CASE STUDY

An anastomosing haemangioma within a lymph node: a rare vascular tumour in a novel location

Yee Sing Lin, Andrew Parasyn and Trent Davidson

ABSTRACT

Anastomosing haemangioma is a rare, benign vascular tumour with a predilection for genitourinary and paravertebral sites, but which has been described in a number of other locations. We report a case of an anastomosing haemangioma in a lymph node presenting as a painful right axillary mass. Microscopic examination revealed a characteristic non-lobular proliferation of anastomosing, thin-walled capillaries, with focal endothelial hob-nailing, and the absence of significant cytologic atypia or mitotic activity.

Keywords: anastomosing haemangioma, lymph node.

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INTRODUCTION

Anastomosing haemangioma is a rare, benign vascular tumour first described in the genitourinary tract (1). Histologically, the lesion is characterised by anastomosing sinusoidal capillary sized vessels with occasional hobnailed endothelial cells, fibrin thrombi, and absence of mitotic activity and cytologic atypia (1) and can be easily misinterpreted as a well differentiated angiosarcoma. This lesion has since been noted in the genitourinary tract (1), para-vertebral soft tissues (2), gastrointestinal tract (3), skin (4), gynaecologic tract (5), upper respiratory airway (6,7), and a variety of other viscera (8-10).

We report a case of an anastomosing haemangioma identified within a lymph node and presenting as a painful right axillary mass.

CASE

A 48-year-old female presented with a tender mass in the right axilla. The initial clinical diagnosis was of a neuroma. MRI revealed a round enhancing lesion with a possible hilum. After resection, a 10mm lymph node was received with multiple attached fragments of fat. Microscopically there was a 7mm benign vascular proliferation expanding and occupying the vascular sinusoid of a compressed lymph node. The lesion was largely circumscribed; however, did not have a lobular architecture, and consisted of anastomosing, variably dilated, thin walled vessels lined by endothelial cells with focally hobnailed nuclei.

Centrally, there were thick walled arteriolar type vessels. There was no endothelial atypia, multilayering, or mitotic activity. The stroma between the vascular lumina contained plump spindled cells and appeared hyalinised and a 3.3mm central portion of the lesion showing marked oedema of the stroma, possibly representing secondary trauma related change. Occasional mast cells were present within the tumour. CD31 and CD34 highlighted the vascular endothelial cells while the intervening stromal cells were negative. D2-40 did not stain the tumour cells, however, highlighted subcapsular lymphatic sinuses. SMA was positive in surrounding pericytic stromal cells.

The associated lymphoid tissue appeared attenuated and reactive. No abnormal lymphoid population was identified on flow cytometry. The morphologic appearances of the lesion were felt to represent an anastomosing haemangioma arising within a lymph node. Three additional reactive appearing lymph nodes were also seen in the submitted fat. These were uninvolved by the anastomosing haemangioma.

DISCUSSION

Lymph nodes are a distinctly rare site for vascular tumours. The morphologic spectrum of such tumours includes haemangiomas, angiomyomatous hamartomas, epithelioid vascular tumours (such as epithelioid haemangiomas, epithelioid haemangioendotheliomas, and epithelioid angiosarcomas), polymorphous haemangioendothelioma (a rare, vascular tumour composed of variable solid, primitive, and angiomatous components with borderline malignant potential), lymphatic malformations, and Kaposi's sarcoma (11). Intranodal papillary intravascular angioendotheliomas, also known as Dabska tumours, have also been described (12).

Anastomosing haemangiomas are rare and were first described in the genitourinary tract (1). They are benign and have been noted in the kidney, testis (1), mediastinum, uterus, para-aortic and para-vertebral soft tissues (2), adrenal (10, liver, small bowel, colon (3), skin (4), breast (9), ovary (5), larynx (6), nasal cavity (7), and atrium (8). They can occasionally be multifocal.

The histologic features of this tumour characteristically include a non-lobular proliferation of tightly packed and anastomosing capillary-like vessels. In addition, there may be fibrin thrombi, hobnailed endothelial cells, intracytoplasmic eosinophilic globules, and adjacent extramedullary haematopoiesis. Given the non-lobular growth and occasional prominent endothelial cells, the main diagnostic differential is of a well differentiated angiosarcoma. Notably, however, there is no multilayering of the endothelial lining, high grade cytologic atypia, or mitotic activity.

