

## *Case report*

# Central intravascular papillary endothelial hyperplasia of the mandible

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**Summary.** Light and electron microscopic findings in papillary endothelial hyperplasia of the mandible in a 49-year-old female are reported. The endothelial cell-lined papillary projection into a cystic lumen was examined by light microscopy and characteristic features of the endothelial cells were found by electron microscopy. Factor VIII-related antigen was demonstrated in the endothelial cells by the immunoperoxidase technique.

**Key words:** Intravascular endothelial hyperplasia – Mandible

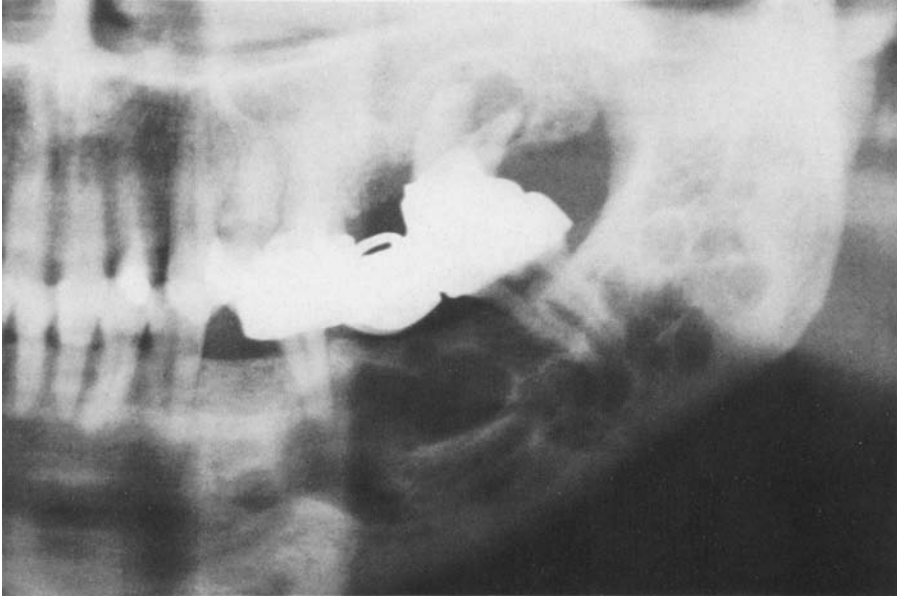
## Introduction

Intravascular endothelial hyperplasia was first described by Ewing in 1922. It is a rare disease, showing predilection for the subcutis of the head and neck and distal parts of the extremities (Hajdu 1979). There are two reports of cases in the oral region (Heyden et al. 1978; McClatchey et al. 1978).

This article records the light and electron microscopic features of a lesion of the mandible and demonstration of the Factor VIII-related antigen in the lesion.

## Case report

A 49-year-old Japanese female was admitted to the 1st Oral Maxillo-facial Surgical Clinic on July 29, 1981, for evaluation of a lesion in the left mandible. Her family history was non-contributory. About 26 years previously, she had had an operation for an ovarian tumor. Two days before admission to this clinic, the patient's dentist had discovered a radiolucent



**Fig. 1.** Rentgenogram of the left mandible showing a multilocular radiolucent region

lesion in the left mandible. On admission to this clinic no other abnormality was found in the oral cavity and the mucosa covering the surface of the lesion appeared normal. On X-ray examination, the region from the first premolar to the second molar on the left of the mandible appeared multilocularly radiolucent (Fig. 1). This lesion was removed on Nov. 12. The post operative course was uneventful, and one year after the operation, the patient is well, with no evidence of recurrence.

