Cervical angiofollicular lymph node hyperplasia (lymphoid hamartoma)

By B. D. Pujari and S. G. Deodhare (Miraj, Maharashtra, India)

Introduction

In 1956 Castleman *et al.* reported a case of a large lymphoid mass in the mediastinum and called it 'benign lymph node hyperplasia' resembling thymoma. Since then about 150 cases have been reported and of these 84 have occurred in the thorax, 16 in the abdomen, 24 in the neck and axilla, and the remainder in diverse other sites (Harrison, 1975). This peculiar lymphoid mass is known by various synonyms, including lymphoid hamartoma, angiofollicular lymphoid hamartoma, angiofollicular lymph node hyperplasia, giant hemolymph node, benign giant lymph node and follicular lymphoreticuloma. We have encountered a case of the hyaline vascular type of angiofollicular lymph node hyperplasia in the neck. At present the major importance of this small class of tumour-like lesions is that they may be mistaken clinically for lymphomas. They have a characteristic histological appearance. They are benign lesions and can be easily treated. The distinctive histological features and the rarity of the lesion have prompted us to report this case.

Report of a case

A male aged 25 years complained of a very slowly growing mass in the neck since childhood. During the last three years or so he had noticed another small swelling about one and half inches below the upper one (Fig. 1). Both were symptomless. The patient had been treated for otitis media on the right side in childhood.

Examination revealed a normally built and developed patient. There was a 7×5 cm swelling in the region of the carotid triangle, on left side of the neck. It was smooth, well defined, and non-pulsatile. The skin over the swelling was normal. There was no movement with deglutition or with protrusion of the tongue. It was non-tender, partly deep to the anterior part of the sternomastoid muscle, and adherent to the deeper structures.

The second swelling was at a lower level in the posterior triangle of the neck, just behind the posterior border of the sternomastoid muscle. It was 1×1 cm. in size, well defined, smooth, non-tender, and free from deeper structures. The routine laboratory investigations and skiagram of the chest were normal. As there was no detectable pathology elsewhere, an excisional biopsy of both the swellings was done under general anaesthesia. The bigger swelling was extremely vascular.

Pathological features

The nodule in the region of the carotid triangle was encapsulated, ovoid, firm, pale brown mass measuring $6 \times 5 \times 2 \cdot 5$ cm and weighing 44 g. The cut surface

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FIG. 1.
Clinical photograph showing two swellings in neck.

showed a solid, well-vascularized greyish-white appearance with areas of haemorrhage at the periphery (Fig. 2). The second small nodule from the posterior triangle was well defined, smooth and measured 1 × 1 cm. It had a solid grey cut surface.

Microscopically the large node showed a fibrous capsule. There was no distinction into cortex and medulla and there was an absence of lymph sinuses both in the subcapsular zone and in the centre of the node. The subcapsular area showed many dilated blood vessels. The main feature was an aggregation of lymphoid follicles with centrally-placed thick-walled arterioles showing endothelial cell proliferation (Fig. 3). The lymphoid follicles were composed of packed lymphocytes arranged in concentric rows like strings of beads and showing no germinal centres. The lymphocytes were mature and of uniform morphology. The endothelial cells of the central arteries had a concentric arrangement. Some of the arterioles were hyalinized with obliterated lumina (Fig. 4). The interfollicular tissue showed an increased number of similar arterioles and also contained sheets of lymphocytes, variable fibrous tissue but no plasma cells.

The silver impregnation method for reticulin revealed that the follicles had very scanty fibres amongst which the thick-walled arterioles were prominent. In between the follicles there was a regular meshwork of reticulin with numerous thick-walled arterioles and no visible sinuses. Most of the reticulin fibres along the circumference of the node were not compressed (Fig. 5).

The second small node showed similar but early histological changes. Few of the lymphoid follicles showed germinal centres. In some of the follicles a definite angiofollicular pattern was lacking. In some places irregular sinuses were noted.