In the present case we did not find eosinophilic globules within the tumour. We also noted occasional mast cells throughout the lesion, which have not been reported in anastomosing haemangiomas in other locations. Consistent with cases reported in other locations (1,3), the tumour stained positive for CD31 and CD34. Negative D2-40 staining excluded a tumour of lymphatic lineage. Fresh tissue was split for routine flow cytometry as the tumour was an unexpected pathologic finding; no abnormal lymphoid population was identified.

Anastomosing haemangiomas have been found to have recurrent mutations in GNAQ and its homologues GNA11 and GNA14 (13), suggesting it is a neoplastic, rather than a reactive process. Activating mutations in GNAQ have also been identified in other benign vascular tumours, particularly congenital vascular lesions associated with Sturge-Weber syndrome (13). However, they have not been reported in angiosarcoma suggesting that the two are not genetically related.

In summary, this is a rare case of anastomosing haemangioma reported in a lymph node, further adding to the documented anatomical extent of this tumour. Awareness of this lesion is important to avoid misinterpretation as a malignancy.

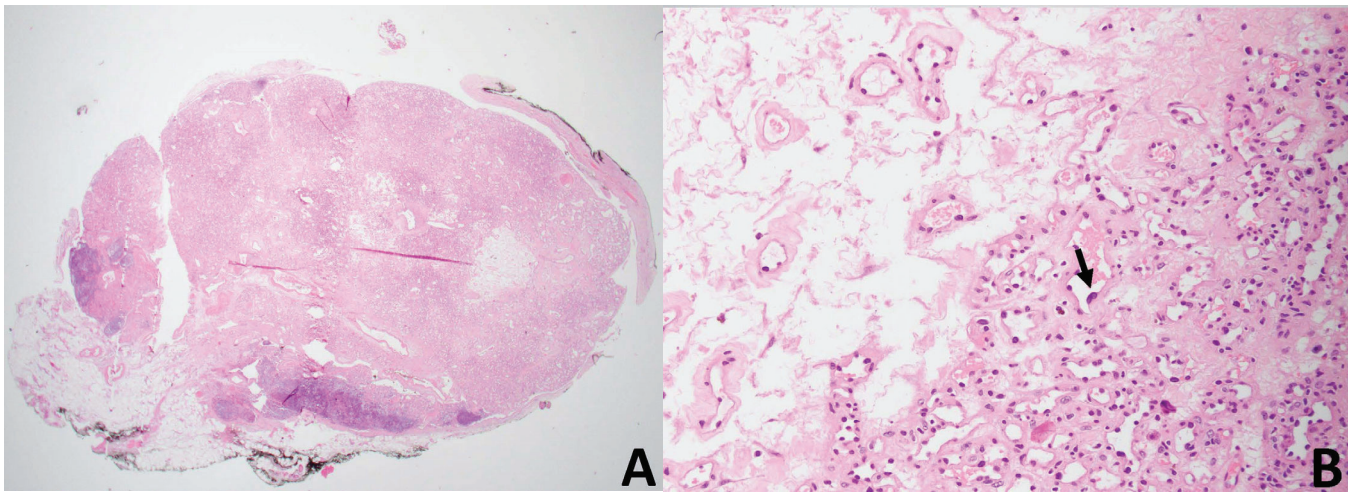

Figure 1. (A) Scanning magnification view of lymph node reveals replacement by a benign vascular lesion. Residual lymphoid tissue is compressed at the periphery. Magnification: 12.5x. **(B)** The central portion of the lesion has a significantly oedematous stroma with hyalinised vessels. Note the hobnailing of the endothelial cells (black arrow). Magnification: 200x.

