

ISSN 1312-1723

Case Study

BACILLARY ANGIOMATOSIS

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ABSTRACT

PURPOSE: Bacillary angiomatosis presents as solitary or multiple skin and/or visceral vascular proliferation that is caused by *Rochalimaea henselae*. It is manifested in patients with compromised immunity, more in HIV - positive cases. Both are with lymph node localization of the disease in HIV positive and in myeolo-dysplastic patients.

METHODS: Biopsy findings on routine haematoxylin and eosin slides along with Warthin-Starry stain and ultrastructure for identification of the aetiological agent are described.

RESULTS: Differential diagnosis to exclude malignant vascular neoplasia or similar looking benign process is discussed.

CONCLUSIONS: To the best of our knowledge these are the first two cases described in Bulgarian medical literature.

Key words: Bacillary angiomatosis (BA), immune deficiency, vascular proliferations, vascular neoplasia

INTRODUCTION

Primary vasoproliferative lesions in lymph nodes are infrequent Most common finding is sinusoidal transformation because of impaired lymph and venous drainage. Quite infrequent is detection of intranodal vascular tumour formation — haemangiomas, haemangioendotheliomas (1), or Karposi sarcoma in HIV positive patients (2, 3).

Bacillary angiomatosis (BA) is comparatively newly described vascular proliferation (4). It can be detected in HIVpositive patients as well as seronegative patients with compromised immunity (3). To the best of our knowledge these are the first two cases described in Bulgarian medical literature.

CASE REPORTS

• Case 1: a-36-year-old drug addicted male, HIV- positive; complaining from malaise and fever for 3 months; inspection of left inguinal region shows hernia and enlarged lymph node measuring 1,5 cm, soliddense and resilient by palpation (Service de Maladies Infectieuses Hopital Rotschild, Paris, France) [poster presentation at National Conference in Pathology, Bulgaria, 1997].

 Case 2: a-61-old HIV-negative male, malaise from 4 months, loss of appetite, rhinitis, dry coughs, diffusely enlarged cervical, axillary and inguinal lymph nodes – painless and tender-hearted (Haematology Department – Medical University of Plovdiv).

MATERIALS AND METHODS

Specimens from two lymph nodes of each case (inguinal from Case 1 and right axillary from Case 2), fixed in 10 % buffered formalin, processed by routine methodology are cut at 4 μ m, stained with haematoxylin and eosin and Warthin-Starry. For electron microscopy materials were fixed in 4 % glutaraldehyde, osmicated; embedded in durcopan; ultra thin sections were stained with uranyl acetate-lead citrate and investigated under transmission electron microscope (Philips CM 12 /STEM).

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RESULTS

Histological findings in both lymph nodes are identical: multiple hyperaemic capillaries with large polygonal epitheloid cells, protruding into their lumens. Both cells and nuclei display mild polymorphism (*Figure 1*). Nuclear chromatin is finely granular, some of the nuclei are vesicular, nucleolar vacuoles are identified, mitosis are absent. Interstitial BELOVEJDOV V., et al.

tissue displays sparse lymphocytes and plasma cells, abundance of polymorphs with apoptotic nuclei (leucocytoclasia). Finely granular Warthin-Starry –positive and eosinophilic material is found deposited in close proximity with the blood vessels (*Figure 2*). On ultrastructure, trilaminaryshaped bacteria are identified in these areas (*Figure 3*).



Figure 1. Intranodal hyperaemic capillaries composed of plump epitheloid endothelium with polymorph nuclei (arrows); interstitial deposits are centrally located.

HE Magnification X 200



Figure 2. Warthin-Starry-positive intercellular material. Magnification X 1000



Figure 3. Ultrastructure trilaminary bacteria. Magnification X 10200

COMMENTS

BA has been first described in 1983 (4) as unusual vascular lesion in HIV-positive patients. The term BA has been applied to delineate capillary proliferation in association with bacterial species. These vascular proliferations can be single or multiple. However, the latter can be found also in viscera, including lymph nodes (6).

Regardless of the specimen, each of these vessels is layered with plump endothelial cells with polymorph nuclei. This finding is deceptive for a malignant vascular tumour such as Karposi sarcoma, which is most common in HIV patients. This malignant sarcoma, however, is composed of slit capillaries. spindle endothelial cells displaying polymorphic nuclei, hvaline globules, abundance of lymphocytes and plasma cells and few granulocytes in the interstitial tissue (1, 3, 5-6). Angiosarcoma is sharing a similar histological picture with atypical hyperchromatic and highly mitotic endothelium. Both malignant tumours lack the characteristic for BA vascular proliferation and the finely granular eosinophilic material in the interstitial tissue (5, 7).

