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Case Report

Intravascular papillary endothelial hyperplasia (masson tumor) of the right wrist: A case report and literature review

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ABSTRACT

Intravascular papillary endothelial hyperplasia (IPEH), also called masson tumor which is a benign vascular tumor named after Pierre Masson, the French pathologist who originally described it in 1923, terming it "hémangioendothéliome végétant intravasculaire." It is characterized by a reactive proliferation of endothelial cells associated with thrombosis. Ultrasound and MRI are the main imaging examinations, but the diagnosis of IPEH was confirmed by biopsy pathology and immunohistochemistry. It is generally accepted that surgical excision is the first choice of treatment. In this report, we report a case of Masson tumor arising in the right wrist.

Introduction

Masson tumor is a rare tumor-like lesion of soft tissue, accounting for 2% to 4% of skin and subcutaneous tissue tumors.[1] Its pathological features are papillary reactive proliferation of endothelial cells, named after French pathologist Pierre Masson, who first described the tumor in 1923. It is called "hemangio endotheliome vegetant intravasculaire". In 1976, Clearkin and Enzinger named this lesion

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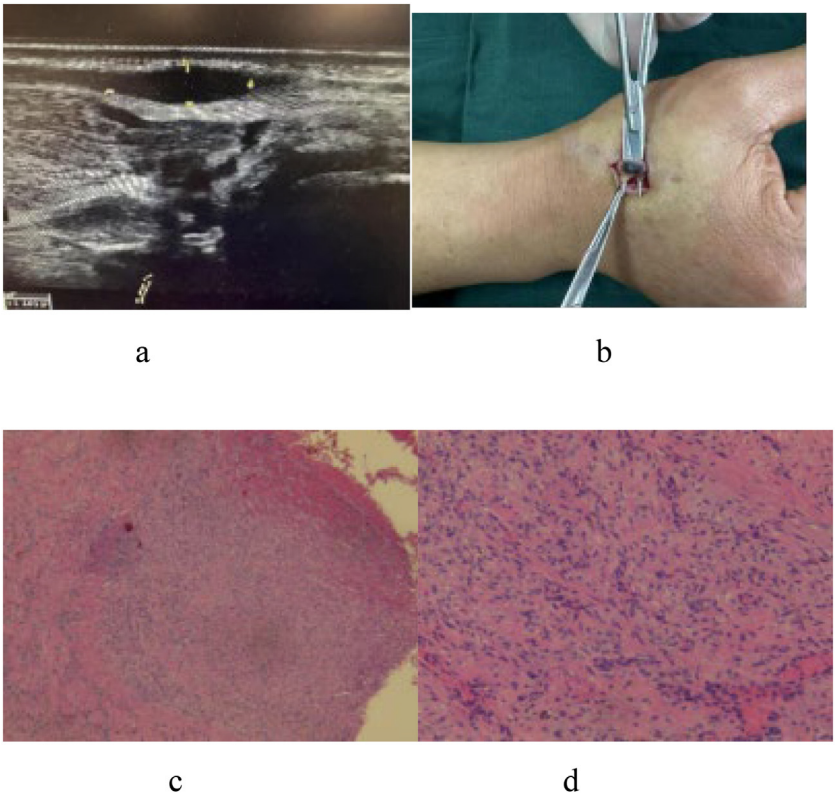

Figure 1. (a): The ultrasonographic characteristics were consistent with the hemangioma formed by thrombus mechanization. (b): The tumor was found to be located in the cephalic vein, and after both ends were ligation. (c.d): The postoperative pathological results showed that the vascular endothelial cells in the vascularlumen were characterized by papillary hyperplasia.

"intravascular papillary endothelial hyperplasia (IPEH)". It is a rare tumor found in the head and neck, limbs, and central nervous system, and has also been reported in the viscera and reproductive system.[2] It is generally considered to be a special form of thrombus organization rather than a primary vascular tumor.[3] It is more common in patients aged 30 to 40 years and slightly more female patients, so hormones are related to this lesion.[4] Levere et al.[5] proposed that IPEH after trauma may be mediated by the autocrine secretion of Basic fibroblast growth factor (bFGF), which is released by macrophages upon arrival at the wound site. Endothelial cell proliferation in turn releases more growth factors, thus activating a positive feedback regulatory loop of endothelial cell proliferation. However, because there are few reports of IPEH patients with a history of trauma, this theory cannot be used as a general explanation for the pathogenesis of IPEH.[6] In this paper, we report a case of masson tumor in the right wrist of a patient with a history of repeated trauma to the right wrist (Figure 1).

