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CASE REPORT

Case Report: Angiomyomatous Hamartoma of the Lymph Node

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Abstract
Angiomyomatous hamartoma (AMH) of the lymph node is a rare, hamartomatous lesion which typically occurs in the inguinal and femoral regions. Although benign, it can clinically and radiologically cause a suspicion of malignancy. Due to its rarity, pathological diagnosis, particularly on small needle biopsies, may be challenging. We present the case of a 67-year-old male with AMH, with a summary of the published cases and discussion of the differential diagnoses.

Keywords Angiomyomatous hamartoma of lymph node · Lymph node hamartoma · Spindle cell lesion · Differential diagnoses

Case Report

A 67-year-old man with no significant past medical history presented to his general practitioner with a lump in the left groin/left upper thigh which had been present for many years. Over recent months, the lump had increased in size and was episodically tender. Clinical examination identified an ill-defined, oval-shaped mass. Due to the recent growth of the lesion, imaging was carried out, and an MRI scan was suggestive of inflammatory changes in a 20 × 9-mm lymph node; however, a primary neoplastic process or a metastasis was in the differential diagnoses. The patient continued to experience discomfort, and three 16G core biopsies were taken from the mass, under ultrasound guidance (Fig. 1).

Microscopic examination of the tissue showed a proliferation of cytologically bland spindle cells with blunt-ended nuclei, which were organised in whirls and interlacing bundles, together with intervening thick-walled medium-size vessels. Some nodular lymphoid infiltrate was included in the material adjacent to the spindle cell proliferation, with retention of the overlying subcapsular lymph node sinus.

On immunostaining, the spindle cell proliferation, including the vessel walls, was strongly positive for smooth muscle actin (SMA) and desmin, with no evidence of expression of S100, CD34, CD99, HMB45, or MyoD1. The lymphoid infiltrate represented a mixture of CD20-positive B-lymphocytes and CD3-positive T cells (Fig. 2).

The morphology and immunophenotype was that of an angiomyomatous hamartoma (AMH) of the lymph node.

The patient was reassured and did not have any additional therapeutic interventions. He remains well and asymptomatic six months after diagnosis.

Discussion

Angiomyomatous hamartoma of lymph node was first described by Chan et al. in 1992 [1]. It is a rare vascular lesion which occurs primarily in the inguinal and femoral lymph nodes and which may be associated with oedema of the ipsilateral limb [2]. It is usually diagnosed following

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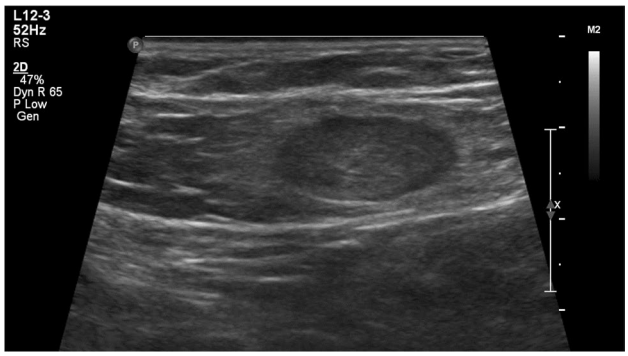

Fig. 1 USS of lymph node, illustrating a dark cortex and bright hilum, which are benign features. The interesting appearances are the slight surface irregularity and a small bright rim immediately surrounding the lymph node, which are suggestive of inflammatory change which could explain the patient's disproportionate pain

lymph node excision. It is more common in males, and the peak age of presentation is the sixth decade [3].

AMH is primarily a localised process which in some patients may present with systemic lymphadenopathy [4]. The aetiology of AMH is not certain, but it is regarded as hamartomatous process, in view of the presence of a mixture of smooth muscle cells, thick-walled blood vessels, and adipose tissue, haphazardly arranged within a fibrous background [3, 5]. Some have suggested that impairment of lymphatic flow may be a factor in its pathogenesis [6]. There are no characteristic molecular features described for the lesion.

Clinical and radiological differential diagnoses of AMH include reactive lymphadenopathy, lymphoma, and metastatic malignancy. A definitive diagnosis can only be made following microscopic evaluation of lesional tissue. Histopathological differential diagnoses include angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), primary nodal leiomyomatosis (LM), lymphangioma, vascular transformation of lymph node sinuses, palisaded myofibroblastoma (PMF), and distinction from normal anatomical structures containing smooth muscle.