### **Materials and methods**

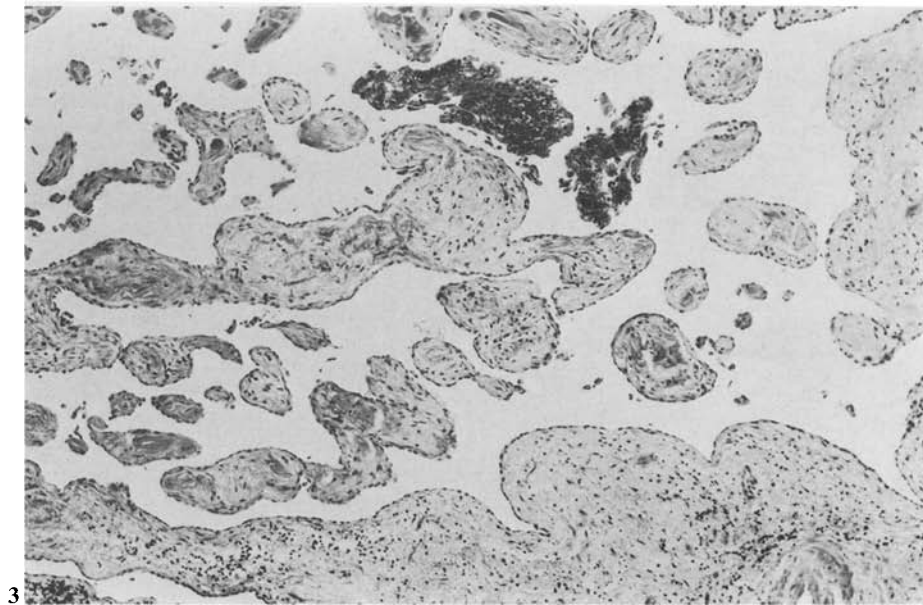
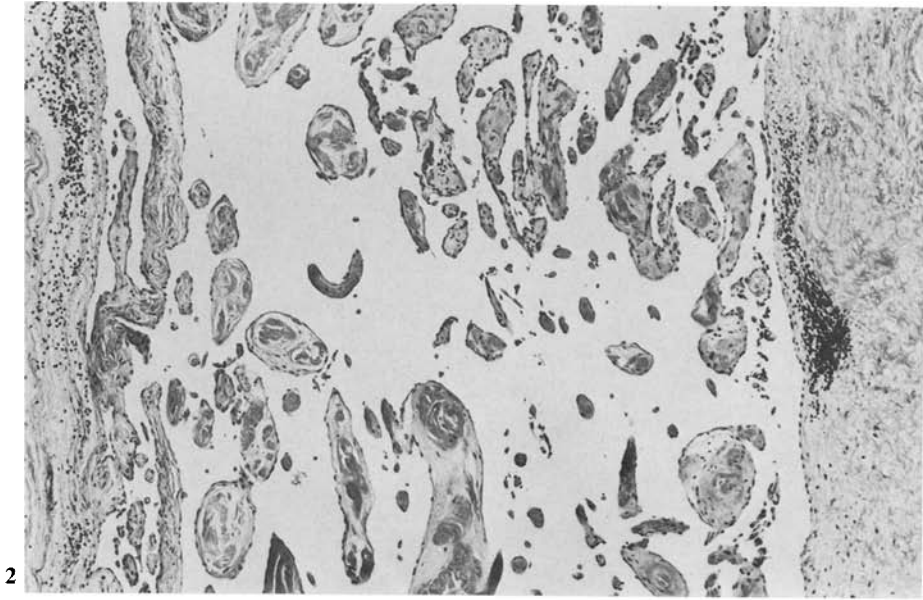
For light microscopic examination, the tissue was fixed in 10% neutral phosphate-buffered formalin and embedded in paraffin. Sections were stained with haematoxylin and eosin, Verhoeff-van Gieson, phospho-tungstic acid haematoxylin (PTAH) and silver stain for reticulin.

For electron microscopic examination, small pieces of the formalin fixed materials were post-fixed in 2% osmium tetroxide, and embedded in Epon 812. Ultrathin sections were stained with uranyl acetate and lead citrate and examined with a Hitachi H-500 electron microscope.

Factor VIII-related antigen was demonstrated by the immunoperoxidase technique (DAKO PAP KIT) in formalin-fixed, paraffin-embedded tissue.

