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Editorial

Ingrained and Jutting-Papillary Intralymphatic Hemangioendothelioma

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Papillary intralymphatic hemangioendothelioma is an extremely exceptional neoplasm appearing within diverse cutaneous surfaces or soft tissues. Neoplasm may emerge as an extra-vascular component of patch stage of Kaposi's sarcoma. Besides, ordinary angiosarcoma may depict focal morphological configurations akin to the neoplasm.

The subtype of hobnail hemangioendothelioma is comprised of components as retiform hemangioendothelioma and papillary intralymphatic hemangioendothelioma.

Additionally scripted as Dabska's tumour, neoplasm was initially described Maria Dabska in 1969 and denominated as malignant endovascular papillary angioendothelioma [1].

Histological assessment is pathognomonic for discerning the infrequent tumefaction. Pre-eminently low grade, neoplasm may manifest with metastasis to regional lymph nodes or distant metastasis into sites as pulmonary parenchyma.

Confined to cutaneous surfaces or subcutaneous tissues, lesion may be discerned within a diverse age range as from birth to 83 years. Generally, paediatric population is implicated. A specific gender predilection is absent [2,3].

The neoplasm is posited to configure as distinct intralymphatic neoplasia and is designated as papillary intralymphatic angioendothelioma. The neoplasm may arise within pre-existing benign vascular lesions as cavernous haemangioma [2,3].

Papillary intralymphatic hemangioendothelioma represents as a painless, gradually progressive tumour nodule. Typically, tumefaction is confined to dermis or subcutaneous tissue of extremities. Infrequently, lesion appears upon sites as the trunk or head and neck. Alternatively, deep seated locales as bone, brain, spleen, tongue, sot tissues or testis may be implicated [2,3].

Clinical representation is variable and the solid to cystic, preponderantly intra-dermal tumefaction may manifest as an ill-defined mass, plaque or nodule which protrudes into circumscribing tissue. Superimposed dermis is atrophic and expounds a pink, blue or violaceous hue [2,3].

Cutaneous tumours vary from two centimetres to three centimetres magnitude. However, lesions of up to 40 centimetres diameter may be expounded.

Neoplasm delineates cogent clinical symptoms as pain, ulceration or haemorrhage from site of lesion [3,4].

Upon microscopy, papillary tufts are layered by plump endothelial cells demonstrating epithelioid-like or histiocytic-like cellular countenance. Constituent papillary structures may be layered by atypical columnar endothelial cells. Subjacent vascular lumens appear distended. Foci of glomeruloid appearance may be enunciated [3,4].

Upon low power, neoplasm appears reminiscent of cavernous lymphangioma. Cuboidal or hobnail endothelial cells coating vascular structures typically demonstrate elevated nucleocytoplasmic ratio with apical nuclei protruding upon cellular surface and articulating 'hobnail' cells or 'matchstick' cells. Individual endothelial cells appear as cuboidal to tall, cylindrical cells impregnated with vacuolated cytoplasm and eccentric, hyperchromatic nuclei with nuclear bulging discernible upon the luminal margin [3,4].

Characteristically, innumerable interconnecting vascular channels delineating papillary projections or tuft like articulations, akin to renal glomeruli are exemplified. Layering endothelial cells demonstrate a classic 'hobnail' or 'matchstick' appearance on account of apical nuclei which protrude upon the cell surface. Also, layering cells enunciate elevated nucleocytoplasmic ratio [3,4].

Intravascular and perivascular infiltration of lymphocytes configure papillary projections pathognomonic of papillary hemangioendothelioma. Innumerable intraluminal lymphocytes appear to adhere to endothelial cells. Mitotic figures are discerned [3,4].

Ultrastructural examination depicts tumour cells incorporated with miniature storage granules pervaded with von Willebrand factor and P-selectin, thereby configuring Weibel Palade bodies. Tumour cells are pervaded with irregular nuclei, abundant perinuclear cytoplasmic filaments and innumerable pinocytotic vesicles. Constituent hyaline globules are comprised of electron-dense basement membrane substance [3,4].

TNM staging of soft tissue tumours [3,4]

Primary tumour

Head and neck

- TX: Tumour grade cannot be assessed.
- T1: Tumour magnitude ≤ 2 centimetre.
- T2: Tumour magnitude > 2 centimetres to ≤ 4 centimetres.
- T3: Tumour magnitude exceeding > 4 centimetres.
- T4: Tumour associated with invasion of adjoining structures:
- T4a: Tumour demonstrating infiltration of orbit, base of skull, dura, central compartment viscera, facial skeleton or pterygoid muscles.
- T4b: Tumour associated infiltration of brain parenchyma, encasement of carotid artery, prevertebral muscle or central nervous system via perineural dissemination.

Trunk and extremities

- TX: Tumour grade cannot be assessed.
- T0: No evidence of primary tumour.
- T1: Tumour magnitude ≤ 5 centimetres in greatest dimension.

- T2: Tumour magnitude > 5 centimetres and ≤ 10 centimetres in greatest dimension.
- T3: Tumour magnitude > 10 centimetres and ≤ 15 centimetres in greatest dimension.
- T4: Tumour magnitude > 15 centimetres in greatest dimension.

Abdomen and thoracic viscera

- TX: Tumour grade cannot be assessed.
- T1: Tumour is confined to organ of origin.
- T2: Tumour extension into circumscribing tissue beyond organ of origin:
- T2a: Tumour infiltrates serosa or visceral peritoneum.
- T2b: Tumour extension beyond serosa or into mesentery.
- T3: Tumour infiltrates adjacent organ.
- T4: Tumour demonstrates multifocal visceral involvement:
- T4a: Tumour is multifocal and confined to two sites.
- T4b: Tumour is multifocal and confined to three to five sites.
- T4c: Tumour is multifocal and implicates > 5 sites.