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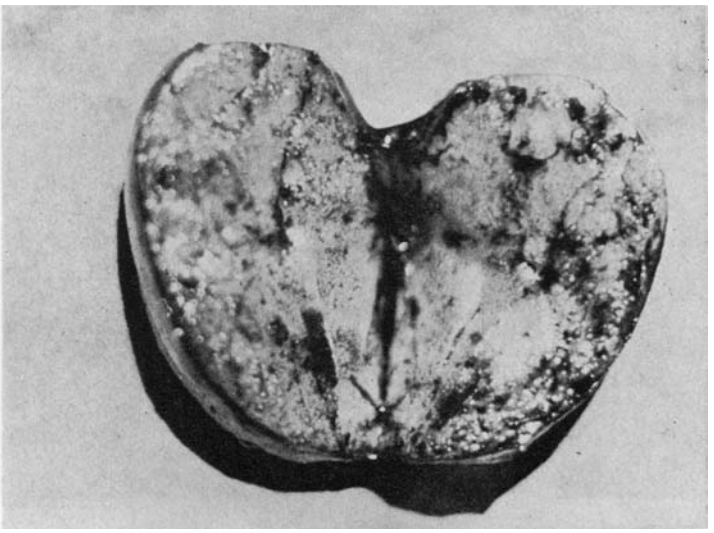


FIG. 2.
Cut of upper lesion showing solid grey appearance with vascularized and haemorrhagic areas.

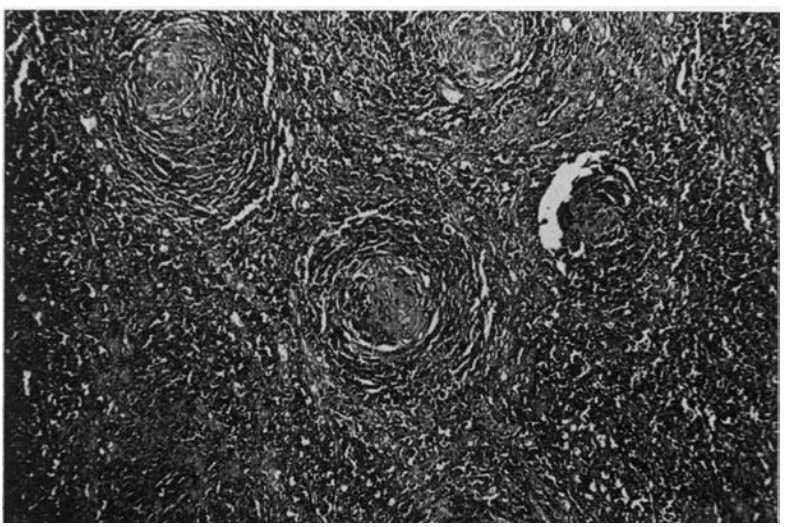


FIG. 3.
The lesion showing lymphoid follicles with vascular centres. (H & E ×240.)

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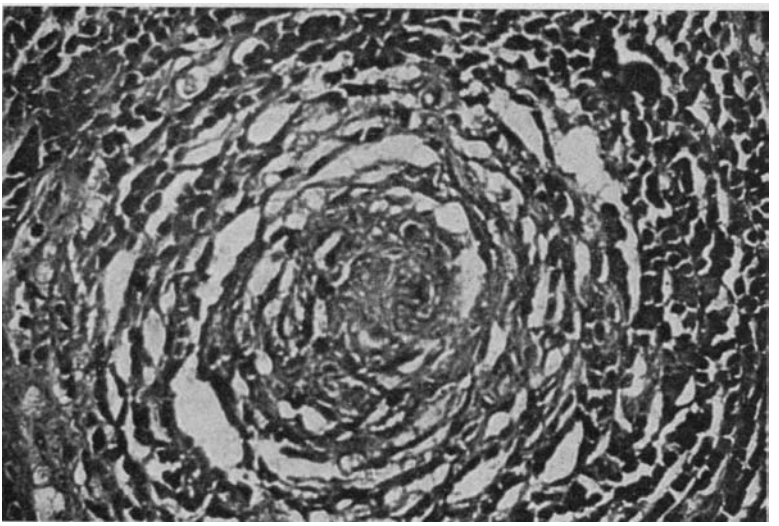


FIG. 4.

A follicle with centrally-placed thick-walled arteriole which shows endothelial cell proliferation and obliterated lumen. The lymphocytes are arranged in concentric rows. (H & E ×960.)