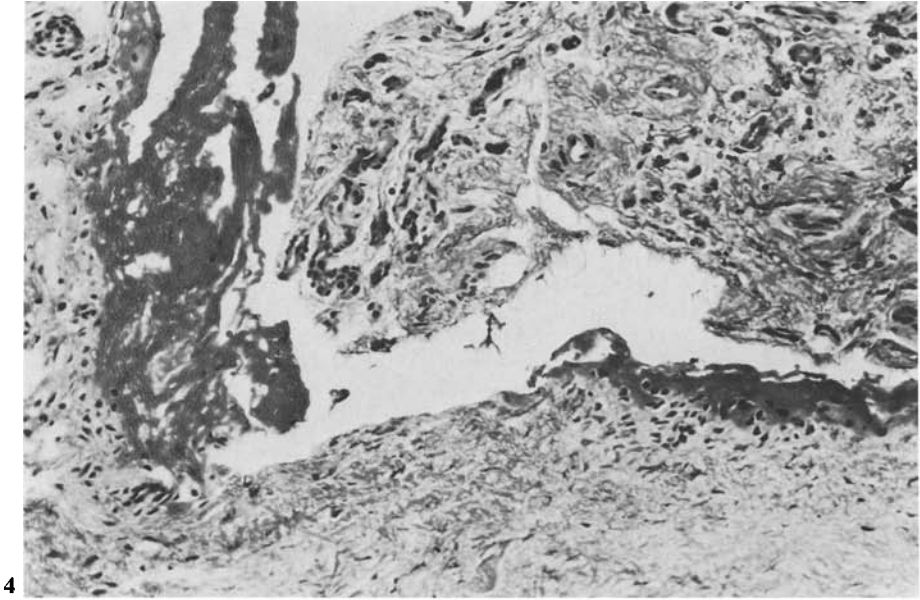
### **Results**

Grossly, the excised lesions appear to be fibrous tissue like that of a cyst wall. On light microscopy, numerous irregular papillary projections are seen within the cystic lumen (Fig. 2). These papillary structures are covered by a single layer of flat or swollen endothelial cells (Fig. 3). There is no cellular pleomorphism and no mitotic figures are seen in endothelial cells. In a few places in preparations stained with PTAH, small amounts of blue fibrin are observed adhering to the internal surface of blood vessels in the fibrous

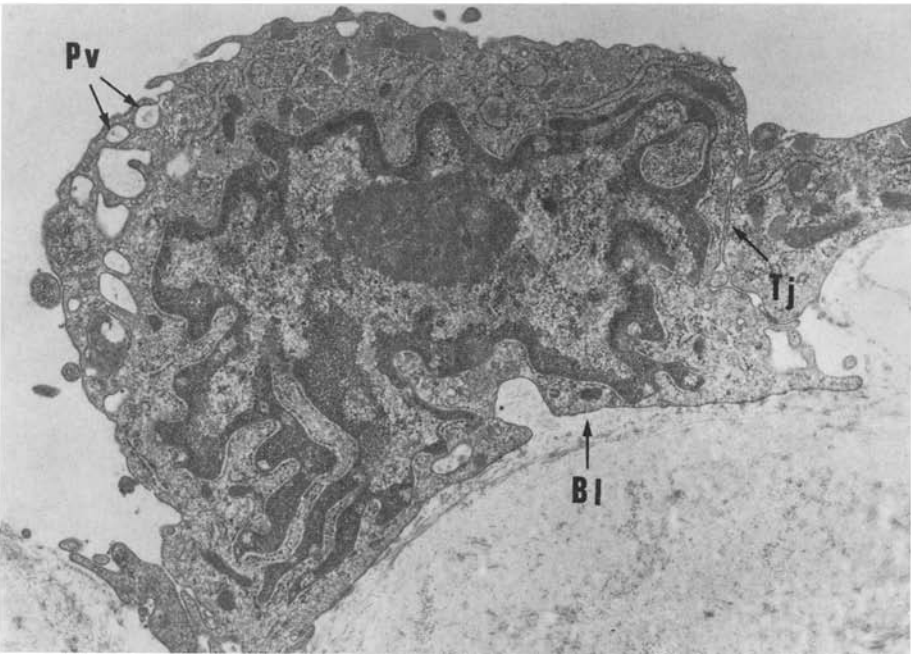


**Fig. 2.** Photomicrograph of the lesion. A papillary projection occurring within a dilated vascular space. (Haematoxylin and eosin stain. Original magnification,  $\times 40$ )

