCH was initially described as a type of haemangioendothelioma of intermediate or low-grade malignancy because the tumour was thought to have limited malignant potential and to have frequent local recurrence. [1] Since then various investigations have been conducted, including immunohistochemical studies, revealing that these lesions are benign. [2]

As in our case, this tumour is commonly detected in the superficial soft tissue of the distal parts of the extremities. The head, neck, chest and abdomen have also been reported as primary sites of origin in a minority of cases. They most often have an indolent course, but can become painful and disfiguring. It usually occurs in young adults and both sexes are equally affected. [1,2] There is usually no associated discoloration of the overlying skin making initial clinical suspicion of a vascular lesion difficult. [3]

SCH can present as a solitary lesion or can be multifocal. Multifocal SCH's have been associated with Maffucci syndrome, Ollier disease, lymphedema, Klippel Trenaunay syndrome and early onset varicose veins. [2]

On magnetic resonance imaging haemangiomas are typically well-defined, lobulated and heterogeneous with no features of local invasion. On T1 the overall signal is often intermediate to slightly higher than muscle and high-signal intensity tends to dominate on T2-weighted images. The lesion shows marked enhancement after contrast medium administration.

The differential diagnosis of SCH includes other types of (cavernous and capillary) haemangiomas, Kaposi's sarcoma, kaposiform haemangioendothelioma and intravascular papillary endothelial hyperplasia. The combination of SCH's cavernous spaces with organised thrombi, plump endothelial cells, multitude of spindle cells and minimal mitotic activity can histologically differentiate it from the other diagnoses. [2] There are also differences in the immunohistochemical staining patterns, including human herpesvirus 8 (HHV-8) positivity in Kaposi's sarcoma, which is negative in SCH. [4]

Preoperative planning should include an MRI to guide biopsy and to assess involvement of deeper structures. [2]

The standard treatment is wide surgical excision. Limited excisions almost certainly reoccur, but there is no consensus on the incidence of reoccurrence. When deciding how to treat each individual case, the slow growth of the tumour and the morbidity that the surgery may cause must be taken into consideration. [2] In our case local surgical excision was performed and no recurrence was reported until 9 months after surgery.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Spindle cell haemangioma

Final Diagnosis: Spindle cell haemangioma

References:

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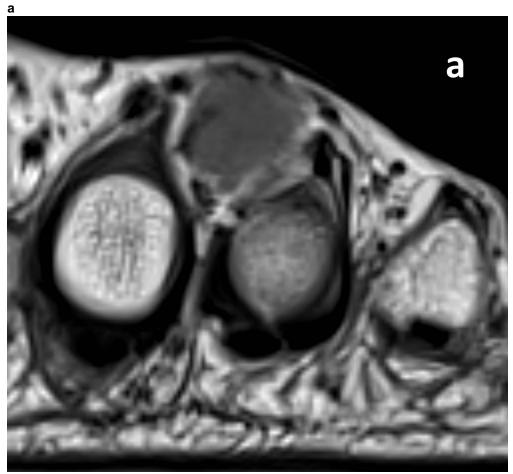
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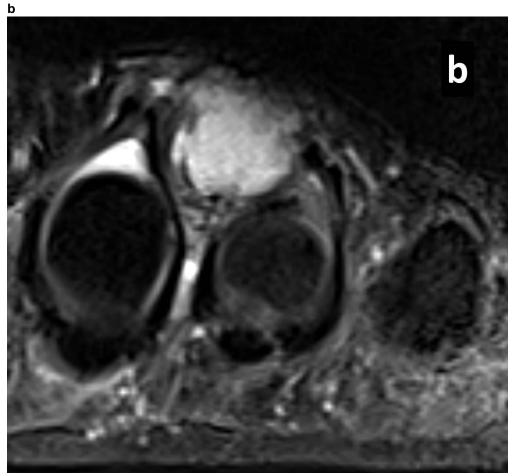
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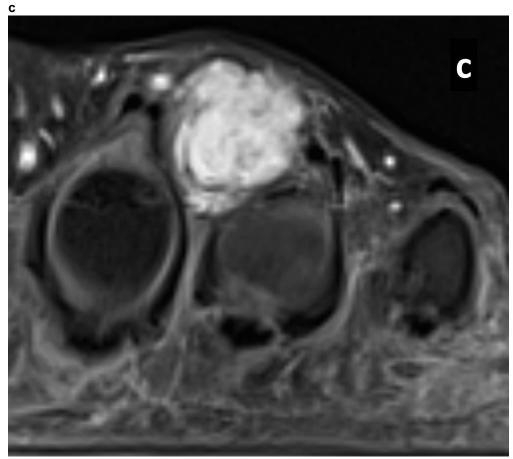
Figure 1