Apart from excluding malignant blood vessel neoplasia, clinico-morphological diagnosis needs to take into consideration other benign lesions – haemangiomas and transformation of lymph node sinuses.

Epitheloid haemangioma (haemangioendothelioma) is layered with quite similar to BA endothelium, a phenomenon mostly related to presence of intracytoplasmic intermediary filaments, whereas in angiomatosis, the ultrastructure shows mitochondria, Golgi-complex and Weibel-Palade-bodies (1, 5).

Capillary haemangioma and pyogenic granuloma as well as all lymph sinus transformations display unremarkable endothelium, which sharply delineates these lesions from BA. Besides, these lesions lack leucocytoclastic features and acidophilic interstitial deposits.

Eosinophilic interstitial deposits are readily seen with silver impregnation methods (Warthin-Starry), which is a proof of their bacterial origin (2-3). The ultrastructure identifies trilaminar bacterial species (1, 5). These morphological methods are of specific value especially when the morphology is ambiguous.

Bacteria causing BA are *Rochalimaea henselae* and belong to Ricettsiae family (8,

9). They are Gram – negative and are responsible for a large number of vascular lesions in the immunocompromised such as peliosis in liver and spleen (8-11). It is considered that these bacteria secrete vascular growth factor (8). With respect to morphology and biochemical processes they are identical to cat-scratch disease (12). Biopsy specimens show similar distribution (perivascular and interstitial) Warthin-Starry-positive deposits. This had misled investigators into believing that BA was a systemic form of cat-scratch disease (3, 5). At present, it is well established that these disease are caused by two different bacteria which is why cat-scratch disease is presented with necrotic epitheloid -cell granulomas.

Bartonella bacilliformis is a similar microorganism causing identical vascular changes, manifested with skin lesions (verruga peruviana) of a systemic infectious disease (Oroya fever) endemic for countries like Peru. However, these bacteria are found intracellularly engulfed by macrophages. Serological tests are in favour of clinical diagnosis (3, 5, 6, 9).

Although benign, BA may be fatal for the organism (3). Erythromycin is the most effective treatment by now. To avoid recurrence a prolonged course of treatment is recommended -4 times for 2 weeks for a period of 2 years (5).

REFERENCES

- Chan J, Lewin K, Lombard Ch, et al. Histopathology of bacillary angiomatosis of lymph node. Am J Surg Path, 15(5): 430-437, 1991.
- 2. Mainguene C, Moreau A, Hofman P, et al. Peliose bacillaire course du Sida. Ann Pathol, 13(5): 341-345, 1993.
- LeBoit Ph, Berger T, Egbert B, etv al. Bacillary angiomatosis. Am J Surg Path, 13 (11): 909-920, 1989.
- 4. Stoler MH, Bonfgilio TA, Steigbigel rt, et al. An atypical subcutaneous infection associated with acquired immune deficiency syndrome. Am J Clin Pathol, 80: 714 - 718, 1983.
- 5. Cockerell C, LeBoit Ph. Bacillary angiomatosis: a newly characterized, pseudoneoplastic, infectious, cutaneous vascular disorder. J Am Ac Dematol, 22: 501-512, 1990.
- 6. Picard-Dahan C, Rober-Knebelmann C, Belaich S. L'angiomatose bacillaire. Ann Pathol, 12 (4-5): 284-290, 1992.

- 7. Schinella R, Greco A. Bacillary angiomatosis presenting as a soft-tissue tumor without skin involvement. Hum Pathol, 21 : 567-569, 1990.
- Reed J, Brigati D, Flyn S, et al. Immunocytochemical bacillary identification of Rochalimaea henselae in bacillary (epitheloid) angiomatosis, parenchymal bacillary peliosis, and persistent fever with bacteremia. Am J Surg Pathol, 16(7): 650-657, 1992.
- 9. Slater L, Welch D, Kyung-Whan M. Rochlimaea henselae causes bacillary

angiomatosis and peliosis hepatic. Arch Intern Med, 152: 602-606, 1992.

- Perkocha L, Geaghan Sh, Yen B, et al. Clinical and pathological features of bacillary peliosis hepatic in association with human immunodeficiency virus infection. N Eng J Med, 323 (23): 1581 -1586, 1990.
- Relman D, Loutit J, Schmidt T, et al. The agent of bacillary angiomatosis. N Eng J Med, 323 (23): 1573 - 1580, 1990.
- Wear D, Margileth A, Ficher G, et al. Cat scratch disease: a bacterial infection. Science, 221:1403 – 1405, 1983.