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Case Report

Angiomyomatous hamartoma: Unusual presentation of a rare entity

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ABSTRACT

Angiomyomatous hamartoma (AMH) is a rare benign vascular growth primarily affecting inguinal and femoral lymph nodes (LNs). Here, we present a unique case of AMH manifesting as a submental neck mass, a location seldom reported in literature. A 20-year-old male presented with a palpable midline neck mass adjacent to the hyoid bone. Ultrasonography suggested a partially cystic lesion, prompting consideration of thyroglossal duct cyst or necrotic lymph node. Fine-needle aspiration (FNA) hinted at a benign cystic lesion, potentially a thyroglossal duct cyst. Surgical excision via the Sistrunk approach revealed no cyst but characteristic features of AMH upon histopathological examination. This case underscores the importance of considering AMH in the differential diagnosis of subcutaneous nodules in unusual locations and highlights the role of surgical excision for both diagnosis and treatment. Our findings expand the understanding of AMH's clinical presentation and emphasize the necessity of a comprehensive differential diagnosis approach for nodal lesions.

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1. Introduction

Angiomyomatous hamartoma (AMH) is a rare benign vascular growth of undetermined aetiology that affects the lymph node (LN). Histologically, it is typified by the substitution of smooth muscle cells, blood vessels, and varying quantities of adipose and fibrous tissue within a collagenous stroma in place of the nodal architecture.[1,2] Since its initial description in 1992, twenty-nine cases have been documented in the literature, with the majority involving the femoral and inguinal LNs. Seldom have descriptions of cervical and submandibular nodal involvement been found.[1,3–7] A congenital condition linked to the persistence of the thyroglossal duct, thyroglossal duct cysts manifest as midline cysts close to the hyoid bone and can have a variety of sonographic appearances.[8,9]

Since sonographic appearances can vary, ultrasonography (US) is a reliable, economical, non-invasive preoperative examination that is typically sufficient to make accurate preoperative diagnoses.[10,11] Despite this, the accuracy of this imaging modality may have limitations. Magnetic resonance imaging and computed tomography serve an additional diagnostic purpose. A definitive diagnosis is obviously possible with surgical removal along with histological evaluation.

Here we are reporting an uncommon instance of AH manifesting as a palpable lump in the anterior midline neck region (submental). The lesion's US preoperative evaluation suggested that it was partially cystic, and because of its unusual placement, a thyroglossal duct cyst, an epidermoid cyst, or a necrotic lymph node were the other possible differentials.

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2. Case Report

A 20-year-old male without other health problems presented with a palpable neck mass of 2 years' duration. Neither dysphonia nor dysphagia affected the patient. The prior medical history was not noteworthy. Upon physical examination, a non-tender, mobile mass in the midline anterior neck, adjacent to the hyoid bone, was noted. There were no swollen lymph nodes found in the side of the neck. The laboratory results fell within the expected range. At USG, a mostly hypoechoic lesion with a well-defined periphery and internal heterogeneous echoes measuring (35 x 20) mm in diameter was found. The thyroid gland displayed no abnormalities. FNA was done and showed presence of cyst macrophages with few squamous cells in a mucoid background suggestive of benign cystic lesion, possibly thyroglossal duct cyst.(Figure 1) The patient underwent surgery to remove the suspected thyroglossal duct cyst. The Sistrunk approach was selected for safety reasons since we were unable to determine intraoperatively whether the lesion was a solid mass or a cystic one, as well as whether it had the potential to be malignant. After a meticulous dissection of the lesion, the hyoid bone's core region was resected and the sample was sent for histopathological examination. Upon gross examination, the removed mass had a maximum diameter of 30 mm (Figure 2). A core fibrous region was seen, but there was no indication of a cyst. The tissue was fixed in 10% buffered formalin, underwent standard processing, and embedded in paraffin. Histopathological examination revealed no epithelial lining and presence of spindle cells in fascicles along with dilated and congested blood vessels (Figure 3). Area of fibrosis, adipose tissue and focal aggregates of lymphocytes was noted. No thyroid tissue was seen. The existence of a cystic lesion was not reported in subsequent sections. The aforementioned results were all in line with the pathological diagnosis of lymph node-derived AH.

