Sclerosing soft tissue haemangioma in an adult patient: an atypical form of a common entity causing the diagnostic dilemma

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DESCRIPTION

A man in his mid-30s presented to our hospital with the complaints of swelling and dull aching pain in the first web space of his left hand (graphic diagram, figure 1) for the last 1 month. The patient first noticed the swelling 5 years earlier, when it was the size of a grain. The swelling gradually increased in size until it became the size of an almond. On examination the swelling was palpable in the first web space and had a soft to firm consistency. Ultrasonography revealed a well-defined, elongated and mixed echogenic lesion (20×10 mm in size) in the first web space in the subfascial and subcutaneous planes. The lesion showed internal vascularity on the colour Doppler examination (figure 2A,B) and abutted the underlying muscles of the first web space. MRI examination revealed a well-defined soft tissue lesion that appeared isointense to surrounding muscles on T1-weighted image and hyperintense on T2-weighted/proton density fat suppressed images (figure 2C,D). The lesion was abutting the first dorsal interosseous muscle. Based on

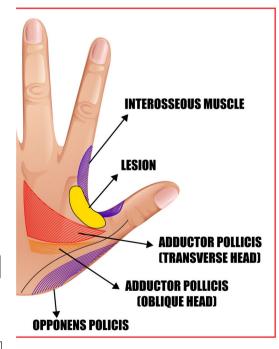


Figure 1 Graphic computed image of the dorsum of the left hand demonstrates location of the lesion with respect to surrounding structures (the figure has been created by the authors).

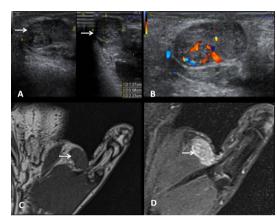


Figure 2 Ultrasonography still images show (A) an elongated, lobulated, well-defined, heteroechoic lesion (20×10 mm in size) in the first web space involving the subfascial and subcutaneous planes (white arrow). (B) Colour Doppler examination of the lesion demonstrates internal colour flow signals that suggest internal vascularity. (C) T1-weighted MRI shows an isointense lesion in first web space of the left hand involving subfascial and subcutaneous planes (white arrow). (D) In PDFS image, the lesion is hyperintense (white arrow) and abutting the first dorsal interosseous muscle (the figures have been created by the authors). PDFS, proton density fat suppressed; USG, ultrasonography.

these findings, differential diagnoses of benign neoplastic lesions such as vascular malformations (haemangioma), nerve sheath tumour and lipomatous lesions were suggested. The patient underwent a local excision and the mass was sent for a histopathological examination (HPE) (figure 3A,B). The HPE demonstrated multiple dense whirls of hyalinised stroma obliterating the endothelium-lined proliferated vascular channels, suggestive of sclerosing variety of haemangioma (figure 3C,D). The patient recovered well without any recurrence.

Soft tissue haemangiomas are benign angiomatous lesions and represent up to 7% of all benign soft tissue tumours. Haemangiomas are more commonly found in children, and are termed vascular tumours. Presentation in the adult age group is highly unusual. Haemangiomas are classified as benign, locally aggressive and malignant lesions according to the International Society for the Study of Vascular Anomalies classification. The patient typically describes the lesion as an indistinct mass that becomes larger or smaller



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Images in...

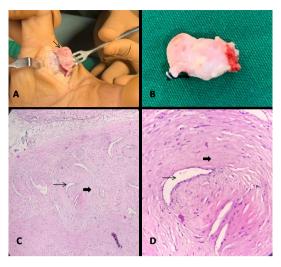


Figure 3 (A,B) Intraoperative and gross specimen images showing an almond-sized lesion excised from first web space of the hand. (C,D) Histopathology images with $10\times$ and $40\times$ magnifications show multiple dense whirls of hyalinised stroma obliterating the endothelium-lined proliferated vascular channels, suggestive of sclerosing variety of haemangioma (the figures have been created by the authors).

with time. Deep-seated haemangiomas cannot be distinguished from malignant soft tissue tumours without imaging and histopathological studies. Different types of typical haemangiomas have been described, which may demonstrate unique radiological manifestations; however, in many cases, the imaging characteristics are insufficient in reaching a diagnosis. Haemangiomas have a proliferation and then a degeneration phase. First, there is a rapid growth of endothelial cells that form syncytial masses with or without increased blood flow, and this phase is linked to high angiogenic factor levels such as vascular endothelial growth factors and basic fibroblast growth factors. This is followed by an end stage, also called the involution stage, in which complete sclerosis or hyalinisation of the haemangioma matrix occurs.²³ These sclerosed haemangiomas are mostly asymptomatic and often incidentally found. Radiological investigations play an important role in diagnosing haemangiomas, as these lesions have typical imaging features. On USG, these appear as hypoechoic lesions with or without vascular flow and as T1/T2-weighted iso/hyperintense lesions on MRI; however, rarely atypical morphologies might also be detected.^{4 5} The process of sclerosis changes the radiological characteristics, making their diagnosis nearly impossible by using imaging characteristics only. This may warrant surgical/pathological correlation.

Patient's perspective

I was having this swelling in my hand for a long while, it was increasing in size and sometimes it was painful. I came to the hospital and doctors helped me a lot. They could not reach the exact diagnosis, however they operated on it very nicely and sent that excised swelling for testing and told me that it was some non-malignant lesion which gave me a big relief.

Learning points

- ► Reduced fat and increased hyalinised material in atypical haemangiomas create diagnostic difficulties in ultrasonography, as this entity mimics nerve sheath tumours and other benign soft tissue neoplastic lesions.
- ► A sclerosing phenomenon is responsible for the change in the radiological appearance of soft tissue haemangioma, and this needs confirmation by histopathological examination.
- MRI is the choice of investigation for soft tissue lesions, as MRI has inherent high tissue contrast resolution properties. We highlight post-resection histopathological examination for sclerosing haemangiomas as they mimic other benign neoplasms.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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