In AML, the tumour cells are plumper than in AMH, and will be reactive for HMB45 [7]. Furthermore, the patient may also have renal AML or a history of tuberous sclerosis. LAM can involve lymph nodes in women and can also be seen in association with tuberous sclerosis [3]. LAM has a different morphology and immunohistochemical phenotype to AMH. LAM can have a subcapsular location, extranodal extension, intralymphatic growth, compact nests, branching lymphatic channels, plump cells, and cells with foamy/clear cytoplasm [3]. In LAM, lesional cells stain positively with HMB45 and cathepsin K [3]. In nodal LM, it is the intraabdominal lymph nodes of females that are largely involved, and the lesion is composed of compact bundles of smooth muscle cells with an insignificant vascular component (reviewed in 1). Furthermore, an association with HIV/immunodeficiency has been reported (reviewed in 1). Lymphangiomas are more commonly seen in females, and the cells lining ecstatic spaces will stain positively with D240. Vascular transformation of lymph node sinuses is a reactive process characterised by anastomosing blood vessels in lymph node sinuses, in contrast to AMH which begins in the hilum of the lymph node then extends out towards the cortex. Palisaded myofibroblastoma is one of the most common primary spindle cell tumours of lymph nodes presenting exclusively in inguinal lymph nodes. Morphologically, it

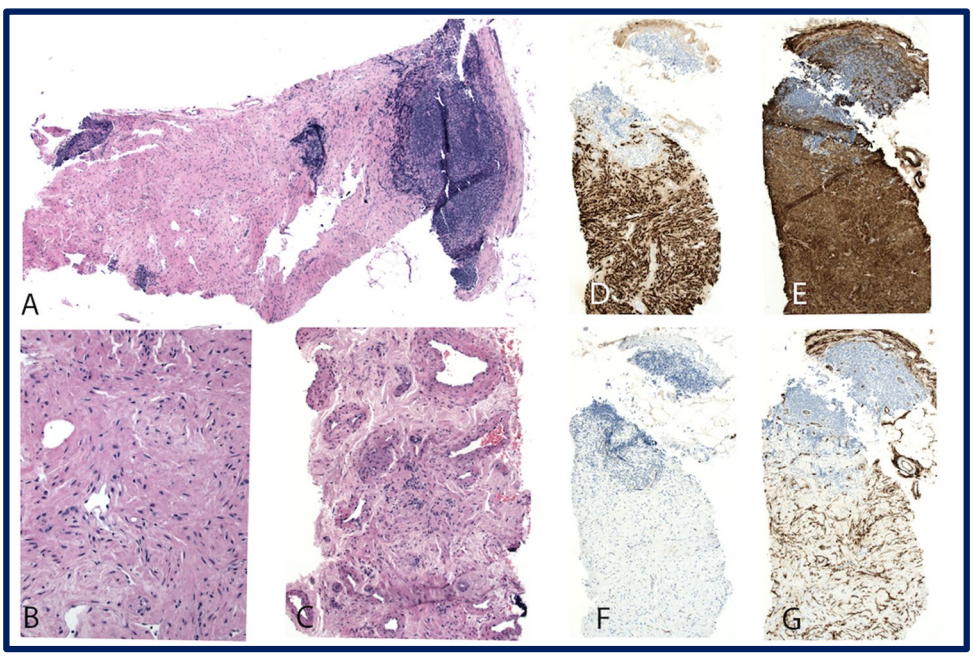

Fig. 2 Histological features and immunohistochemistry. (**A**) Cytologically bland spindle cell proliferation with residual lymph node tissue and lymph node capsule (H&E, ×4); (**B**) The cytologically bland spindle cells form short interlacing cellular bundles with (**C**) area containing thick-walled medium-size vessels (H&E, × 10); There is strong expression of desmin (**D**) and SMA (**E**), with no expression of S100 (**F**) and CD34 (**G**) (immunohistochemistry, × 10)

resembles a Schwannoma or a smooth muscle proliferation. The distinctive features include striking nuclear palisading, frequent perinuclear hyaline globules, and amianthoid fibres, a particular appearance of collagen fibres. Their myofibroblastic, rather than smooth muscle derivation, is characterised by a strong expression of SMA, nuclear positivity for beta-catenin, and no desmin positivity [8].

Smooth muscle in lymph node tissue is present in small quantities in the capsule, trabeculae, and also in the hilum [9, 10]. Smooth muscle and vascular hilar proliferations have been noted in axillary lymph nodes, considered to originate from the native nodal smooth muscle [10].

This is the first case report of AMH being diagnosed on a core biopsy as opposed to lymph node excision. It is important to be aware that lymph node pathology is often diagnosed on a resection specimen rather than on a biopsy, and that interpretation of morphology must take into consideration the amount of lesional tissue being examined. If there is any diagnostic uncertainty, the case should be reviewed by an expert histopathologist. This case highlights the importance of being aware of the entity and its differentials, so that appropriate morphological analysis and immunohistochemistry can be applied to a limited tissue sample.

It is vital that AMH is correctly diagnosed, so that the patient can be reassured and avoid unnecessary diagnostic or therapeutic interventions. The symptomatic lesions are cured by excision, but conservative management may be appropriate, as in our case.

Declarations

Ethics Approval Not applicable

Consent to Participate and Consent for Publication The patient has emailed written consent for publication and has reviewed a copy of the case report

Conflict of Interest The authors declare no competing interests.

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