Angioleiomyoma: a clinical, pathological and radiological review

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SUMMARY

Angioleiomyoma is a benign tumour arising from the vascular smooth muscle (tunica media) and presents commonly between third and fifth decades of life. Although there are sporadic reports about this tumour in the literature, none describes all the information in detail. This review is an attempt to collate all the facts in one concise article. Angioleiomyoma presents as a painful mass in approximately 60% of the cases. One of the distinct clinical feature noted is the increase in size of the swelling with physical activity of the involved part, especially in the hand. It should be considered in the differential diagnosis of painful nodular lesions of the extremity. Pre-operative diagnosis is difficult, but with a high index of suspicion and awareness, it is possible. The use of ultrasound and magnetic resonance imaging should be considered. It causes minimal morbidity and excision is usually curative. Histological examination using smooth muscle Actin stain portraits the smooth muscle bundles clearly.

Introduction

Angioleiomyomas are benign solitary smooth muscle tumours that occur uncommonly in the hand. They are also known as angiomyoma, vascular leiomyoma or dermal angioma. The peak incidence is between the third and sixth decades of life and has a female preponderance. This tumour is rarely diagnosed before surgery. The usual treatment is excision of the mass and at times ligation of feeder vessel. Although the occurrence of these tumours is thought to be uncommon in the hand, we have treated three patients with angioleiomyoma in their hands within a short span of time. We present four cases here along with a concise review of clinical, pathological and radiological features.

Angioleiomyoma is a rare, benign smooth muscle tumour that originates in the tunica media of veins. It can occur anywhere in the body and can be found in the dermis, subcutaneous fat and fascia. This tumour causes pain in approximately 60% of the patients and occurs most commonly on an extremity, particularly the lower leg.

Incidence

Angioleiomyomas account for 5% of all benign neoplasms of soft tissues (16). The prevalence of angioleiomyoma in East Africa is 10 times that of a Caucasian population living in temperate climates.

Aetiology

Minor trauma (7–10), venous stasis and hormonal changes especially that of oestrogen have been proposed as aetiological features. The presence of chronic inflammatory cell infiltrates (1,2) in some lesions support the venous stasis theory.

Genetics

A few karyotypic abnormalities like 6p, 13q, 21q rearrangements and translocation (t(X; 10) (q22; q23.2) have also been described in the literature (13, 14).

It has been suggested that angioleiomyomas may vary from angioma to a solid leiomyoma depending on the amount of smooth muscle proliferation (12). Another theory suggests that they could be hamartomatous lesions as demonstrated by the presence of mature fat cells (1) (12).

Pathology

In 1973, Morimoto (4) studied 241 cases of angioleiomyoma and classified them into three histological types:

1 Solid: the most common type which has closely compacted smooth muscle and many small, slit-like vascular channels. These type of tumours are three times more common in females than in males.
2 Venous: thick, easily identifiable muscular walls distinguish this type. These tumours occur more commonly in males.
3 Cavernous: the vascular channels are dilated with less smooth muscles. This is the least common type of the three. These tumours occur four times more common in males.
Morimoto (4) also grouped these tumours into two groups:
1 The larger group of extremity tumours, where the tumours are mainly of the solid type and often painful. The lower extremity tumours are twice as common in female patients.
2 The smaller group of head tumours, where the tumours are usually of the venous type and painless. These tumours are more common in males.

Histological examination demonstrates smooth muscle bundles and the vascular channels surrounded by a thin capsule (1,2,11,12). Myxomatous and hyaline degeneration were documented by a few authors (1,2,12) who feel that the changes could be due to circulatory disturbance. Organising thrombus (1,2), mature fat cells (1,11) and lymphocytic infiltrate (1,2) was found in some tumours.

Fine needle aspiration features of angioleiomyoma were not sufficient to permit histotype diagnosis. Cytological specimens showed uniform spindle cells admixed with smooth muscles, collagen, macrophages and fat cells (11).

Imaging

On ultrasound examination, angioleiomyoma shows well-defined margins and a homogenous structure, suggesting benign nature of the lesion. Colour Doppler examination shows high resistance in intratumour arteries, suggesting the presence of muscular arteries (16).

Magnetic resonance imaging (MRI) features of angioleiomyoma are well documented (16–18). T2-weighted MR images showed mixed areas that were hyperintense and isointense to skeletal muscle (Figures 4 and 5). Hyperintense areas on T2-weighted MR images showed strong enhancement after intravenous contrast injection. Isointense areas on T2-weighted MR images did not show enhancement after intravenous contrast.

Hyperintense areas on T2-weighted MR images correspond to the smooth muscle bundle cells and isointense areas on T2-weighted MR images correlate to the tough fibrous tissue or intravascular thrombi. MR images cannot differentiate between different histological subtypes of angioleiomyoma.

For an extremity mass with mixed areas of hyper and isointensity to skeletal muscle on T2-weighted MR images and with a hypointense rim, a diagnosis of vascular leiomyoma should be considered (18).

Clinical Features

The typical lesion is a solitary, small, slow-growing, firm, mobile, subcutaneous nodule. Majority of these tumours measure <2 cm in size (1,3). Freedman et al. (3) documented significant decrease in the size of the lesion with increasing age.