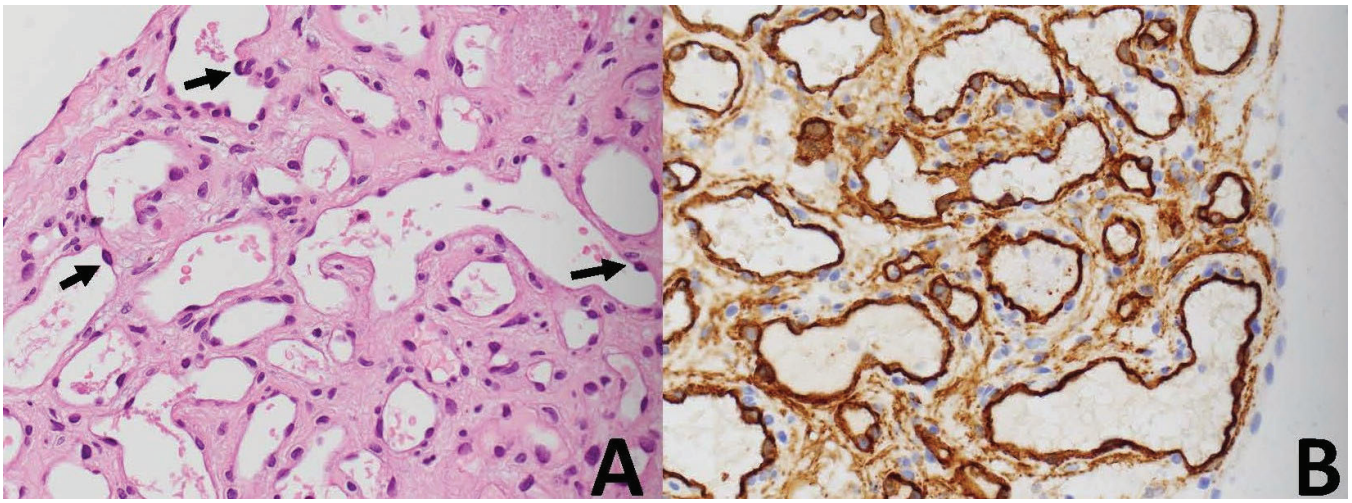

Figure 2. (A) The lesion is composed of anastomosing capillary like vessels without cytologic atypia or multilayering. Note the hobnailing of the endothelial cells (black arrow). Magnification: 400x. **(B)** CD34 stains the endothelial cells and highlights the vessels. Magnification: 400x.

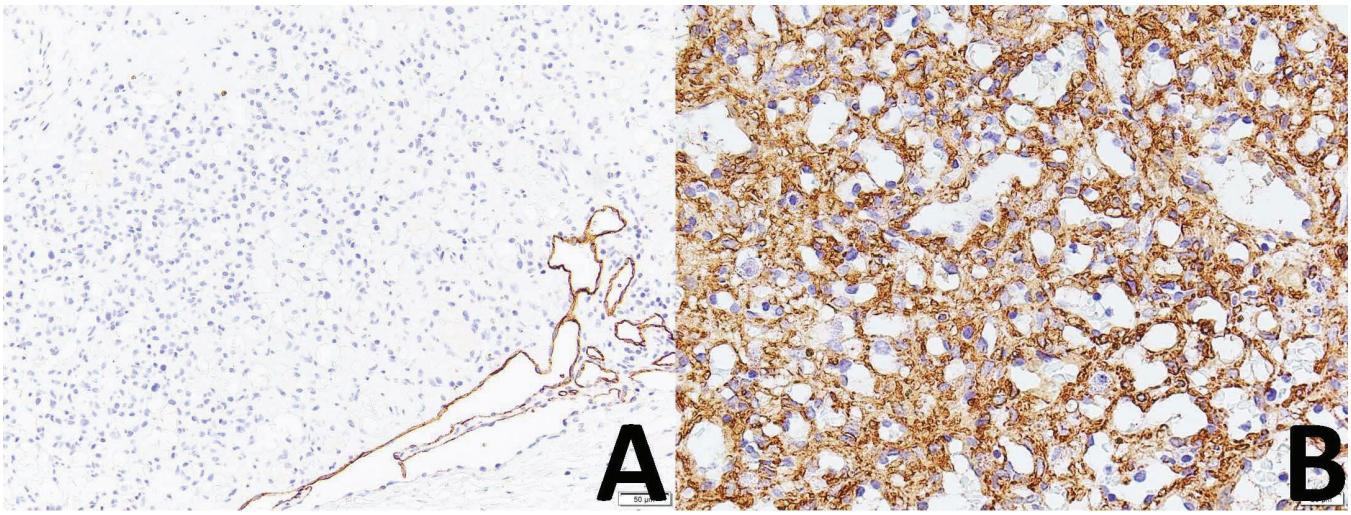

Figure 3. (A) D2-40 staining is negative in the tumour cells. The lymphatic sinuses are highlighted in the subcapsular region. Magnification: 200x. **(B)** SMA highlights the surrounding pericytic layer. Magnification: 400x.

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