ANGIOKERATOMA OF THE WRIST

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ABSTRACT

A case of angiokeratoma was presented. The patient had a red-to-purple soft tissue mass about 1 cm size at volar aspect of the right wrist that gradually enlarged with time to be 9x8x7 cm. Multi-modality imagings were performed and revealed a large, fat-containing mass with multifocal small soft-tissue densities within the fat. The presumptive diagnosis was hemangioma. Angiographic study revealed nearly complete occlusion of the interosseous (feeding) artery with collateral circulation filled its distal part subsequently. This pattern is not common in hemangioma. The histologic diagnosis was angiokeratoma which although being a benign vascular tumor of skin, need wide surgical excision margin to prevent local recurrence. Careful observation of uncommon imaging findings in the case presumptively diagnosed as hemangioma will help to raise the possibility of other entities.

INTRODUCTION

Angiokeratoma is a benign vascular tumor of skin which may be presenting in various characteristics in the body.[1,2] It usually arises as a soft, compressible, pink to red papules 1-3 mm in diameter. With the passage of time, many lesions develop keratotic changes, become darker, and present a warty appearance.[3] Because angiokeratoma is usually asymptomatic and small, with average size of 3 mm,[1,4] imaging investigation and surgical intervention are rarely performed. We presented a case of angiokeratoma of the wrist which was large and diagnosed as angiosarsoma by clinical findings and as hemangioma by imaging findings.

CASE REPORT

In January 1995, a 39-year-old Thai woman came to the Hand clinic because she had a right wrist mass. This mass has appeared since birth as a nodule of 1 cm size involving the skin of volar aspect of right wrist, showing dark red coloration with lobulated contour. At that time surrounding skin was normal. This lesion slowly enlarged with time, and multiple small red-black dots lately developed in the surrounding skin. In some instances, she had associated pain and low-grade fever. She had normal range of motion of the right wrist and fingers. She did not have any relatives with this disease. Physical examination revealed a married, female agriculturer with normal consciousness. She was not pale, and did not have icteric sclera. Heart, lungs, and abdominal examinations were normal. Extremity examination disclosed a lobulated mass of 9x8x7 cm in size, soft consistency, with red-purple coloration involving the volar aspect of the right wrist. Multiple small red-black nodules are observed on

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the surrounding skin (Fig. 1). The radial and ulna pulses could not be palpated. The motor and sensory functions were intact. The circumferences of the right wrist was 21 cm whereas of the left was 15 cm. The presumptive diagnosis was hemangioma with suggested malignant transformation (angiosarcoma). Plain radiograph revealed soft tissue swelling at the volar aspect of right distal forearm all through the wrist and palm. Large area of fat density was seen in the lesion. No bone involvement was demonstrated (Fig. 2). Incisional biopsy was performed, yielding a result of angiokeratoma. Computerized tomography (CT scan) (Fig. 3) and magnetic resonance imaging (MRI) (Fig. 4) revealed that this lesion was fat-containing, with multiple small soft tissue densities distributing within the fat. Multifoci of skin thickening were observed. Bony structure appeared normal. Contrast-enhanced CT scan showed minimal enhancement of the soft-tissue component of the lesion. Right brachial angiography was subsequently performed (Fig. 5). In arterial phase; there was nearly complete occlusion of the interosseous (feeding) artery at the level of mid forearm with collateral circulation filling its distal part. The distal part of radial artery was displaced laterally by the mass. No significant arterial abnormality was seen at level of the wrist. In capillary phase; there were diffuse, multiple small contrast medium retentions which remained opacified late into the venous phase. The angiographic diagnosis was hemangioma involving soft tissue of the right wrist. Surgical exploration revealed that the mass could be easily removed from underlying muscles and tendons except at the wrist where a part of the mass involved the flexor retinaculum resulting in this structure having to be resected. The wound defect was covered with split thickness skin graft (STSG) harvested from left thigh. Post operative conditions were satisfactory. Follow-up at 1, 2, and 3 months revealed normal functions and range of motion of the right wrist and fingers. No evidence of local recurrence is detected.

Left **Right**

Fig.1 Preoperative photograph of the lesion shows an ill-defined mass with red-purple coloration involving the volar aspect of the right wrist. Multiple small red-black nodules are observed in the surrounding skin.

Fig. 2 Oblique plain radiograph of right distal forearm including hand reveals soft tissue swelling in the volar aspect of the wrist with fat density distributing throughout. No bone destruction is observed.

