CLINICAL CASE

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Hemangiopericytoma of the Hand – Case Report

Obłoniak dłoni – opis przypadku

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Abstract

An 18-year-old boy developed a *hemangiopericytoma* in the metacarpal region, after the second excision of recurrent capillary hemangioma in the same location. There was no recurrence of the tumor 6 years after wide margin surgical excision (**Adv Clin Exp Med 2006, 15, 1, 171–173**).

Key words: *hemangiopericytoma*, capillary hemangioma, hand neoplasm.

Streszczenie

U 18-letniego chłopca rozwinął się obłoniak w obrębie śródręcza (okolicy metakarpalnej), po drugim wycięciu nawracającego naczyniaka włośniczkowego w tym samym miejscu. Po chirurgicznym usunięciu z dużym marginesem nie odnotowano nawrotu przez 6 lat (**Adv Clin Exp Med 2006, 15, 1, 171–173**).

Słowa kluczowe: obłoniak, naczyniak włośniczkowy, nowotwór ręki.

Hemangiopericytoma is an uncommon, slowly growing vascular tumor which usually develops in soft tissues [1, 2]. The tumor is composed of Zimmerman's pericytes, and biologically it can be benign or malignant, however, it is difficult to predict tumor's behavior based on histopathology alone [3, 4]. For many years *hemangiopericytoma* has been a controversial entity and was classified with hemangioma. The WHO International Classification of Soft Tissue Tumours in 1969 included both forms of *hemangiopericytoma* [5]. The authors report another case of *hemangiopericytoma* of the hand located in metacarpal region.

Case Report

An 18-year-old boy was hospitalized for a 6-cm mass located between adductor policis and flexor policis brevis of the right hand.

The mass appeared soon after the second excision and slowly increased to its current size. Patient had been operated 4 and 2 years before due to recurrent mass at the same location. Both times capillary hemangioma was recognised.

At operation the mass was found to be ill defined from surrounding tissues and was closely adherent to the first metacarpal bone. Wide excision of the tumor was performed including resection of adjacent fragment of the first metacarpal bone. Meticulous hemostasis was accomplished and the wound was closed.

Gross examination showed bulky mass rubbery in texture and soft-red appearance. In light microscopy there were seen relatively uniform, round to ovoid and occasionally spindle-shaped cells, tightly packed among numerous branching thin-walled blood vessels.

Mitotic activity was low and ranged from 0 up to 3 mitoses per 10 high power fields. Sparse areas of degenerative changes caused by hemorrhage were seen. Silver reticulin preparation revealed dense meshwork of reticulin fibers enveloping the individual tumor cells or small clusters of the cells, which were closely related to the reticulin sheath of the vascular channels. Immunoperoxidase stains for factor VIII-related antigens and for

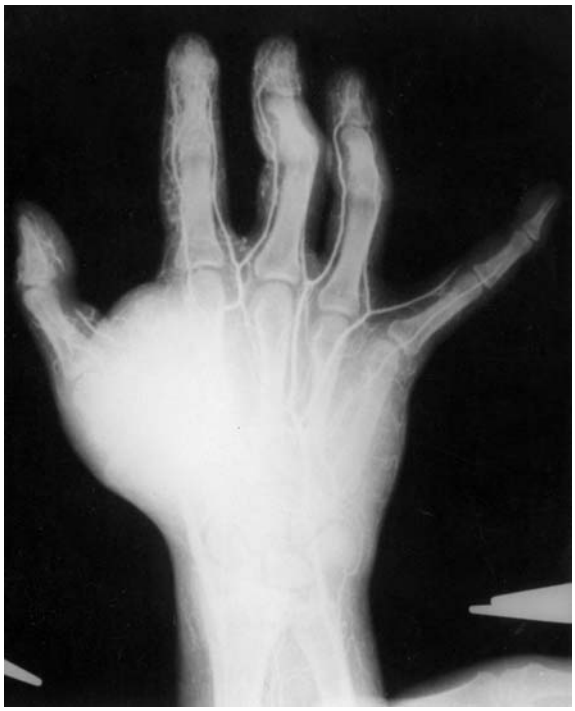

Fig. 1. X-ray picture of the tumor located in metacarpal region

Ryc. 1. Radiogram nowotworu umiejscowionego w obrębie śródręcza

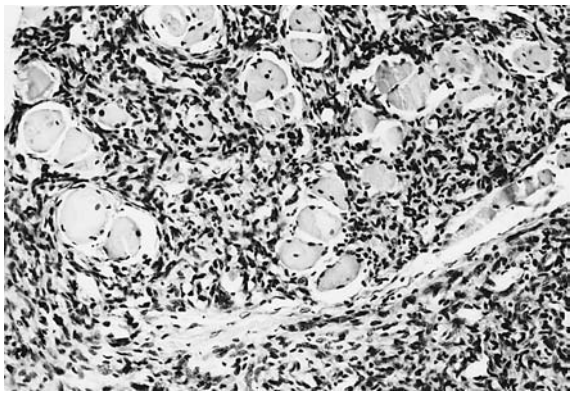

Fig. 2. Capillary hemangioma of metacarpal region, histological picture of the first excised tumor (the same patient at 14 yrs of age). Stain HE, magnification 120×

