CASE REPORT

A rare tumour of hand: angioleiomyoma

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SUMMARY

Angioleiomyoma is a benign tumour composed of smooth muscle and vascular tissue. Because of the paucity of smooth muscles in the hand other than tunica media of the blood vessels, its occurrence is quite rare in the hand and only few cases are reported in the English literature. We present the case of a 49-year-old man with benign painless swelling on the dorsum of hand. Differential diagnosis of ganglion cyst and tendon sheath tumour were considered. However, excision biopsy revealed angioleiomyoma. At 2-year follow-up, the patient remained asymptomatic with no evidence of recurrence.

BACKGROUND

Angioleiomyoma is a rare benign tumour composed of smooth muscles and vascular tissue. They are also called vascular leiomyoma, angiomyoma or dermal angioma.¹ According to the WHO soft tissue tumour classification, they are classified in pericytic (perivascular) benign tumour group.² They are common in middle-aged women with preponderance to occur in lower limbs. They are small, slow-growing, firm, occasionally painful swelling occurring most commonly in legs with no characteristic clinico-radiological findings. They are uncommon in hand as there is paucity of smooth muscles in the hand other than tunica media of the blood vessels. And hence diagnosing angioleiomyoma preoperatively in the hand is difficult.³

CASE PRESENTATION

A 49-year-old right hand dominant man with no pre-existing comorbidities presented with a swelling on the dorsum of the right hand which was insidious in onset and gradual in progression since 5 years. It was associated with mild pain. There were no similar swellings in other parts of the body. On examination, the swelling was 3 cm×2 cm over the dorsum of the hand in the second web space. It was ovoid in shape, non-tender, firm in consistency with smooth surface and well-defined edges. It was mobile horizontally and was in the subcutaneous plane.

INVESTIGATIONS

Radiographic examination revealed soft tissue swelling without bony changes. Ultrasound showed hypoechoic solid lesion in subcutaneous plane in the second web space measuring 2.8 cm×1.3 cm, showing internal vascularity abutting adjacent bone, muscle and tendons (figure 1).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of ganglion cyst, tendon sheath tumour, schwannoma, neurofibroma and fibrolipomatous hamartoma were considered (table 1).

TREATMENT

Patient underwent excision biopsy. The mass was well encapsulated and did not involve the extensor tendons and digital nerves. Small branches of the vessels were seen entering the tumour mass (figure 2). Grossly the specimen appeared as a single nodular mass of flattened grey-white tissue, weighing about 9 g and measuring about 3 cm×2 cm (figure 3).

OUTCOME AND FOLLOW-UP

Histopathological examination revealed a tumour composed of spindle cells arranged haphazardly, interspersed with thick-walled and thin-walled blood vessels. The cells showed vesicular cigar-shaped/fusiform nuclei and were seen to merge with the outer smooth muscle layer of blood vessels. Interspersed collagen, few lymphocytes and foci of retiform lymphatics were also seen (figure 4). At 2-year follow-up there was no evidence of recurrence and patient was asymptomatic.

DISCUSSION

Angioleiomyoma constitutes less than 1% of all soft tissue tumours in the upper limb.⁴ They are common in the third to sixth decades with a male to female ratio of about 1:1.7.⁵ Most commonly they are distributed in the appendicular skeleton (89%) predominantly in lower limbs (75%).⁵ Pain is

Figure 1 Ultrasound image showing the tumour (TUM), tendon (TEN) and adjacent metacarpal (MC) bone.

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Table 1 Differentials for benign painless swellings of the hand