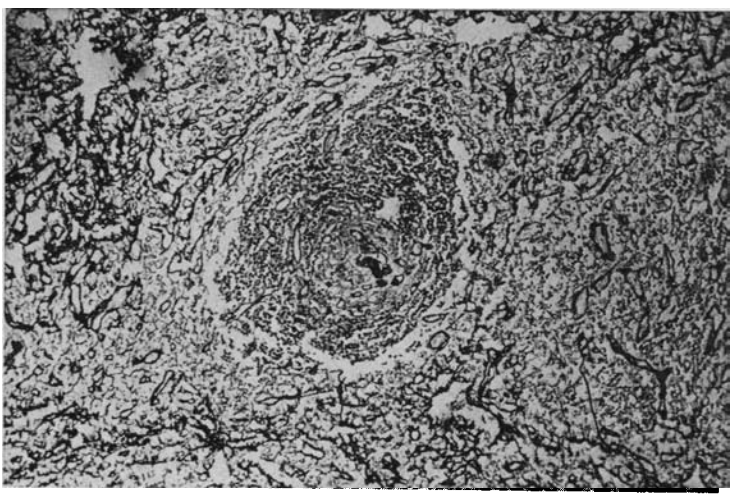


FIG. 5.

A small blood vessel in the follicle is distinctive. The reticulin fibres are scanty in the follicle. Perifollicular tissue shows meshwork of reticulin with numerous vascular channels. (Reticulin stain ×240.)

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Discussion

Angiomatous lymph node hyperplasia occur most commonly in the thorax and less commonly in the neck. They have been reported rarely in pelvis, vulva, mouth, axilla, mesentery, retroperitoneum, maxillary antrum, retro-orbital region and soft tissues of the extremities (Tung and McCormack, 1967).

They are divided into two histological types, the hyaline vascular and the plasma cell type (Keller *et al.*, 1972). The hyaline vascular type is five times more common and is usually without general symptoms. It might be discovered accidentally or because of local pressure symptoms. The plasma cell type is often associated with such abnormalities as fever, growth retardation, splenomegaly, anaemia, increased erythrocytic sedimentation rate, leucocytosis, thrombocytosis and altered serum proteins. These abnormalities disappear or become less severe within 1 or 2 months after excision of the lesion (Humphreys *et al.*, 1975). Recently Humphreys *et al.*, (1975) have reported a case of plasma cell-type mesenteric angiofollicular lymph node hyperplasia with nephrotic syndrome in an 18-year-old female. Removal of the lesion resulted in remission of the nephrotic syndrome. It is postulated that the lymphoid mass produced a toxic substance which affected the metabolic properties of the glomerular basement membrane.

The plasma cell type has all the histological features of the hyaline vascular type as described in the present case with an additional feature of heavy plasma cell infiltration in the intervening lymphoid follicles. At places some clusters of eosinophils may be seen. Also true lymphoid follicles with germinal centres are present in appreciable numbers.

Some lesions share features of both types and are transitional (Hartsock, 1968). All these types grow very slowly (Stanford, *et al.*, 1966).

The hyaline vascular type is often very vascular and bleeds readily during surgery. It seems to be related to the vascular proliferation seen microscopically (Castleman *et al.*, 1956; Cheen, 1957; Krasznai and Juhasz, 1969; Mackay, 1969; Tung and McCormack, 1967). The term angiofollicular is coined because of the characteristic vascularity of the lesion.

Lattes and Pachter (1962) consider the following five criteria essential for histological diagnosis of the lesion:

1 A nodular of follicular architecture.

2 The presence of epitheloid (endothelial) cells in the follicular centres arranged to mimic Hassall's corpuscles.

3 Absence of the subcapsular and medullary sinuses found in normal lymph nodes.

4 A distinctive vascular pattern with prominent arterioles in the centre of lymphoid follicles and prominent vascularity of the inter follicular tissue.

5 A population of entirely normal lymphocytes.

The well-vascularized lymphoid follicles have been confused with the Hassall's corpuscles of a thymoma and with the red pulp of an ectopic spleen. Sometimes these lesions can be mistaken for follicular hyperplasia of a lymph node or for follicular lymphoma. However, the absence or extreme paucity of germinal centres excludes follicular hyperplasia. The recognition of thick-walled

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arterioles and the absence of compression of reticulin fibres along the circumference of the follicles should exclude a follicular lymphoma. Similarly a rheumatoid lymph node might be mistaken for the plasma cell type. Sometimes the histological manifestations are similar, but the clinical features are quite different.

