Dendritic fibromyxolipoma adherent to the median nerve in the forearm

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Dendritic fibromyxolipoma adherent to the median nerve in the forearm

Case report

Short running title: Dendritic fibromyxolipoma and median nerve

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Abstract

We describe a 65-year-old woman with a tumour adherent to the median nerve in the left forearm that was found to be a dendritic fibromyxolipoma, a distinctive benign soft tissue lesion possibly related to a myxoid spindle cell lipoma; it is a solitary fibrous tumour that may be mistaken for a sarcoma. The tumour was successfully excised with no complications.

Key words: Median nerve; Dendritic fibromyxolipoma; Myxoid spindle cell lipoma; Lipoma; Mesenchymal tumour

Introduction

Benign mesenchymal tumours from the main group of lipomas have many different variants. The clinicopathological features of such lesions have recently been described under the name of “dendritic fibromyxolipoma” [1, 2].

This lesion shares many clinical and pathological features of myxoid spindle cell lipoma, solitary fibrous tumour with myxoid change, and angiomyxolipoma [3-5]. It is characterised by a mixture of spindle and stellate cells, mature adipose tissue, and abundant myxoid stroma with prominent collagenisation. It may be confused with a low grade myxoid liposarcoma [2]. Suster et al. described 12 cases, particularly in men, in whom the tumours were located in the neck, shoulder, upper back, chest wall, or nasal area. Dendritic fibromyxolipomas did not recur, were well circumscribed, and simple excision was curative in all patients [2, 3]. In one case the tumour was mistaken for a schwannoma, but the dendritic fibromyxolipoma has spindle and stellate cells that stain strongly for CD34 and bcl-2 but not for S-100 protein or for epithelial and muscle cell markers [1, 6]. Extremely few extrathoracic solitary fibrous tumours reported by - for example - Hasegawa et al. [4] have been located in the forearm [4]. We report a 65-year-old patient with a dendritic fibromyxolipoma that was topographically connected to the median nerve in the forearm.

Case report

A 65-year-old woman, who had hypertension treated with propranolol (Atenolol®) but was otherwise healthy, was referred to the Department of Hand Surgery, Malmö University Hospital, Sweden, with a 3 x 4 cm tumour in the left volar forearm. The tumour was soft

and not adherent to the skin. Sensation in the hand and arm was normal and she had no paraesthesia on percussion of the tumour. There was no atrophy of the muscles and the range of movement was normal. We suspected that the tumour was a lipoma. At exploration there was a well-circumscribed tumour that was connected to the ulnar side of the median nerve, but there was no distinctive small fascicle approaching the tumour as is seen in schwannoma. At exploration one could see superficial fascicles in the uninjured median nerve (Figure 1) covered by epineurial and mesoneurial tissue. The wound healed uneventfully and she had no complications after the exploration. The median nerve functioned normally postoperatively.

Materials and methods

Tissue specimens were fixed in 10 % neutral-buffered formalin. Representative sections of the lesion were cut, processed routinely, and embedded in paraffin. Sections for microscopy were cut at 4-6 μm and stained with haematoxylin and Eosin, Giemsa and van Gieson stains. Representative sections were also studied immunohistochemically using an automatic immunostaining system, Benchmark® XT, with a standard protocol (Ventana Medical Systems, Illkirch, France), using the following antibodies: ACTSM (Neomarkers, Fremont, USA; RB-9010-P), bcl-2 (Ventana Medical Systems, Illkirch, France, 26-4240, prediluted), CD34 (Ventana Medical Systems, Illkirch, France, 760-2927, prediluted), desmin (Dako, Glostrup, Denmark, M0724, 1:50), Ki67 (Dako, Glostrup, Denmark, M7240, 1:50), S-100 (Dako, Glostrup, Denmark, Z0311, 1:2000), and vimentin (Ventana Medical Systems, Illkirch, France, 790-2917, prediluted).

