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Volume 1 Issue 2
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ES Journal of Surgery

Angiolymphangioma at Femoral Neck: How to Diagnosis and Which Surgical Method is better for Children with Femoral Neck Pathological Fracture

Case Report

Xiangyu Deng, Wenbo Yang, Hang Liang, Zhicai Zhang, and Zengwu Shao

Department of Orthopaedic Surgery, Huazhong University of Science and Technology, China

Received: May 28, 2020; **Accepted:** June 17, 2020; **Published:** June 18, 2020

***Corresponding author:** Xiangyu Deng, Wenbo Yang, Hang Liang, Zhicai Zhang, and Zengwu Shao

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Abstract

Hemolymphangioma is a rare vascular developmental error. It comprises malformed venous and lymphatic component in various proportion. To the best of our knowledge, only a few cases have been reported in the literature so far. Here, we report a case of hemolymphangioma which happened in a 13-years-old patient's femur and also accompanied with a femoral neck fracture. He has undergone X-ray, CT, MRI and two biopsies without a clear diagnosis. Considering the high rate of avascular necrosis of the femoral head after children's femoral neck fracture, we performed femoral neck tumor resection, artificial bone grafting, internal fixation and iliac bone grafting for this patient and after the surgery, hemolymphangioma was the clear diagnosis. For hemolymphangioma happened in bone is such a rare diseases and the patient had a fracture in femoral neck, diagnosis and therapy were both difficult, we reported this case to help more doctor to recognize hemolymphangioma and choose the best surgery methods.

Keywords

Hemolymphangioma; Bone grafting, X-ray; CT; MRI

Introduction

Angiolymphangioma, also called as vascular tumor, is a rare bone occupying lesion involving a mixture of vascular and lymphatic malformations. Its main pathological manifestation is that the vascular tissue of tumor-like hyperplasia locates between the bone trabeculae and travels through the bone to cause the dissolution destruction of bone expansion, which can then cause a series of pathological changes, such as pathological fractures [1]. Although the disease is a benign vascular and lymphangioma hyperplasia lesions, but the hyperplastic lesions are often invasive growth, no significant capsular, showed cavernous, cut cavity filling light yellow lymph, cancellous bone trabecular scarce, assumes the dilatability of bone destruction, and lymphangioma bone blood vessels can be connected to the periosteum and the blood vessels in the surrounding soft tissue lymphangioma. The disease was first identified by immunohistochemistry in the dermis of Menzel and Kutzner [2]. It has been reported to be found in the spleen [3], lungs [4] and other organs [5], but the occurrence of the disease in the skeletal system is rarely reported.

Similar to other bone tumors, bone angiolymphangiomas can cause pathological fractures in patients, as well as pain, numbness, dysfunction and other symptoms caused by nerve compression because of the local occupying of tumor, so it is necessary to pay enough attention to this disease.

Medical treatment

The check of local hospital

A male patient of 13 years old came to our hospital for examination because of the pain in the right lower extremity for more than 2 months. After asking about medical history, we know that he fell on the ground and his right leg suffered severe pain two months ago, however, after the rest for a while, this pain eased. About a month later after that, the pain in the right lower extremity became more serious, so he came to hospital for imaging and biopsy. On January 24, 2019, he was treated at a local hospital. The local hospital performed magnetic resonance examination and showed his upper femur-femoral neck had a space-occupying lesion and the femoral neck suffered complete pathological fracture, the soft tissue around the lesion was swollen, and it was identified as a bone tumor-like lesion or tumor. What's more, eosinophilic granuloma has to be excluded, another left hip joint had a little fluid, both sides have several bilateral groin area lymph nodes, the MRI imaging were showed in Figure 1.

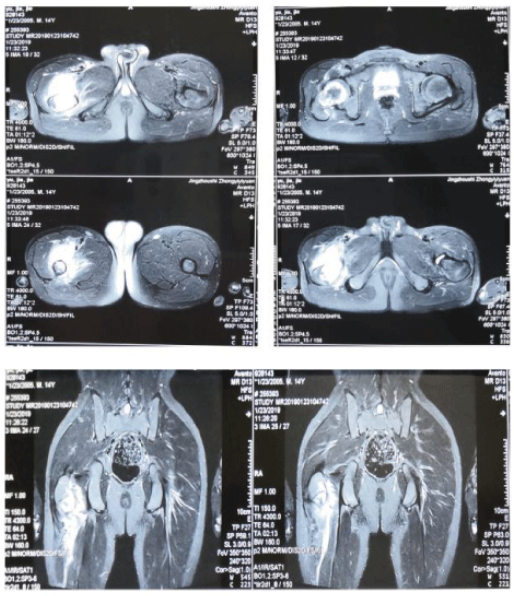

Figure 1: The MRI imaging of hip joint prompt there was a bone tumor-like lesion or tumor. What's more, eosinophilic granuloma have to be excluded.