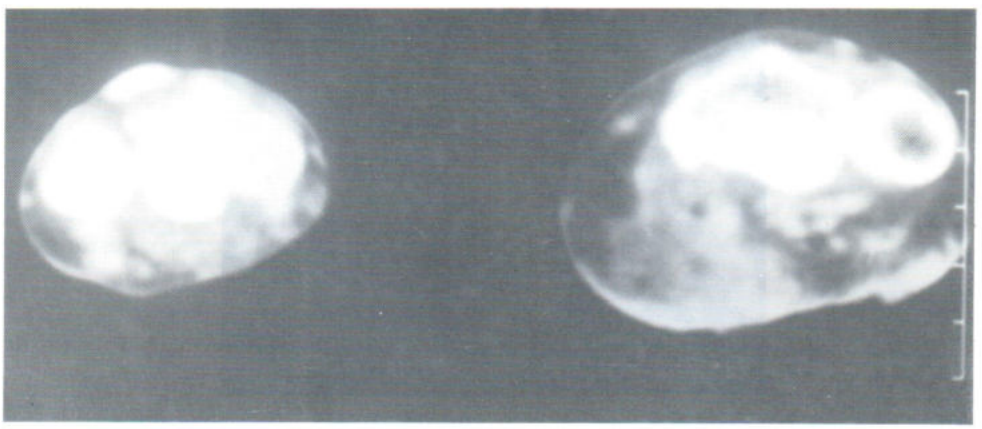
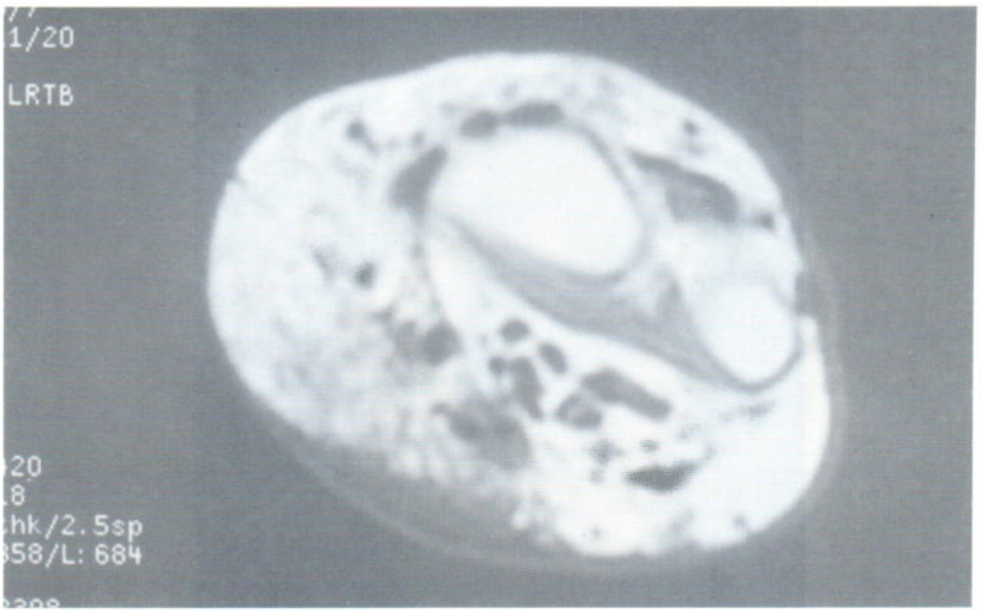


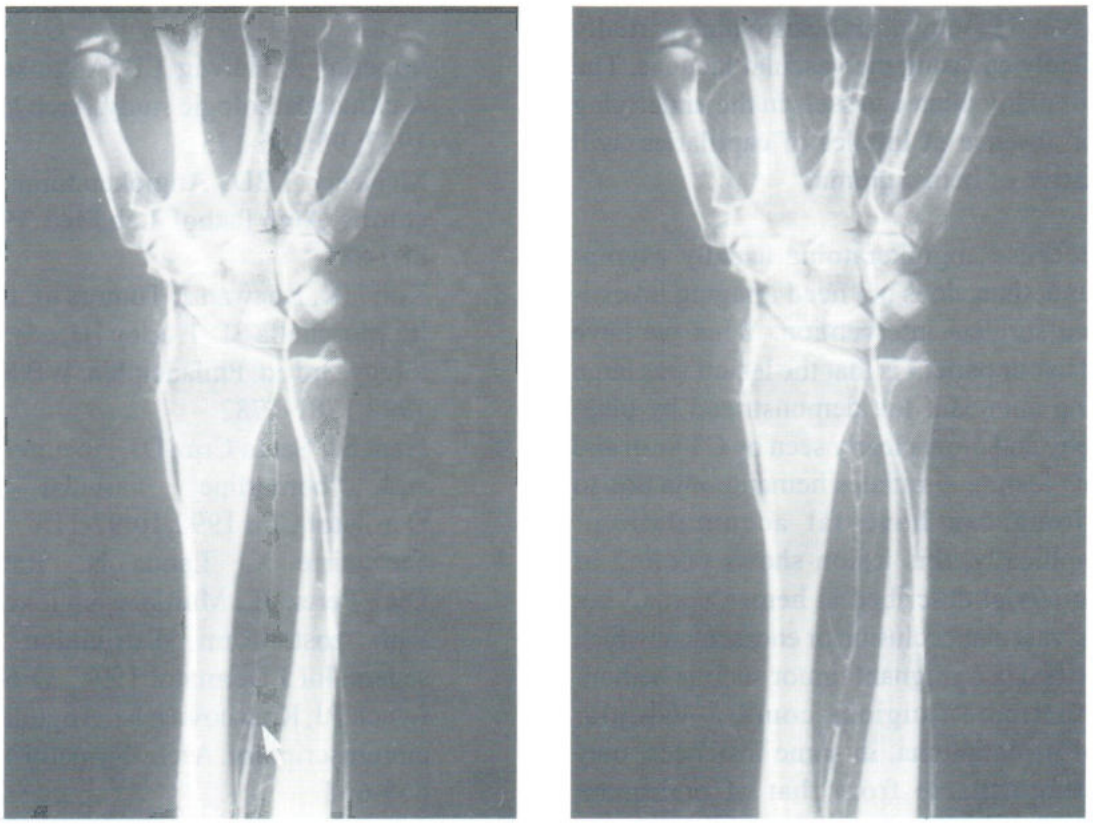
Fig. 3 Axial CT scan demonstrates that the lesion has a large amount of fat, with multiple small soft tissue densities distributing within the fat. Skin thickening is noted.