Ryc. 2. Naczyniak włośniczkowy śródręcza, obraz histologiczny pierwszego usuniętego nowotworu (ten sam pacjent w wieku 14 lat). Barwienie HE, powiększenie 120×

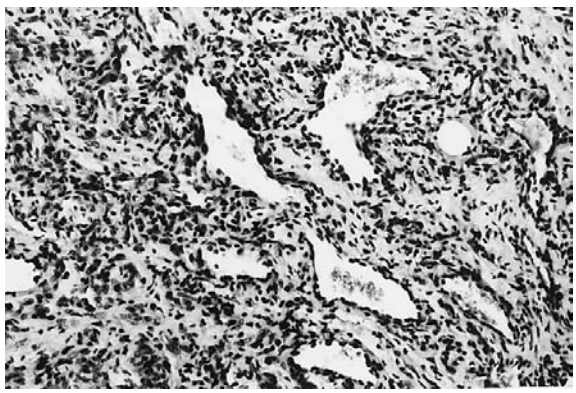

Fig. 3. Intramuscular capillary hemangioma (the same patient at 16 yrs of age), histological picture of the second tumor excised from metacarpal region. Stain HE, magnification 120×

Ryc. 3. Wewnątrzmięśniowy naczyniak włośniczkowy (ten sam pacjent w wieku 16 lat), obraz histologiczny drugiego nowotworu usuniętego ze śródręcza. Barwienie HE, powiększenie 120×

CD34 revealed positive reaction of the vascular endothelial cells. The tumor cells were positive for vimentin, CD34 and were negative for actin and S-100. The tumor tissue was closely adherent to the first metacarpal bone but without infiltration of the periosteum.

The function of the hand was fully preserved. Four years after the third operation the patient has been free of the disease.

Discussion

Benign vascular neoplasms are common and form a large group of hemangiomas with distinctive clinicopathologic features [6]. Almost one-third of all babies have a hemangioma of some type due to developmental abnormalities. Benign vascular neoplasms are usually classified according to their cell lineage of differentiation (e.g. endothelial, glomus cell or pericytic differentiation). In hemangioma, the main element that proliferates is an endothelial cell. *Hemangiopericytoma* (HPC) is relatively rare among soft tissue tumors and it is a tumor primarily of adults whose median age is about 45 years, although they are seen at any age from infancy to the last decades of life and at any site. Most of them are benign tumors, but local recurrence or distant spread eventuates in about 20% of patients. Large HPC localized in abdominal cavity might be associated with clinically observed hypoglycemia [7]. The diagnosis of HPC is debated, because the HPC-like formation may be seen in variety of neoplasms, including "true" HPC, synovial sarcomas, mesenchymal chondrosarcomas, infantile fibrosarcomas, malignant fibrous histiocytomas, malignant peripheral nerve sheath tumors, leyomyosarcomas, endometrial sarcomas, solitary fibrous tumors, myofibromas, malignant mesotheliomas, thymomas, sarcomatiod carcinomas, malignant melanomas, and "phosphaturic mesenchymal tumors" [8]. Some authors used to classify the

dural-type HPC as subtype of meningeoma, until Ono at al. showed that both these entities are genetically different [9].

According to Nappi at al. true HPC may be defined immunohistochemically by reactivity for vimentin, with or without CD34 and CD57, but without showing neural or myoepithelial differentiation [8]. In 1995 Nielsen at al. described a unique variant of HPC – lipomatous HPC, positive immunohistochemically for vimentin and negative for actin, desmin, S-100, GFAP, EMA or keratin [10].

In our case, neoplastic cells were positive for both, vimentin and CD34 without immunoreactivity for S-100 and actin. Localization of the HPC in upper extremity is extremely rare [11]. Appearance of HPC in the same localization where twice-excised hemangiomas had appeaed still needs an explanation. There have been reports in world literature of HPC associated with Kasabach-Merritt syndrome [12] or glomus tumor [13]. In these cases vascular neoplasms existed simultaneously. In described case the authors cannot preclude that HPC was a part of both previously arisen vascular tumors, but was not included in incompletely excised biopsies.

HPC with diameter less than 6 cm and with low or absent mitotic activity are usually benign. The treatment of vasculary benign neoplasms is wide excision as is emphasized elsewhere, because the rate of recurrence is high. In presented case HPC showed low mitotic potential and was regarded as potentially benign. There was no recurrence of the tumor 6 years after wide margin surgical excision.

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