Pain is the most striking clinical feature of angioleiomyoma (58% in Hachisuga et al. (1) series and 62% of patients from Mayo clinic). Pain is often paroxysmal and is provoked by exposure to cold and wind. It is thought be due to the active contraction of smooth muscle resulting in local ischaemia. Rest appears to be a relieving factor. We found that in our cases swelling increased in size after physical activity. It is probably due to accumulation of blood in the vascular spaces which in turn stretches the capsule and nerve endings (Figures 1–3).

Examination may show compressibility with pain on compression. Small vascular channels of solid type tumours are more prone for compression (explaining frequency of painful
lesions, 70% compared to 30% of cavernous and 37% of venous in Hachisuga et al. (1) series.

Angioleiomyoma is uncommon in hand, but it is interesting to note that we have three cases involving the hand in our series. These tumours tend to occur most often at the base of the fingers near the neurovascular bundle and usually not seen distal to the DIP joint. This is useful in differentiating from the glomus tumour (35).

CASE REPORTS

Case 1

A 39-year-old, right-handed engineer noticed a swelling on the left hand ulnar border at the base of the little finger approximately for 6 months. The swelling had grown slowly and was painful when he knocked it on objects. He also had mechanical interference with his work which was disabling. There was no known history of injury. Examination showed a 1.5 cm swelling on the ulnar border of the neck of the fifth metacarpal. The skin was free, and the swelling was soft, cystic, not tender and mobile. Grip strength was normal and equal. The provisional clinical diagnosis was either a ganglion or an implantation dermoid.

This pale grey coloured swelling was excised under local anaesthetic in total. There was no feeder vessel found. The histology proved it to be an angioleiomyoma.

Case 2

A 39-year-old male presented with a painful swelling of the left palm of one year duration, which increased in size after exercise of one year duration. Examination showed soft, cystic, tender swelling over the distal palmar crease at the metacarpophalangeal joint level measuring approximately 1 cm. This was excised in total under General Anaesthetic with ligation of vascular pedicle. Histology of the swelling confirmed angioleiomyoma, and patient was symptom-free 1 year after surgery.

Case 3

A 30-year-old male computer programmer presented with a painful swelling of the left palm of 18 months duration. The swelling gradually increased in size. Preoperative diagnosis of ganglion was performed, and the swelling was excised. Histological examination showed angioleiomyoma.

Case 4

A 51-year-old lady presented with a lump on plantar aspect of right foot of 18 months duration. This interfered with her walking and was painful. She was weightbearing more on the lateral border of the foot. The swelling was tender, freely mobile and was superficial to the plantar fascia. Excision biopsy confirmed the diagnosis. Patient has recovered well, although the scar is still tender (Table 1).

DISCUSSION

Angioleiomyoma can be found throughout the body which may arise in the dermis, subcutaneous fat, fascia or bone. It should be distinguished from all nodular lesions of the extremity like lipoma, ganglion, fibroma, schwannoma, haemangioma, foreign body granuloma, pseudoaneurysm, inclusion cyst, giant cell tumour of tendon sheath and glomus tumour (3, 18, 23). Rare cases of tumours arising from sweat glands such as aggressive, digital papillary adenocarcinoma should be borne in mind.

The histogenesis is still debated. Duhig and Ayer (12) suggest that proliferation of smooth muscle within a haemangioma produces a vascular leiomyoma and that further proliferation produces a simple leiomyoma. Most authors agree
that (1,4,9) vascular leiomyoma arises from veins. These lesions may be hamartomas. Duhig and Ayer (12) also suggest that an appreciable number of angioleiomyomas are not true tumours but are instances of vascular malformations.

The histology shows bundles of mature smooth muscle cells orientated around blood vessels. The deep soft tissue tumours, which are often solid, show similar features. In addition, marked degenerative changes with hyalinisation, myxoid changes and calcification are seen.

Hasegawa et al. (15) after clinicopathological and immunohistochemical study of angioleiomyomas suggested that the pain in these tumours may be mediated by the nerve fibres located within the tumour parenchyma. Small nerve fibres (immunoreactive for S-100 protein and PGP 9.5) were identified within the stroma of 69% of painful tumours.

Angioleiomyoma is rarely diagnosed preoperatively (2). An unusual case of (24) Epstein–Barr virus-associated multiple subcutaneous angioleiomyomas was reported in a patient with acquired immunodeficiency syndrome.

Angioleiomyoma is considered as a soft tissue tumour, but angioleiomyoma of the finger extending in a horsehoe-shaped fashion between the flexor tendons and proximal phalanx causing bone erosion has also been reported (27).

Angioleiomyomas account for 5–12% of hand tumours (1,13,28). Angioleiomyomas of the upper extremity are more common in males but are less painful. Calle et al. (23) reported a case of angioleiomyoma arising from a digital artery in the hand and reviewed a number of these tumours reported in the English literature. So far three cases of angioleiomyoma of the hand arising from the digital artery were reported (23). One of our cases had feeding vessel probably arising from a digital artery.

Radial nerve palsy (29) and Sciatic nerve dysfunction due to mechanical compression by an angioleiomyoma have been reported (30). Intra-osseous angioleiomyoma was postulated to arise from smooth muscle cells of vessel walls in the bone (19,20), and so far, nine cases have been documented in the literature (22).

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