4A

4B

Fig. 4 a) Axial and b) coronal T1-weighted MR images exhibit the fat to insinuate between the normal-appearing tendons. Some small hypointensities are seen within the fat.



5A 5B

Fig. 5 Angiographic study discloses nearly complete occlusion of the interosseous artery (arrow in Fig. 5 a) which was filled subsequently by the collateral vessels (Fig. 5 b)

DISCUSSION

Five clinical forms of angiokeratoma are generally recognized:[1,3-5] 1) the Mibelli type, occurring as hyperkeratotic lesions on the hand and feet,[4] particularly on the dorsum of fingers and toes,[5] 2) the Fordyce type, involving the scrotum and vulva, 3) angiokeratoma corporis diffusum, associated typically with Fabry's disease, and also with fucosidosis,[4] 4) angiokeratoma circum-scriptum, is present at birth or early childhood, and females are affected three times more commonly than males. The process is almost always unilateral.[3] On histopathologic examination, it is a true hemangioma,[5,6] 5) solitary or multiple papular angiokeratomas which may locate in any part of the body[5] but favors the lower extremity.[1,3,4] Histopathologically, it has the criteria of a true angiokeratoma.[5]

Because angiokeratoma appears as variably hyperkeratotic lesions with coloration ranging from pink-red to purple, brown, or black; differential diagnosis can include hemangioma, nevus, verruca, and melanoma.[1,4] Although usually asymptomatic, angiokeratoma may become irritated and bleed, perhaps as the result of local trauma.[4]

Histopathologic features suggesting the diagnosis of angiokeratoma are combination of marked vascular dilatation of the papillary

vessels, forming large lacunae in the papillary area of dermis; and having acanthosis which partially or completely encasing the vascular lacunae. The absence of many dilated vessels in the underlying dermis or absence of lobules of capillaries, will be suggestive of hemangioma.[1]

Because angiokeratoma usually asymptomatic and, thus, does not need imaging investigation and surgical intervention. What we have observed in our patient is that the lesion was large and having abundant fat, demonstrated by plain radiography and more clearly seen by CT scan and MRI. This feature simulates hemangioma due to hamangioma can have fat accumulation.[7] Angiographically, this lesion shows pooling of contrast material described in hemangioma,[8] but exhibiting vascular occlusion or encasement which was described in malignant tumor such as hemangioendothelioma (angiosarcoma).[9] Vascular pattern of hemangioma, in some instances, may be indistinguishable from that of malignant tumor.[8,10] The diagnosis can be made only by histology as in this case. Differentiation of angiokeratoma from hemangioma is important because angiokeratoma needs wider excision margin to prevent recurrence whereas hemangioma can be treated by local excision.[10] In our case, nearly complete occlusion of the feeding artery demonstrated angiographically is uncommon in hemangioma, and raise the possibility of other disease.

In summary, from this present case, angiokeratoma is a benign vascular lesion of skin that may enlarge with time, and may simulate hemangioma in multi-modality imaging findings. Careful observation of uncommon imaging findings in the case presumptively diagnosed as hemangioma may help to raise the possibility of other entities. Correct diagnosis obtained by histologic examination is of importance for proper surgical treatment.

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