Case

A 47-year-old right-handed, construction worker male patient with a history of trauma to the right wrist presented to the hand surgery clinic with mild pain due to a dorsal mass on the right wrist for 3 months. Physical examination revealed that there was a tender fusiform mass in the tiger mouth area of the proximal carpometacarpal joint on the dorsal side of the right wrist. The size was 1.5 × 0.5 cm. The texture was hard and the skin was normal. Doppler ultrasound examination showed low echo in

the cephalic vein of the right hand tiger mouth area, with the size of 1.08 × 0.27 cm. MVI: little blood flow signal was seen inside. The ultrasonographic characteristics were consistent with the hemangioma formed by thrombus mechanization (a). After admission, surgery was performed under local swelling and anesthesia. During the operation, an "S" shaped incision was designed along the tumor area, skin and subcutaneous tissue were cut layer by layer to expose the tumor. The tumor was found to be located in the cephalic vein, and after both ends were ligation (b), the tumor was completely removed without any tumor infiltrating any adjacent structures, and no perioperative complications were observed. The postoperative pathological results showed that the vascular endothelial cells in the vascularlumen were characterized by papillary hyperplasia (c, d). The surface of the papillary structure was covered by a single layer of slightly hypertrophic endothelial cells, and the core structure under the endothelial cells was fibrous connective tissue. In the proliferating endothelial cells, there were no obvious atypia and mitotic signs, and no necrosis. And the patient has recovered well after surgery and no recurrence has been observed.

Discussion

Masson tumor is a rare soft tissue tumor-like lesion, accounting for 2−4% of skin and subcutaneous tissue tumors,[1] and in aggressive cases of IPEH are recurrence rates up to 10%. Its pathological feature is papillary reactive proliferation of endothelial cells. In addition, it is more common in patients aged 30–40 years, with slightly more female patients, so hormones are related to this lesion.[6] It tends to occur in the third and fourth decades of life.[2,7] The exact cause of Masson pine tumors is unknown.[8,9] Several factors may play a role, but the most likely cause is prior trauma, with IPEH forming after trauma and vascular disease present in 30% of cases. Radiation therapy has been proven to be a risk factor for inducing IPEH in the central nervous system.[10] Endothelial cell proliferation, in turn, releases more growth factors, thus activating the positive feedback regulatory ring of endothelial cell proliferation. However, since few IPEH patients have a history of trauma reported, this theory cannot be used as a general explanation for the pathogenesis of IPEH.[6]

IPEH can occur anywhere in the human body, mostly in the skin and subcutaneous tissues of the head, neck and upper limbs,[7,11,12] and its surface skin is mostly bluish purple and red, and it has also been found in the oral and tongue mucosa, heart, liver, small intestine and renal veins.[8] IPEH located in the brain and spinal cord has also been reported, but it is rare in ten.[9] In addition to the patient's medical history and physical examination, preoperative imaging examinations, such as ultrasound or MRI, must be performed. The diagnosis of IPEH is challenging to distinguish from venous malformations, ganglion cysts, ganglion giant cell tumors, schwannomas, and neurofibromas, especially angiosarcoma, because both IPEH and angiosarcoma exhibit enlarged endothelial cells, papillary structures, thrombosis, and irregular blood cell-filled Spaces. Therefore, the differential diagnosis of IPEH and angiosarcoma requires special attention.

Conclusions

IPEH is a kind of benign vascular tumor, which is very rare and often occurs in the soft tissues of limbs. The common shortcoming of relevant studies is the insufficiency of retrospective studies and the small number of cases. Preoperative diagnosis is not difficult through clinical features and imaging findings, but the final diagnosis is still pathology. A complete and correct diagnosis enables the clinical surgeon to solve the problem using the simplest surgical method. Simple surgical resection of IPEH in skin and soft tissue has a good effect, but there is a lack of clinical studies on IPEH that cannot be completely resected in the nervous system.

Declaration of competing interest

The authors declare there is no conflict of interests.

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Ethical approval

Not required.

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