Disease	Salient clinical features	MRI findings	Histopathology
Ganglion cyst	Most common on dorsum of the wrist Firm, smooth fluctuant Arising from the synovium of the joint or the tendon sheaths	Focal, fluid cystic mass (T2W axial section)	Myxoid wall, benign fibroblastic collagen No epithelial synovial lining
Tendon sheath tumour	More commonly on the lateral three rays of the hand (middle, index and thumb) Benign swelling Firm lobulated mass	Well-defined mass adjacent to or enveloping a tendon Signal intensity similar to or less than that of skeletal muscle (T1W images) Low signal intensity with variable degree of heterogeneity (T2W images)	Spindle cells, fibrous tissue Cholesterol-laden histiocytes Multinucleated giant cells Hemosiderin
Schwannoma	Rarely seen in hand, eccentrically or centrally located Minimally tender Moves at right angle to the course of the nerve	Isointense or slightly hypointense to muscle (on T1W images) and hyperintense (on T2W images) Usually a homogenous internal pattern is evident	Hypercellular (Antoni A) cells—arranged in palisades, composed of plump and fusiform nuclei (Verocay bodies) Hypocellular (Antoni B) cells—non-uniform cells in a myxomatous matrix
Neurofibroma	Solitary or multiple, centrally located, non-tender, nodular, non-encapsulated Involving skin and nerves	Homogeneous mass Isointense signal in T1W sequences Hyperintense signal in T2W sequences	Plexiform mass of irregular, thickened nerve fibres separated by increased endoneurial matrix
Fibrolipomatous hamartoma	Rare benign tumour Presenting up to third decade Swelling in the wrist or digits Involving the median/digital nerves	Characteristic low intensity 'serpentine nerve bundles' embedded in abundant hyperintense adipose material, with fine fibrous tissue septa coursing along the nerve (T1W and T2W images) Also characteristic 'cable-like' appearance (on axial T1W images)	Fibro-adipose tissue infiltrating the epineurium They separate and compress the fascicles

T1W, T1 weighted; T2W, T2 weighted.

a presenting feature in half of the patients which is often paroxysmal and aggravated by physical stimulation.

Aetiology about the origin of these tumours is unclear.⁴ Hormonal changes particularly oestrogen, repetitive micro trauma and venous stasis have been proposed as causative factors.⁶ Some authors have considered these lesions as proliferation of smooth muscles in haemangioma or as hamartomas.^{5,7}

Histologically, they can be classified into three types (table 2) as per Morimoto.⁸ Majority of these tumours are in the extremities which are mainly solid type and often painful. Remaining are seen in the axial skeleton and are usually of venous type and painless. Immunohistological markers will be positive for smooth muscles (small muscle actin (SMA)) and vascular tissue markers (CD31 and CD34).⁹ Areas of calcification, myxoid

degeneration, hyalinisation, haemorrhage, nerve fibres and fat cells may be seen.^{5,6} Presence of nerve fibres and contraction of the smooth muscles which constricts the vessels lead to ischaemia and pain in more than half of the patients.

Ultrasound imaging shows well-defined margins with homogenous structure.¹ MRI studies show slightly hyperintense round or oval mass when compared with skeletal muscle in T1-weighted images. Mixed hyperintense and isointense regions when compared with skeletal muscles are seen in T2-weighted images, which correspond to smooth muscle and connective

Figure 2 Intraoperative photograph showing well-encapsulated tumour with small vessels entering the tumour mass (black arrow mark).

Figure 3 Gross (A) and cut section of the specimen (B) showing homogenous grey-white tissue and focal grey-brown areas.

Figure 4 (A) Photograph of the microscopic appearance showing (black arrow marks) uniform smooth muscles (B) with slit-like vascular spaces (H&E, x10 and x40).

Table 2 Histological classification of angioleiomyoma (Morimoto)⁸

Type	Characteristic features
Solid type (66%)	Common in women Abundant compact mature smooth muscle cells Absent mitotic figures Few slit-like vascular channels
Venous type (23%)	Common in men Smooth muscular wall in the vascular channels
Cavernous type (11%)	Common in men Abundant dilated vascular channels Very less smooth muscles

tissue, respectively.¹⁰ Peripheral rim shows hypointense signals on T2-weighted images corresponding to fibrous capsule.

Angioleiomyoma is a benign tumour which is seen rarely in the hand. More than half of them are painful. It should be considered in the differential diagnosis of any benign nodular swelling in the hand. Due to lack of tumour-specific clinical and radiological findings, it is difficult to diagnose this lesion preoperatively, but MRI findings may be suggestive at times. Excision of the swelling is the treatment of choice as it is both diagnostic and

Learning points

- ▶ Angioleiomyoma is a benign tumour which is seen rarely in the hand, of which more than half are painful.
- ▶ It should be considered in the differential diagnosis of any benign nodular swelling in the hand.
- ▶ Though clinically preoperative diagnosis is difficult, radiological findings can be suggestive.
- ▶ Excision of the swelling is the treatment of choice and recurrences are very less.

curative. Very few cases of malignant transformation to vascular leiomyosarcoma have been reported.^{11 12} The risk of recurrence is very less with complete excision.

Contributors MSK was responsible for the study concept and design; carried out the literature search. SV and MN were responsible for initial proof reading. SV, MN and SKR critically reviewed the manuscript for important intellectual content. SKR was the study supervisor. All authors read and approved the final version of the manuscript.

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