Pathological findings

The tumour measured 20 x 32 x 10 mm and was sharply demarcated with a polycyclic outer contour and a smooth surface. It was rather soft with slightly lobulated cut surfaces and contained yellowish adipose tissue with gelatinous myxoid areas. Histologically it consisted of mature adipose tissue with fibrous and myxoid areas containing spindle-shaped and dendritic stromal cells. The tumour tissue was divided into smaller compartments by fibrous, collagen-rich bands, assuming a lobulated or multinodular architecture (Figure 2a). Scattered round cells and mast cells were present throughout the lesion. The adipose tissue component consisted of mature, normal-looking fat cells. No lipoblasts were seen. The lesion was well-vascularised with normal-looking small and medium-sized vessels. No vascular component as seen in angiomyxolipoma or myxoid liposarcoma was found. The myxoid areas showed a fibrillary and vacuolated intercellular substance that was strongly basophil with haematoxylin and eosin (Figure 2b), and showed a distinctive metachromatic reaction with Giemsa stain (Figure 2c). Dendritic stromal cells were common, lying in a small lake of mucinous material and surrounded by a clear halo, mimicking an empty lacuna (Figure 2c). In other areas the mucinous substance formed larger, confluent masses embedding many stromal cells, each lying in an empty lacuna of its own (Figure 2d).

Both spindle-shaped and dendritic stromal cells were strongly immunoreactive to CD34 (Figure 2e) and vimentin (Figure 2f), but did not stain for S-100 or smooth muscle markers. There was little or no immunostaining for Ki67, indicating low proliferative activity. Bcl-2 staining was seen in mast cells and also in spindle cells in the stroma. However, the dendritic stromal cells did not invariably stain for bcl-2. In conclusion the tumour resembled a dendritic fibromyxolipoma.

Discussion

We have described a 65-year-old woman with a six-year history of a slowly-growing tumour in the left distal forearm, in which macroscopic findings at exploration showed a tumour closely adherent to the median nerve but with no fascicles approaching the tumour. The microscopic finding showed a dendritic fibromyxolipoma. Such tumours are extremely rare in the extremity, and are mainly located in the neck, shoulder, and back regions [1-4, 7]. The spindle-like cells in the tumour stained for CD34 but not for S-100, which is typical for dendritic fibromyxolipoma [2]. Initially there was a suspicion that the tumour was a schwannoma, but at exploration we could see no single fascicle going into the tumour, which showed no microscopic features of schwannoma that stains for CD34 in 15% of cases [6]. The tumour showed no morphological characteristics of either nerve sheath myxoma or malignant peripheral nerve sheath tumour [6], the latter tumour not staining at all for CD34 [6]. It did not stain immunocytochemically against Ki67, which indicated a low proliferative tendency. There were no mitoses in the tumour cells and no cytological atypia, indicating that the tumour was benign. It lacked the vascular component of angiomyxoma where spindle stromal cells, in addition, fail to stain for CD34 [5]. Spindle cell lipoma and solitary fibrous tumours both stain for CD34 in stromal cells and may have a myxoid component, but lack the dendritic cells typical of a dendritic myxofibrolipoma [3, 4, 6]. Bcl-2 was observed in mast cells and in stromal spindle cells, but the dendritic stromal cells invariably stained for bcl-2, which is a marker for apoptosis.

After exploration the superficially exposed fascicles of the median nerve were covered with epineurial and meseoneurial tissue. The patient had no symptoms or impaired sensation after exploration, which is usually the case after excision of a benign

schwannoma [8]. The tumour was easily excised, which is not the case for a granular cell tumour that may occur in the median or the ulnar nerve [9]. The dendritic fibromyxolipoma in our case had a maximum size of around 3 cm, which is in the range of previously reported tumours from other parts of the body but they mainly occur in middle-aged to elderly men [1]. Previous reports have shown that simple excision is curative in all patients with maximum follow-up of 13 years [2]. Our patient had had no recurrence after three years.

In conclusion, we report a patient with a dendritic fibromyxolipoma that strongly adhered to the left median nerve in the forearm, which to our knowledge has not previously been described. Excision caused no sensory disturbances and seemed to be curative, but the patient is being followed up regularly.

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Figure legend

Figure 1. At exploration there was a circumscribed tumour on the ulnar side of the median nerve (thick arrows in a and b), where the tumour was clearly and closely connected to the epineurium and mesoneurium (thin arrows in b and c) of the median nerve. No minor fascicles approached the tumour, but there were superficial fascicles in the median nerve (star in c), which could be covered by epineurial and mesoneurial tissue (thin arrows).