**Fig. 3.** A single layer of endothelial cells cover the papillary structures. (Haematoxylin and eosin stain. Original magnification,  $\times 100$ )



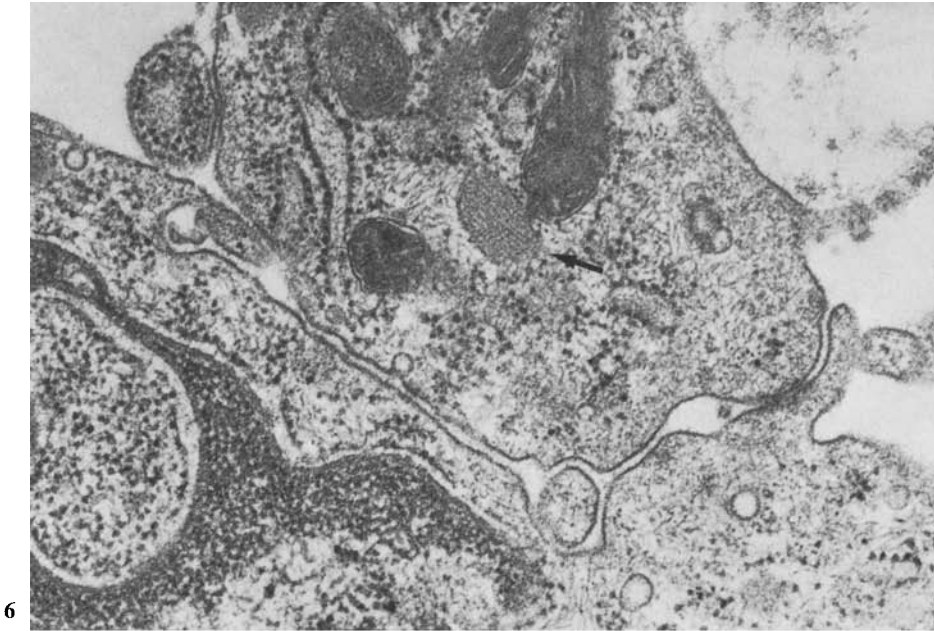
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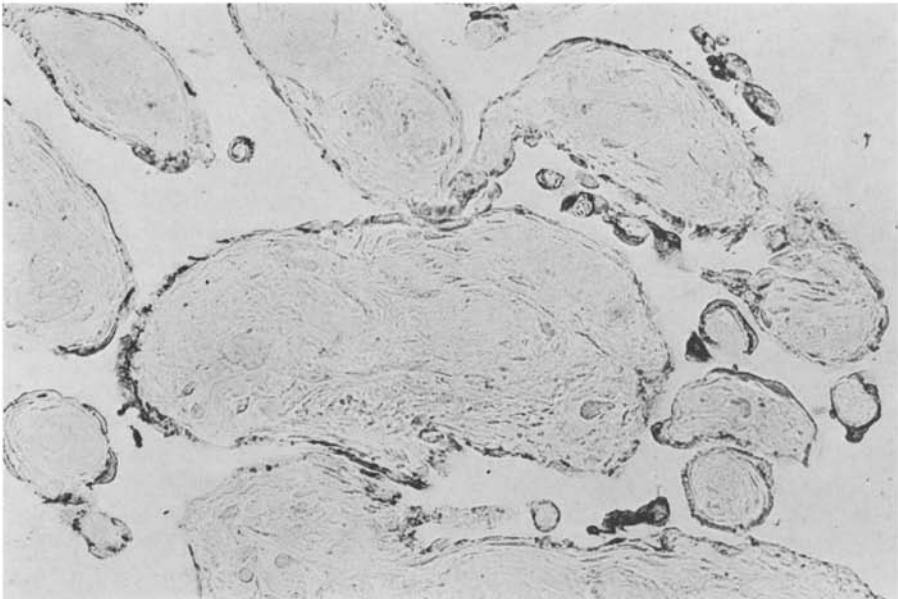
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**Fig. 4.** Microphotograph of blood vessels in the fibrous tissue of the luminal wall. Adhesion of fibrin to the surface is seen. (Haematoxylin and eosin stain. Original magnification,  $\times 200$ )