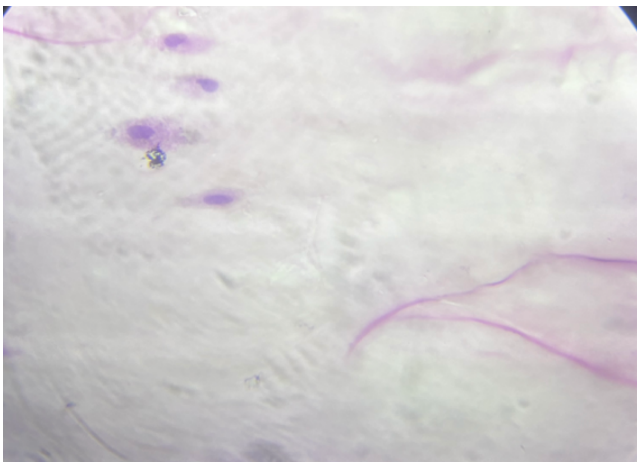

Figure 1: FNAC; presence of cyst macrophages against mucin

Figure 2: Cutsection of the excised tissue sent for histopathology

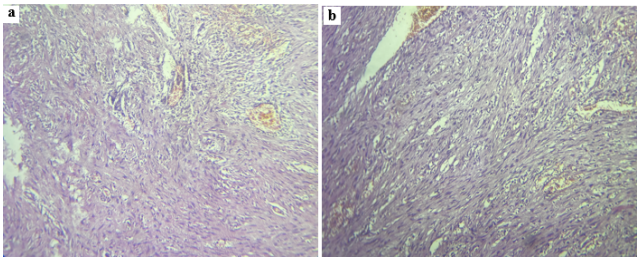

Figure 3: a,b: HPE; Spindle cells with presence of blood vessels

3. Discussion

A rare and benign type of modification to the hilar lymph nodes, AH is characterised by the proliferation of smooth muscle and the replacement of parenchymal tissue with fibrous tissue that contains capillaries. It typically affects the inguinal lymph nodes, which seem to begin in the hilum and extend to the brain. Since Chan et al. originally described this entity in 1992, 29 examples have been documented in the literature.[3,6,7,12–21]

Our case shows unusual involvement of the submental node and validates prior findings that AMH may present outside the inguinal or femoral nodal basins, even though the rarity of AMH may be related to underreporting.

There is still much to learn about the pathophysiology and epidemiology of AMH. The age range at presentation is 8 months to 83 years, with no discernible preference for either sex or age.[1,4] Our case was also presented in a 20-year male, similar to previous studies. There has been considerable debate regarding whether AMH of LN is a unique histologic entity or a variation of previously recognised conditions such lymphangioleiomyomatosis (LAM).[1] It has been hypothesised that the illness, which seems to be idiopathic, results from a reparative response to nodal inflammation.[3]

As our case shows, AMH of LN can also affect the head and neck area. Clinically, it manifests as an indolent subcutaneous nodule or mass within the inguinal or femoral LNs. Common benign disorders like lipoma or cysts are included in the differential diagnosis, along with malignant conditions like lymphoma. Lesions of AMH of LN do not have an overlaying punctum or a history of inflammation and drainage, in contrast to epidermal cysts.[6] While ultrasound is neither sensitive nor specific, it can be useful in distinguishing it from other differentials and in indicating that the lesion is inside an LN.[4] Ultimately, the diagnosis of AMH of LN is made by histological evaluation.

Surgical excision is frequently used to treat and diagnose AMH of LN, and it is frequently successful.[1,5,8] However, a case of AMH that recurred 14 years after the initial surgical excision and showed signs of pain and burning due to compressive neuralgiademonstrated an exception to this usual treatment. Instead of undergoing another excision, the lump was treated with intralesional steroid injections, and it stopped growing after that.[5] On the other hand, our case on follow-up did not show any evidence of recurrence.

To the best of our knowledge, this is the first instance of AMH of LN in the submentalregion. Our results also support earlier reports showing that LNs outside of the inguinal and femoral areas can be affected by AMH of LN. Thegoal of the index caseis to raise awareness of this vascular growth as a differential diagnosis, emphasise its benign clinical course, and demonstrate how surgery can be used to cure it. The need of keeping a high index of suspicion and considering a wide differential diagnosis while assessing nondescript subcutaneous nodules or masses close to nodal basins is highlighted by our case.

4. Source of Funding

None.

5. Conflict of Interest

None.

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