Reply to 'Letter to the Editor' concerning a case report of endovascular papillary angioendothelioma (Dabska tumor) of the tongue

The authors would like to thank Dr Ide for his interest and comments on our case report (1).

He recommends the mature type of intravascular papillary endothelial hyperplasia (IPEH) as the pathologic diagnosis in our case. However, in the light of both clinical and pathologic features, we are convinced that the diagnosis we made is appropriate for endovascular papillary angioendothelioma (EPA).

Microscopically, the lesion was composed of a large number of ovoid cells with quite dense proliferation and micropapillary growth, and encapsulated with thin fibrous connective tissue closely resembling the vascular wall. Minimal cytologic atypia and only rare mitotic figures were seen in these proliferative cells. It is clear from the immunohistochemical analysis that the proliferative lesion is derived from the endothelium. We considered that the lesion showing dense proliferation is unlikely to classify as hyperplastic lesions. However, he claims that our case is more feasible as a mature form of IPEH rather than as any kind of EPA or EPA-like tumor. Accordingly, for the differential diagnosis between tumor and hyperplastic or reactive lesions, we performed immunohistochemical analysis in various cell-proliferative markers; proliferating cell nuclear antigen (PCNA), Ki-67 and DNA Topoisomerase II α (DNA Topo II α; Novocastra Laboratories, Newcastle-upon-Tyne, UK); in this lesion together with our previously experienced 13 cases of IPEH. As a result, PCNA was strongly positive and labeling index (LI) was 32.4, and Ki-67 LI and DNA Topo II α LI was 16.3 and 14.9 in the proliferative cells of the present case. In contrast, LIs in IPEHs were 5.6 ± 2.0, 1.7 ± 1.1, and 1.4 ± 0.7, respectively, that is, cell proliferative activity in vascular hyperplastic lesion remained low. As shown by these facts, this proliferative lesion is undoubtedly a vascular tumor including EPA.

On the other hand, there is considerable difference between our and his opinion as to the existence of capsule in EPA. The discrepancy may result from the difference of origin in each tumor. We speculate that the origin of the tumor is the endothelium of the capillaries, as described in the majority of previous reports. Therefore, the capsule in tumor may become ill-defined. In contrast, thin fibrous capsule microscopically recognizable in our case is considered to correspond to a remnant of vascular wall. If our case corresponds to the early stage of EPA, the capsule in the lesion may become ill-defined as an outgrowth with the course of the lesion.

We think that a key finding in the histologic feature of EPA is the existence of glomerulus-like appearance. This finding is the most frequently detectable feature in EPA, according to this peculiar findings is the most important feature in pathologic diagnosis of EPA. In the present case, the microscopic examination revealed glomerulus-like

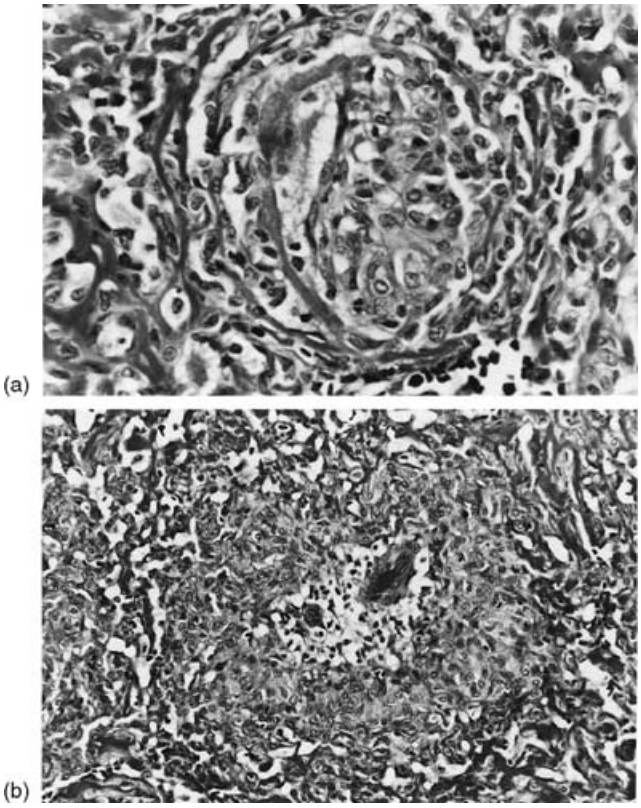

Figure 1 (a) Showing glomerulus-like appearance. (b) Lymphocytes aggregation was observed in the central area of glomerulus-like appearance.

appearance everywhere in the lesion (Fig. 1a) and hobnail-like pattern (Fig. 2d in (1)). Moreover, lymphocytes aggregation was observed in the central area of some glomerulus-like appearance (Fig. 1b). However, we could not find out the tuft and true rosette formation as one of the findings in EPA that Dr Ide pointed out. This difference may be linked with the degree of maturation in this tumor.

Dr Ide described that the term hemangioendothelioma should at least be restricted to low-grade malignant or borderline vascular tumor (2, 3), but we cannot necessarily favor his opinion. Basically, the term Dabska tumor (DT) in the strict sense implies a malignant neoplasm, which originates in skin or subcutaneous tissue of the infant, indicating malignant EPA. DT is characterized by unusual distribution in the predilection of age, namely the distribution is biphasic in juvenile people of 18 years and under (approximately 60%) and adults of 30 years and over (approximately 40%) (4). It is speculated that the former is almost malignant, and the latter is largely benign lesion, including borderline and low-grade malignancy. There is a similarity in the clinical characteristics except for the age distribution and pathologic findings. It is well recognized that the differentiation between benignancy and malignancy in vascular tumors is often difficult because of a conflict between the histologic and the biologic characters. We also feel that the differential

diagnosis in the previous reports of EPA might not necessarily be made with proper interpretation.

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