There is a lot of controversy about the nature of these lesions. The cause of lymphoid proliferation itself is not known. However, chronic infection and viruses have been suggested as possible causes (Harrison, 1975; Keller *et al.*, 1972). But so far no virus has been isolated. Probably the plasma cell type represents an early process, and the hyaline vascular type is its late stage (Flendring, 1972). This can explain the transitional lesions but these findings cannot be correlated with the age of the patient and the duration of the mass. They could be due to a different type of immunological response by the host. Similarly they could be due to two different but closely related antigens (Keller *et al.*, 1972).

Some authors suggest that it may be a hamartoma of lymphoid tissue (Abell, 1957; Lattes and Pachter, 1962; Tung, and MacCormack, 1967). Willis (1962) points out that they have some of the major characteristics of hamartomas. The extranodal formation of these lesions is suggestive of hamartoma (Climie *et al.*, 1964; Keller *et al.*, 1972). They occurred in regions in which lymph nodes are not situated. Keller *et al.* (1972) and Hartsock (1968) explain their occurrence due to the potential of benign lymph nodule formation in extranodal sites as a result of chronic inflammation. The absence of lymph sinuses and the irregular arrangement of the lymphoid follicles is suggestive of hamartoma (Lattes and Pachter, 1962). But in some reports residual lymph node structure is observed (Timme, 1965). In the present case the second node showed a partly normal lymph node structure and was partly replaced by lymphoid hyperplasia suggesting that the disease is acquired, and is a benign slowly growing lesion. The larger node in the present case showed no normal lymph node structure and appears to represent the late phase while the smaller node seems to be an early lesion.

Complete surgical excision is the treatment of choice and is curative. If total excision is not possible, partial excision does not appear to alter the biological behaviour of these lesions. Radiotherapy and chemotherapy have no value in their treatment.

Summary

A case of the hyaline vascular type of angiofollicular lymph node hyperplasia (lymphoid hamartoma) in the neck of a 25-year-old Hindu male is reported. A brief review of the literature is presented and relevant clinical, pathological and therapeutic aspects of this lesion are discussed.

REFERENCES

Abell, M. R. (1957) *Archives of Pathology*, **64**, 548.

Castleman, B., Iverson, L., and Menedex, V. (1956) *Cancer*, **9**, 822.

Cheen, H. (1957) *Journal of Mount Sinai Hospital*, **24**, 750.

Climie, A. R. W., Waggpmer, L. G. and Krabbenhoft, K. L. (1964) *Laryngoscope*, **74**, 1831.

https://doi.org/10.1017/S0022215100084024 Published online by Cambridge University Press

FLENDRING, J. A. (1972) Cited by Keller *et al.*
HARRISON, C. V. (1975) *Recent advances in pathology*. Churchill Livingstone, Edinburgh, No. 9, pp.87, 92.
HARTSOCK, R. J. (1968) *Cancer*, **21,** 632.
HUMPHREYS, S. R., HOLLEY, K. E., SMITH, L. H. and MILLARTH, D. C. (1973) *Mayo Clinic Proceedings*, **50,** 371.
KELLER, A. R., HOCHHOLZER, L. and CASTLEMAN, B. (1972) *Cancer*, **29,** 670.
KRASZNAI, G. and JUHASZ, I. (1969) *Journal of Pathology*, **97,** 148.
LATTES, R., and PACHTER, M. R. (1962) *Cancer*, **15,** 197.
MACKAY, J. S. (1969) *Canadian Medical Association Journal*, **100,** 434.
STANFORD, W., GIVLER, R. and LAWRENCE, M. S. (1966) *Journal of Throacic and Cardiovascular Surgery*, **52,** 303.
TIMME, A. H. (1965) *South African Medical Journal*, **39,** 356.
TUNG, K. S. K. and MCCORMACK, L. J. (1967) *Cancer*, **20,** 525.
WILLIS, R. A. (1962) *Borderland of embryology and pathology*, Butterworths, London, 2nd ed. p. 351.

Dr. B. D. Pujari, MS, FICS,
Shree Hospital, MIRAJ–416 410,
Maharashtra.

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