CASE REPORT

Angiosarcoma arising within a Malignant Endovascular Papillary Angioendothelioma (Dabska tumor)

Zbigniew Antosz [1], Maciej Zaniewski [2], Jacek Kostecki [2], Ryszard Poreba [3]

1 Department of Pathology, District Specialist Hospital, Tychy, Poland
2 Department of Surgery, Medical University of Silesia, District Specialist Hospital, Tychy, Poland
3 Department of Gynecology Obstetrics, Medical University of Silesia, District Specialist Hospital, Tychy, Poland

Correspondence to: Jacek Kostecki, MD.
Department of Surgery, District Specialist Hospital,
ul. Edukacji 102, 43-100 Tychy, Poland.
tel: +48-32-325 42-97; e-mail: kosteckj@op.pl

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Abstract

We report an angiosarcoma arising within a malignant endovascular papillary angioendothelioma (Dabska tumor) in soft tissue of the upper thigh/buttock of a 42-year-old woman. Although neoplastic progression within a vascular tumor of an existing low-grade lesion into DT has been described so far, we seem to be the first to report transformation of DT into an angiosarcoma.

INTRODUCTION

The Dabska tumor is a rare vascular neoplasm also known as malignant endovascular papillary angioendothelioma. There have been 31 cases reported in literature to this day.

Although Maria Dabska, who originally described the condition in 1969, found it only in infants and young children (6 cases), so far there have also been 13 patients aged 18–83, reported to have DT (Schwartz *et al.* 2000). Affecting primarily the skin or subcutaneous tissue, DT was found to involve the tongue, spleen, and bone as well (Takaoka *et al.* 2003; Katz *et al.* 1988; McCarthy *et al.* 1999).

Histologically, it is characterized by papillary proliferation of endothelial cells within vessel lumina, accompanied by intra- and peri-vascular lymphocyte infiltration. DT shows some traits of malignancy like increased mitotic activity, focal necrosis and potential for metastasizing (most frequently to regional lymph nodes), but because of its relatively good long-term prognosis the lesion was often regarded as a low grade vascular tumor.

The case we are reporting is, to our knowledge, the first one in which transformation of DT into angiosarcoma with accompanying pulmonary and liver metastases has been confirmed.

CASE REPORT

A 44-year old female was admitted to the Department of General and Vascular Surgery in our hospital for elective surgical treatment of a tumor in the left buttock area. The tumor had first been noticed by the patient about 5 years before and had been periodically disappearing and reappearing since that time. For the last 3 months prior to admission it had been growing and becoming painful, especially in a sitting position. The patient also had a history of varicose veins and had undergone appendectomy 20 years earlier. She was preg-

nant 5 times and had 4 spontaneous deliveries. Family history was unremarkable apart from varicose veins. Physical examination revealed a tumor, about 5 cm in diameter, located within skin on the border of the left buttock and thigh. It was soft, elastic, bluish in coloration and presented no tenderness on palpation. Skin above the lesion was unaffected and its temperature was normal. Laboratory findings like urinalysis, blood cell count and biochemical parameters were within normal range. Local excision was performed in conduction anesthesia with following Redon drainage.

Histopathological examination of the mass removed at surgery, revealed a 2.9 × 1.8 × 2.5 cm cystic tumor within subcutaneous tissue. It consisted mainly of both venous-like and lymphatic-like spaces lined predominantly by flattened endothelial cells giving appearance entirely consistent with an ordinary hemangioma.

However, within the ectatic, hemangioma-like vascular lumina one could observe papillary structures, locally forming glomerulus-like aggregations. Papillary structures were formed by solid nests of cuboidal cells containing central, thin, hyalinized collagen core. These cells had evident hobnail-like appearance, with apical nuclei projecting into the lumen and basally oriented cytoplasm. Focally, intracytoplasmatic vacuoles were seen and intravascular and perivascular lymphocytic inflammatory infiltrate was present (Figure 1).

After 8 months, because of local recurrence, the patient underwent consecutive tumor resection in another center.

At operation a 2.5 cm cystic tumor was found in subcutaneous tissue. Histological studies showed both ordinary hemangioma and locally the same hobnailed appearance like in the first lesion suggesting DT (Figure 2)

About 10 months after the second resection, another recurrence took place. This time, apart from the primary tumor CT examination demonstrated 7 tumors 1.3–1.4 cm in diameter located in the right lung and 2 tumors 0.6cm and 0.4 cm in the left lung. All of them were diagnosed as metastases. CT of the liver also revealed 7 tumors, the biggest one 2.5 cm in diameter. Histopathological examination of the excised primary tumor (9 × 7 cm) showed locally persistent traits of DT as well as evident angiosarcoma with infiltration of soft tissue and muscle of the buttock (Figure 3).

Immunohistochemical stains were performed on the paraffin-embedded tissues from the first and the last lesion. Tumor cells were positive for CD34 and weakly positive for factor VIII-related antigen.

The patient died three years after having been diagnosed with DT.

DISCUSSION

Malignant endovascular papillary angioendothelioma, also known as the Dabska tumor, was first described in 1969, when Dabska reported six cases (Dabska 1969).

Enzinger and Weiss in 1995 referred to DT as a form of low-grade angiosarcoma occuring predominantly in children (Enzinger & Weiss 1995).

