Primary Nodal Hemangioma

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Benign vascular tumors arising primarily in lymph nodes are rare. The importance of identifying these entities is to avoid misdiagnosing them as malignant vascular tumors, which occur more often in lymph nodes. Hemangioma is a benign vascular tumor, but its occurrence in lymph nodes is extremely rare. Hemangiomas can occur at any age, mostly in females. It is usually asymptomatic, affects only one node, and does not recur. Four histologic types of hemangioma have been identified: capillary/cavernous, lobular capillary, cellular, and epithelioid. This review highlights the key features of previously reported cases and discusses the differential diagnosis.


Benign vascular tumors arising primarily in the lymph nodes are rare.2 Apart from a study of 39 cases by Chan et al,3 there have been only occasional case reports in the literature. Although benign nodal vascular proliferations are uncommon, identifying these entities can help to avoid misdiagnosing them as malignant vascular tumors, which occur more often within lymph nodes.

Hemangioma is 1 of the 4 types of benign nodal vascular tumors classified by Chan and colleagues.3 Although hemangioma is common in skin, mucosa, and soft tissue, its occurrence in lymph nodes is extremely rare.2 To date, 18 cases have been reported in the English language medical literature, to our knowledge. This review highlights the key features of previously reported cases and discusses the differential diagnosis.

CLINICAL FEATURES

The age reported in the literature for presentation of nodal hemangiomas varies, ranging from 4.5 to 75 years. There is a female predominance, and usually only a single node tends to be involved. Hemangiomas occur in both peripheral and more centrally located lymph nodes, such as supraclavicular, submental, cervical, axillary, common iliac, pelvic, inguinal, and oral soft tissue lymph nodes.2–9 Some nodal hemangiomas are diagnosed incidentally when lymph nodes are surgically removed in a radical mastectomy for breast cancer or radical hysterectomy for endometrial adenocarcinoma, without any antecedent radiotherapy.2,8 Most patients are asymptomatic. However, when superficial and peripheral nodes are affected, a mass is often the presenting feature.2–9 Some nodal hemangiomas have been associated with other vascular lesions, such as cavernous hemangioma of the liver, intestinal angiodysplasia, oral hemangiopericytoma, and vascular esophageal polyp.5,9,10

GROSS AND MICROSCOPIC EXAMINATION

Grossly, the size of the involved lymph nodes ranges from 2 to 35 mm.2–9 Microscopically, 4 histologic types have been identified: capillary/cavernous, lobular capillary, cellular, and epithelioid.2–9,11 Capillary/cavernous hemangioma is more often centered on the lymph node hilum or medulla with well-preserved nodal parenchyma,11 and is either a well-defined or poorly defined mass of closely packed capillaries or cavernous vessels lined by flat endothelial cells, and which can be empty or filled with blood.2,9 The lobular capillary type can almost replace the entire nodal parenchyma and has an appearance similar to a pyogenic granuloma.2 The cellular type is composed of closely packed, nearly solid to rarely canalized, vascular channels that can be outlined by periodic acid–Schiff and reticulin stains.3 The epithelioid type is characterized by plump endothelial cells.1,3 In all types, no cytologic atypia, necrosis, mitoses, or extravasated erythrocytes are present.2–4 The endothelial cells in hemangioma show immunopositivity for smooth muscle actin, CD31, CD34, and factor VIII–related antigen.1,11 Much of the nodal parenchyma is effaced by a vascular proliferative lesion with remnants of the residual cortex containing reactive follicles present (Figure, A). The vascular lesion extends from the subcapsular sinuses through to the medulla of the lymph node. The lesion is composed of vessels of varying caliber; however, there is a predominance of small capillary-sized vessels with an occasional edematous myxoid stroma (Figure, B). Under low magnification, the lesion has a lobular appearance in areas. In other areas, the vessels have a more anastomosing, slitlike appearance. The individual vessels are composed of endothelial cells with a characteristic hobnail appearance, and, in some areas, the lining is thrown into small, papillary tufts (Figure, C). Solid sheets of tumor cells are not usually seen. Interspersed among these small-caliber vessels are occasional larger vessels, which are presumably feeder vessels (Figure, D). Mild nuclear hyperchromasia and pleomorphism of the endothelial cells may be encountered occasionally; however, mitoses, hemorrhage, and necrosis are not evident.