Figure 2. Low power view of the tumour with a mixture of adipose and myxoid tissue and fibrous collagen-rich bands in a lobulated arrangement (a, haematoxylin and eosin, original magnification x2); Detail showing myxoid area with basophil mucinous material (b, haematoxylin and eosin, original magnification x10); Dendritic stromal cells lying in empty-looking lacuna and surrounded by mucinous material (c, Giemsa stain; original magnification x40); Confluent mass of mucinous substance with many stromal cells embedded, each lying in an empty-looking lacunae of its own (d, Giemsa stain; original magnification x20); CD34-stained stromal cells, visualising their slender cytoplasmic processes, but small blood vessels also showed endothelial immunoreaction (e; original magnification x20) and Vimentin-stained stromal and fat cells (f; original magnification x40).

Figure 1

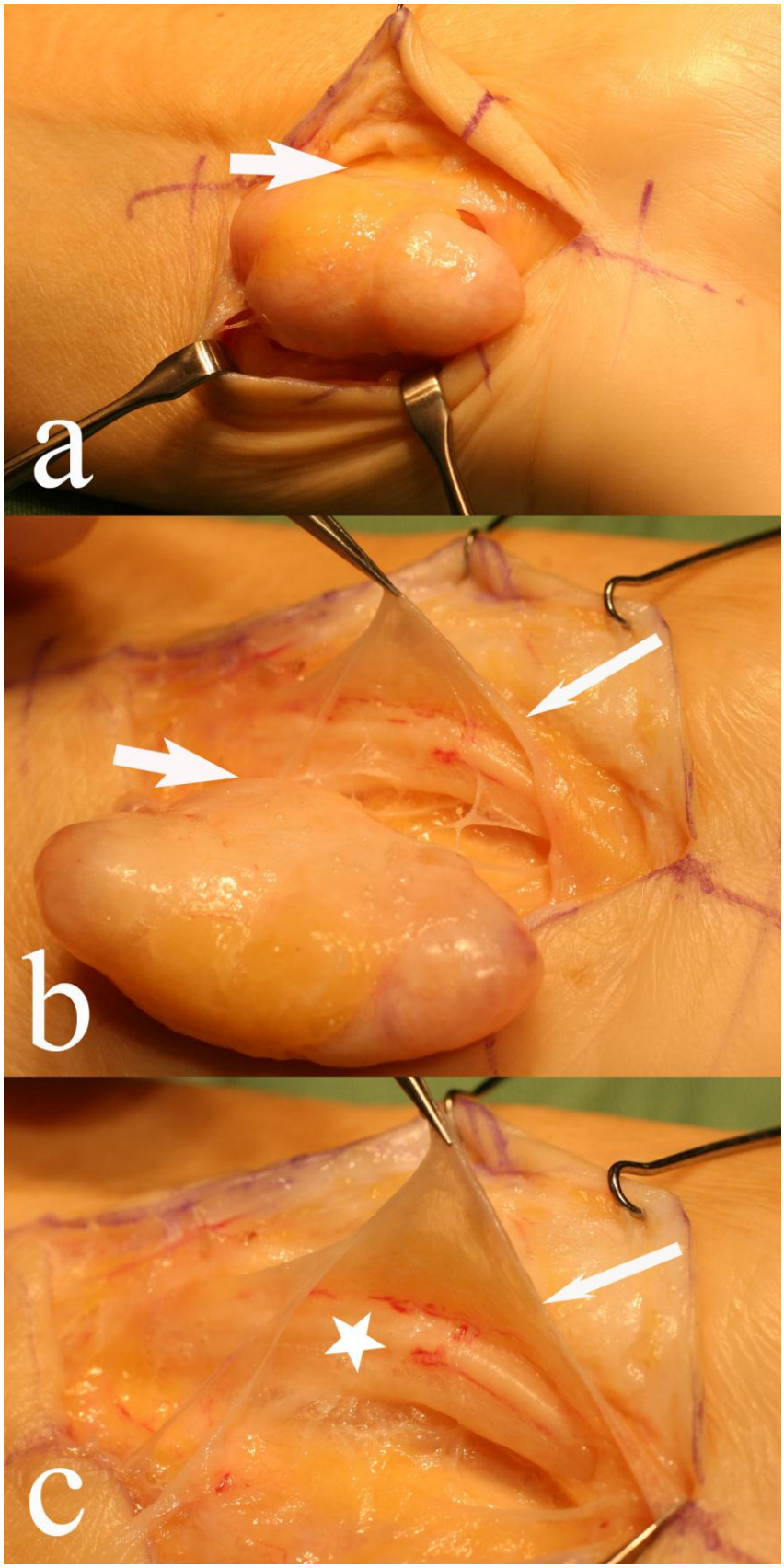

Figure 2