In a few cases of mainly adult patients, the diagnosis of DT or DT-like tumor has been challenged, because there are also other vascular tumors known to have

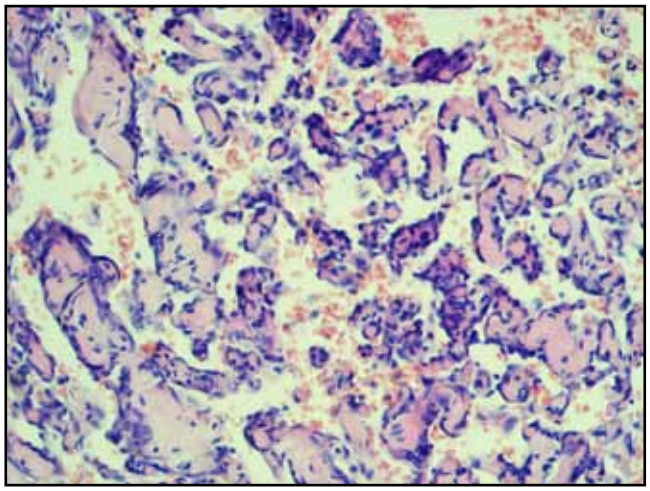

Fig. 1. Scale 1:400, hematoxylin/eosin stain.

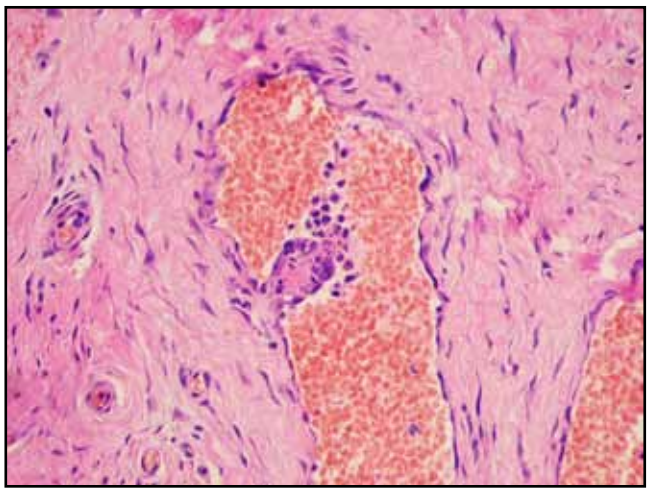

Fig. 2. Scale 1:400, hematoxylin/eosin stain.

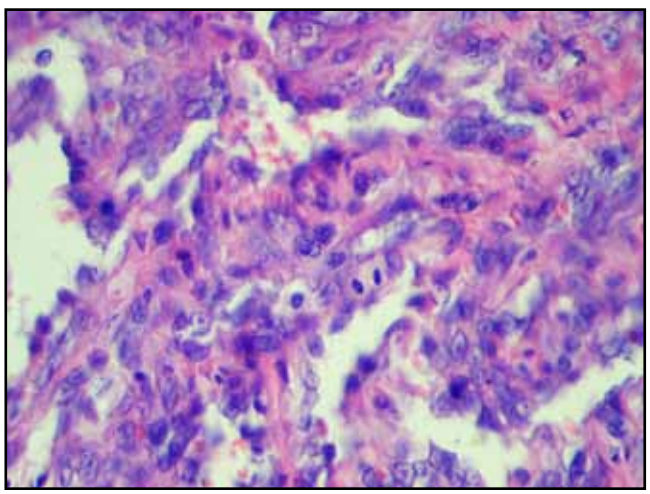

Fig. 3. Scale 1:400, hematoxylin/eosin stain.

similar histolological appearance (Allen *et al.* 1992; Requena & Sangueza 1998; Miyachi & Imamura 1981; Fukunaga *et al.* 1995).

DT typically presents as a violaceous, pink or bluish black tumor growing slowly within dermis or subcutis up to 40 cm in diameter and sometimes fluctuating in size (Fanburg-Smith *et al.* 1999; Dabski 1993; Enzinger & Weiss 1995). Although the head and extremities seem to be most commonly affected sites, the following cutaneous locations have also been described: palm, forearm, heel, knee, cheek, temple, pinna of the ear, neck, buttock, abdomen and upper back. Development of DT often results in degeneration of its underlying vascular tumor.

Reported cases of DT include: focal degeneration within a deep cavernous hemangioma of leg soft tissue in an 8-year-old girl (Argani & Athanasian 1997), lesion within an angiomatous vascular malformation on the buttocks of a boy with the Kasabach-Merritt syndrome (Quecedo *et al.* 1996), and a part of subcutaneus cavernous hemangioma fixed to underlying fascia on the forearm of a 6-year-old boy (Patterson & Chandra 1985).

Histologically, the lesion resembles a cavernous lymphangioma at low power, with large dilated vascular channels and an infiltrate of small lymphocytes. The channels are predominantly lined by plump endothelial cells which have characteristic hobnail-like or matchstick-like appearance with apically placed nuclei projecting into the vessel lumen.

Unique for DT is a pattern of papillary structures, sometimes similar to renal glomeruli, with hyalinized collagenous cores, which develop within the lumen and are lined with atypical columnar endothelial cells.

There remains some controversy regarding DT's malignancy though currently it is more and more often classified as a tumor of intermediate biologic malignancy (Enzinger & Weiss 1995).

Two of Dabska's initial cases were associated with metastases to regional lymph nodes and one to lung. One patient subsequently died of the disease. Dabski reported lung metastases and death of one of the initial patiens 6 years after local resection and regional lymphadenectomy (Dabska 1969). In our case we observed widespread pulmonary and liver metastases.

Complete surgical excision is the treatment of choice. Prognosis is good following complete surgical excision of the primary lesion, although spread to regional lymph nodes is possible (Enzinger & Weiss 1995).

Our case proves that DT, once regarded as a low-grade lesion has the potential to transform into angiosarcoma which makes it a tumor of intermediate malignancy.

Therefore, the Dabska Tumor should never be overlooked.

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