Endovascular Papillary Angioendothelioma (Dabska Tumor) of Bone

TOMITAKA NAKAYAMA, MD*; MIZUKI NISHINO, MD†; KOSHO TAKASU, MD‡; KATSUMI HAYAKAWA, MD‡; JUNYA TOGUCHIDA, MD§; CHIAKI TANAKA, MD*

Endovascular papillary angioendothelioma, first described by Dabska, is a rare vascular tumor characterized by anastomosing vascular channels with intraluminal papillary proliferations of endothelial cells. This low-grade malignant tumor, which occasionally metastasizes, usually occurs in children and involves the skin or subcutaneous tissue. A few cases involving other tissues and adults, as well as bone, have been reported.

This article presents an intraosseous endovascular papillary angioendothelioma, which was successfully treated surgically.

CASE REPORT

A 39-year-old woman presented with right knee pain of 1 year’s duration. Pain worsened at night, and was significantly reduced by nonsteroidal anti-inflammatory drugs. No swelling was observed, and tenderness was demonstrated over the medial aspect of the right distal thigh.

Plain radiographs showed dense, continuous periosteal thickening on the medial distal metaphysis and epiphysis of the femur, with a small radiolucent area. Figure 2: CT demonstrates an intraosseous radiolucent lesion with a sclerotic margin. Figure 3: Coronal low intensity T1-weighted (A) and high intensity T2-weighted (B) MRIs. The lesion is surrounded by a hyperintense area on T2-weighted MRI, which was demonstrated in bone and extraosseous soft tissue (C).

From the Departments of *Orthopedic Surgery, †Radiology, and ‡Pathology, Kyoto City Hospital; and the §Institute for Frontier Medical Sciences, Kyoto University, Kyoto, Japan.

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Reprint requests: Tomitaka Nakayama, MD, Dept of Orthopedic Surgery, Graduate School of Medicine, Kyoto University, Shogon-Kawahara-cho 54, Sakyo-ku, Kyoto 606-8507, Japan.
metaphysis and epiphysis of the femur, and a small radiolucent lesion within it (Figure 1). Computed tomography (CT) demonstrated a 1-cm, well-circumscribed, radiolucent intraosseous lesion with an accompanying thin sclerotic margin (Figure 2). Magnetic resonance imaging (MRI) revealed a hypointense lesion on T1-weighted images and hyperintense on T2-weighted images (Figure 3). Marked thickening of the periostea was noted. Prominent high-signal intensity on T2-weighted images was present in the surrounding intraosseous region and extraosseous soft tissue and was considered to be peritumoral edema (Figure 3). Technetium bone scan showed abnormal accumulation in the medial femoral metaphysis (not shown).

From the clinical and radiographic evaluations, the lesion was diagnosed as osteoid osteoma. The patient underwent curettage of the lesion. A small, soft brownish mass was resected; however, the margin of the lesion was ill-defined. Pain resolved postoperatively.

Histologic examination of the resected specimen revealed a lesion consisting of a proliferation of spindle cells with numerous vascular spaces, which contained papillary tufts of hobnail endothelial cells.

Re-excision with a wider margin was performed 3 months postoperatively. The lesion was resected with approximately 2 cm of surgical margin and the defect formed after the resection was filled with autologous fibula graft. No residual tumorous proliferation was found in microscopic examination of the re-resected specimen. The patient was pain-free with no signs of recurrence 50 months postoperatively.

**DISCUSSION**

Most endovascular papillary angioendotheliomas reported have involved the skin or subcutaneous tissue.2,4,5 Recently, the proliferative endothelial cells in this lesion have features of lymphatic vessels, which was suggested by the immunohistochemical analysis.2,5 Only one known case involving bone has been reported.3 Endovascular papillary angioendothelioma was first described as a distinctive subtype of malignant vascular neoplasm by Dabska.1 Although two of six patients presented with lymph node metastases in the initial report, clinical behavior of this condition is not well understood because the cases reported thereafter showed few apparent distant metastases.

In larger series, Fanburg-Smith et al2 described no metastasis in 8 patients, and Weiss and Goldblum4 reported 4 local recurrences and 1 regional lymph node metastasis in 10 patients. The difference of tumor prognosis between intraosseous lesions and lesions of conventional soft-tissue origin is unknown, and no consensus exists concerning the proper treatment of intraosseous lesions.

In the case reported previously, curettage was the treatment of choice providing a successful outcome at 1-year follow-up. In the present case, wide resection, which is the most common procedure for malignant bone tumor, was chosen. An accumulation of knowledge about this rare condition is essential for standardization of treatment strategy.

**REFERENCES**