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DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes other benign vascular lesions, such as angiomymomatous hamartoma, which is more common in males and has been reported only in inguinal lymph nodes. It starts in the hilum of the node and the parenchyma is replaced by blood vessels, fat, smooth muscle, and fibrous tissue without cellular fascicle formation.\(^1,2,12\) Lymphangioma of the lymph nodes is always associated with involvement of other organs, and the immunohistochemical marker D2-40 will highlight the lymphatic vessels.\(^2,13\) Epithelioid hemangioendothelioma occurs more commonly in soft tissue and bones than in lymph nodes. It consists of sheets and cords of plump spindle cells with nuclear pleomorphism and abundant eosinophilic cytoplasm with vacuoles occasionally containing erythrocytes, set in a myxoid stroma.\(^2,14\) Polymorphous hemangioendothelioma is a rare vascular tumor of borderline malignant potential that most frequently involves peripheral nodes but occasionally affects internal or central nodes, soft tissues, and internal organs such as the liver. Adjacent nodes can be involved in visceral-based cases. This tumor is known to recur and metastasize but has not been described either in children or in women. Histologically, it is characterized by solid areas of polygonal cells with oval nuclei and angiomatous areas, with up to 14 mitoses per 10 high-power fields. The lesional cells show positivity for endothelial markers, such as CD31, factor VIII, and CD34, and they do not show reactivity for epithelial markers.\(^2,15,16\) Vascular transformation of lymph node sinuses is characterized by conversion of subcapsular, intermediate, and medullary sinuses into capillary-like channels associated with extravasated erythrocytes, vascular obstruction, and often, fibrosis.\(^17\) Rarely, if ever, is the entire lymph node architecture effaced and replaced by the vascular proliferation.

Other vascular tumors, such as Kaposi sarcoma, should be excluded. Kaposi sarcoma shows a proliferation of slightly atypical spindle cells that form slitlike vascular spaces, sometimes showing prominent mitoses, extravasated erythrocytes, hemosiderin, and closely associated

An example of a hemangioma from an enlarged inguinal node of an asymptomatic, 23-year-old woman. A. Much of the nodal parenchyma is effaced by a vascular proliferative lesion with remnants of the residual cortex containing reactive follicles. B. Predominance of small, capillary-sized vessels with an edematous myxoid stroma. C. The lining endothelial cells with a characteristic hobnail appearance, and, in some areas, the lining is thrown into small, papillary tufts. D. Occasional larger vessels interspersed among small-caliber vessels (hematoxylin-eosin, original magnifications ×40 [A and B], ×200 [C], and ×100 [D]).
plasma cells. The clinical setting for primary nodal Kaposi sarcoma is also quite characteristic because it is predominantly found in children.\(^1\)

Composite hemangioendothelioma is a rare, low-grade, malignant vascular tumor showing varying combinations of benign, low-grade, malignant, and malignant vascular components with significant potential for local recurrence but little, if any, potential for distant metastasis. It usually presents in the nodes as a metastasis from a soft tissue primary.\(^1\)

Angiosarcoma is usually metastatic rather than primary. It has vascular spaces or channels that are usually obvious and lined by tumor cells showing cytologic atypia.\(^1,11\) Dabska tumor is a rare, low-grade angiosarcoma with metastatic potential, which often affects the skin of children. It may spread to regional lymph nodes and produce disseminated lung metastases. Histologically, it is characterized by anastomosing vascular spaces with intravascular papillary outpouchings projecting, sometimes in a glomerulus-like pattern, into a lumen lined by atypical columnar endothelial cells.\(^19,20\)

**TREATMENT AND PROGNOSIS**

Surgical excision is curative in primary nodal hemangioma. Although follow-up has not been reported in all cases, in those with follow-up, no recurrences have been documented for nodal hemangiomas.

**CONCLUSIONS**

Primary nodal hemangioma is an extremely rare, benign vascular tumor that is important to identify to avoid misdiagnosing it as a malignant vascular tumor, an entity that occurs more often within lymph nodes.

**References**