**Fig. 5.** Electron-micrograph of the endothelial cells lining the papillary projection. The cells have irregular nuclei, a few micro-pinocytotic vesicles (*Pv*) and organelles, a basal lamina (*Bl*) and tight junctions (*Tj*). (Magnification,  $\times 16,000$ )



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**Fig. 6.** An endothelial cell at higher magnification. A Weibel-Palade body (*arrow*) is seen in the cytoplasm. (Magnification,  $\times 66,500$ )

**Fig. 7.** Preparation stained with immunoperoxidase for factor VIII-related antigen. A marked positive reaction is seen in the endothelial cells. (Original magnification,  $\times 200$ )

connective tissue of the luminal wall (Fig. 4). No elastic or reticulin fibers are seen in the fibrous connective tissue of the luminal wall, but a small amount of irregular-shaped material staining with Verhoeff's elastic stain is found in fibrous tissues in the papillary projection. A few smooth muscle fibers are seen scattered in part of the fibrous connective tissue of the luminal wall in preparations stained with PTAH.

Electron microscopy show that fibrous papillary projection is lined by a single layer of flat or plump endothelial cells. The nuclei of these cells are comparatively large and irregular in shape. A few micro-pinocytotic vesicles, mitochondria, and free ribosomes and some rough-surfaced endoplasmic reticulum are observed in the cytoplasm of these cells (Fig. 5). Only a few microfilaments and small, oval dense bodies (Weibel-palade bodies) showing cross striation are found in the cytoplasm (Fig. 6). The endothelial cells are characterized by tight intercellular junctions and are separated from the collagen fibrils by thin basal lamina.

The endothelial cells lining the papillary projection give a strong positive reaction for Factor VIII-related antigen, seen as a fine granules in their cytoplasm (Fig. 7).

## Discussion

The lesion described in this paper has various names (Hajdu 1979), and was recently named intravascular papillary endothelial hyperplasia by Clearkin and Enzinger (1976). This lesion is rare: Dabska (1969) reported 6 cases, Clearkin and Enzinger (1976) reported 44 cases, Kuo et al. (1976) reported 17 cases, Hajdu (1979) reported 8 cases and Amerigo and Berry (1980) reported 24 cases. The lesion occurs predominately in adult females, and shows predilection for the subcutis of the head and neck. Our case was a 49-year-old female and the lesion was located in the mandible.

Endothelial cell-lined papillary projections into the lumen of dilated blood vessels or cavernous vascular spaces have been described as the most characteristic histological feature of the intravascular papillary endothelial hyperplasia. The histological features of the present lesion were identical with those described by others.

By electron-microscopy, the endothelial cells are characterized by the presence of Weibel-palade bodies, numerous pinocytotic vesicles, tight junctions and a basal lamina (Erlandson 1981). The endothelial cells lining the papillary projection into the lumen in our case also showed these features.

Factor VIII-related antigen has been shown to be synthesized by endothelial cells, and this antigen has been used in differentiation of vascular lesions (Hoyer et al. 1973; Mukai et al. 1980; Sehested and Hou-Jensen 1981). In our case, the endothelial cells lining the papillary projection gave a strong positive reaction for factor VIII-related antigen.

The similar papillary lesion was described as a papillary fibroelastoma in the heart (Abu Nassar and Parker 1971; McAllister and Fenoglio 1978). Although the histological structures were slight different in the papillary endothelial hyperplasia and the endocardial papillary fibroelastoma, it was

thought that the both papillary lesions developed from the thrombi (Salyer et al. 1975; Salyer and Salyer 1975; Fishbein et al. 1975). In this case, the finding of fibrin adhering to the wall of blood vessels in fibrous tissue of the luminal wall has been considered to be related to thrombosis rather than to a neoplastic process to its histogenesis.

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