Retroperitoneum

- TX: Tumour grade cannot be assessed.
- T0: No evidence of primary tumour.
- T1: Tumour ≤ 5 centimetres in greatest dimension.
- T2: Tumour > 5 centimetres and ≤ 10 centimetres in greatest dimension.
- T3: Tumour magnitude > 10 centimetres and ≤ 15 centimetres in greatest dimension.
- T4: Tumour magnitude > 15 centimetres in greatest dimension.

Orbit

TX: Tumour grade cannot be assessed.

- T0: No evidence of primary tumour.
- T1: Tumour magnitude ≤ 2 centimetres in greatest dimension.
- T2: Tumour magnitude > 2 centimetres in greatest dimension in the absence of invasion of bony walls or globe.
- T3: Tumour of variable magnitude along with invasion of bony walls.
- T4: Tumour of variable magnitude along with invasion of globe or periorbital structures, eyelids, conjunctiva, temporal fossa, nasal cavity, paranasal sinuses or central nervous system.

Regional lymph nodes

- NX: Regional lymph nodes cannot be assessed.
- N0: Regional lymph node metastasis absent.
- N1: Regional lymph node metastasis present.

Distant metastasis

- MX: Distant metastasis cannot be assessed.
- M0: Distant metastasis absent.
- M1: Distant metastasis present.

Relevant prefixes

- m: Multiple.
- y: Adoption of preoperative radiotherapy or chemotherapy.
- r: Recurrent tumour stage.

Tumour cells are immune reactive to diverse vascular and lymphatic markers as CD34, vascular endothelial growth factor receptor 3 (VEGFR 3), CD31, D2-40, factor VIII related antigen or Von-Willebrand factor [5,6].

Papillary intralymphatic hemangioendothelioma requires segregation from neoplasms associated with intravascular proliferation as intravascular papillary endothelial hyperplasia (Masson's tumour), organized thrombi, epithelioid hemangioendothelioma, angiosarcoma, lymphangioma-like Kaposi sarcoma or retiform hemangioendothelioma.

Appropriate disease discernment necessitates comprehensive physical evaluation and assessment of regional and distant metastasis. Surgical procedures as orchidectomy emerge as a recommended therapeutic option [5,6].

Prognostic outcomes of the essentially low grade neoplasm are superior. However, tumefaction may exhibit a propensity for metastasis into regional lymph nodes or distant viscera as pulmonary parenchyma. Neoplasm may enunciate localized aggression and tumour invasion into deep seated anatomical structures as bone, skeletal muscle, fascia or abutting tendons [5,6].

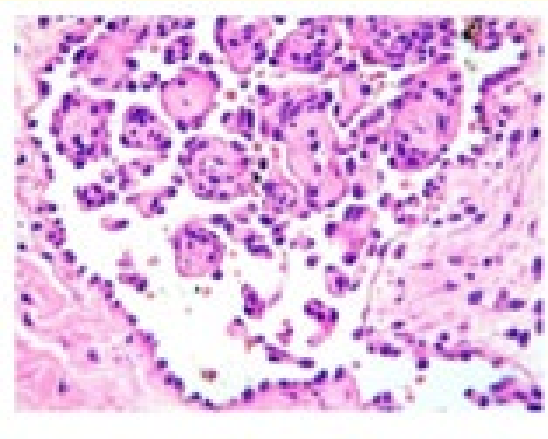

Figure 1: *Papillary intralymphatic hemangioendothelioma delineating papillary structures lined by endothelial cells with apical nuclei protruding upon cellular surface and elevated nucleocytoplasmic ratio. Intravascular and perivascular lymphocytes are observed [7].*

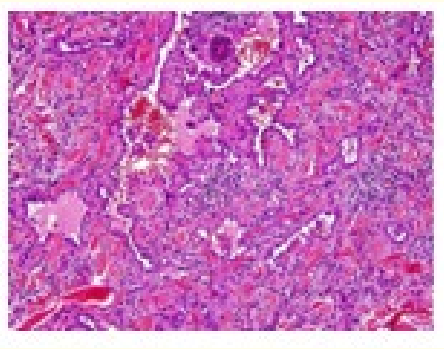

Figure 2: *Papillary intralymphatic hemangioendothelioma demonstrating papillary articulations lined by endothelial cells with apical nuclei protruding upon cellular surface and elevated nucleocytoplasmic ratio. Intravascular and perivascular lymphocytes are encountered [8].*

Bibliography

1. Dabska M. "Malignant endovascular papillary angioendothelioma of skin in childhood". *Cancer* 24 (1969): 503-510.
2. Al-Refai R., *et al*. "Locally invasive papillary intralymphatic angioendothelioma arising within a lymphatic/venous malformation". *American Journal of Case Reports* 24 (2023): e940602.
3. Li T., *et al*. "A huge spleen with papillary intralymphatic angioendothelioma". *Digestive and Liver Disease* 56.2 (2023): 365-366.
4. Kaplan GO., *et al*. "Papillary intralymphatic angioendothelioma: An extremely rare tumor". *Joint Diseases and Related Surgery* 32.1 (2021): 245-248.
5. Thadeu Santos Silva., *et al*. "Papillary intralymphatic angioendothelioma: Dabska tumor". *Anais Brasileiros de Dermatologia* 95.2 (2020): 214-216.
6. Bhatia J and Tadi P. "Endovascular papillary angioendothelioma". Stat Pearls International. Treasure Island. Florida (2023).
7. Image 1 Courtesy: Annais brasileiros de dermatologia.
8. Image 2 Courtesy: Semantic scholar.

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