Description: Transverse (a,b,c) and coronal (d,e,f) T1, T2 FAT SAT and T1 FAT SAT after gadolinium administration: sharply delineated, slightly lobulated oval lesion with flow voids (black arrow) from high flow vessels traversing the tumour. Signal intensity is intermediate on T1 (a little higher than muscle *), high on T2 and high after gadolinium administration. **Origin:** Department of Radiology, Ghent University Hospital, Medical University of Ghent/ Belgium 2018.



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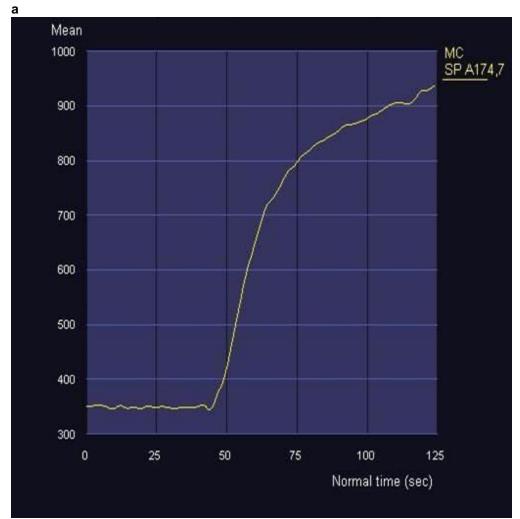


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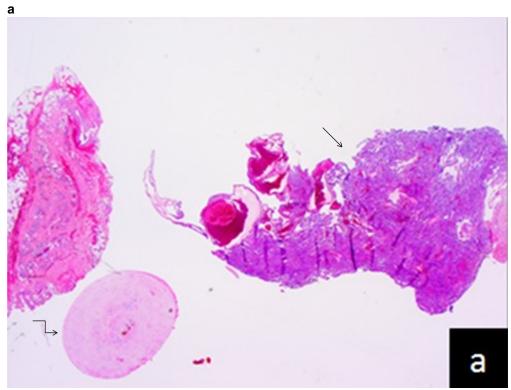
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Figure 2



Description: Time-intensity curve of the lesion shows steep first pass, indicating high vascularity and perfusion, followed by continuous further but slower increase, indicating large interstitial space. **Origin:** Department of Radiology, Ghent University Hospital, Medical University of Ghent/ Belgium 2018.

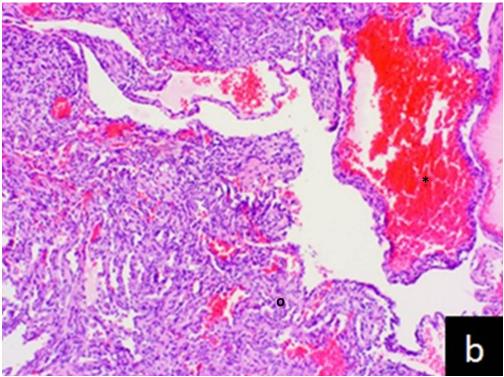
Figure 3



Description: Haematoxylin-eosin stain overview (Fig 3a) demonstrates the vascular lesion (arrow) on the right side and a little phlebolith (hooked arrow) on the left side.

Detail 100x (Fig 3b) shows the dilated thin-walled cavernous type vascular channels (*) on the right side and the central solid spindle cell areas (o) with a slit-like growth pattern on the left side. **Origin:** Department of Pathology, Ghent University Hospital, Medical University of Ghent/ Belgium 2018.





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