CT-guided right thigh puncture picture examination in local hospital. On January 25, he was admitted to the local hospital. He underwent a CT-guided right thigh puncture picture examination. On January 28, he performed cytology puncture and both the results show that the tumor is possible; sarcoma cannot be ruled out (Figure 2).

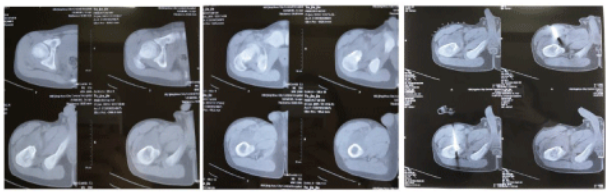

Figure 2: The imaging of CT showed tumor was suspected and the sarcoma could not be ruled out, and further examination is recommended.

The cytological puncture in our hospital. For it was difficult to diagnosis, he went to our hospital to went on deeper examination, on January 25, he performed cytology puncture again and the microscopic observation of a large number of red blood cells and some monocytes, villonodular synovitis cannot be rule out **(Figure 3).**

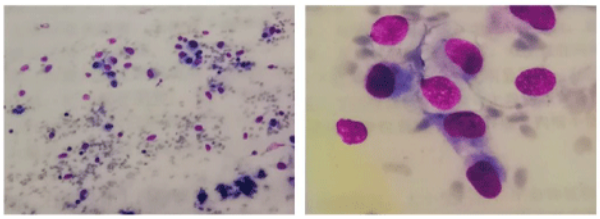

Figure 3: The microscopic observation of a large number of red blood cells and some monocytes, villonodular synovitis cannot be rule out.

Consultation on pathological examination of local hospital. At the same time, Wuhan Union hospital and Wuhan Tongji hospital conducted consultation on local pathology report and both thought immunohistochemistry is necessary, so local hospital went on immunohistochemistry and on February 3, the results suggested: (The right upper femoral puncture tissue) Microscopically see lymphocytes in skeletal muscle tissue, plasma cell infiltration, partial skeletal muscle tissue atrophy, partial papillary structures can be seen in some areas, central axis is red-stained collagen-like substance, surface covering the squamous epithelium cannot be ruled out as intravascular papillary vascular endothelium. Immunohistochemistry: CD31 and D2-40 were positive in the papillary structure, FLI-1 was positive, CD34, myogenin and myoD1 were negative,

striated muscle Desmin was positive, vascular smooth muscle SMA was positive, PCK was negative, and ki67 was low proliferative. In summary, lymphangioma may not be ruled out, and more specimens need to be diagnosed. The HE pictures were showed in Figure (4).

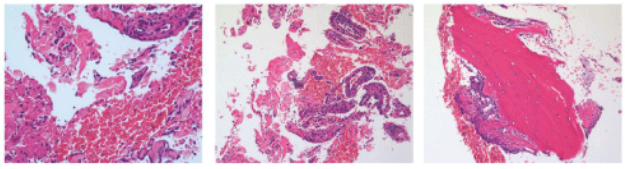

Figure 4: local hospital went on the biopsy and reported that lymphangioma may not be ruled out, and more specimens need to be diagnosed.

Cut biopsy in our hospital. For a clear diagnosis, we decided to cut the patient's lesion for biopsy. On February 24, the pathology report of our hospital: the examination (in the bone marrow lesions, the cancellous bone lesions in the femoral bone marrow) were all taken from the production. The microscopic mature lamellar bone was seen under the microscope, and more thick-walled blood vessels were seen in the bone marrow cavity. More lymphocytes and plasma cells infiltrated around the blood vessels. Immunohistochemistry suggested: S100, langerin, CD1a (-), CD68 (a small amount of tissue cells +). Eliminate Langerhans cell histiocytosis (also known as eosinophilic granuloma). On February 28, Tongji Hospital consulted the pathology of our hospital: (intramedullary lesions, cancellous bone lesions in the femoral bone marrow). The bone fragments were seen under the microscope. The bone marrow tissue and the lymphocytes and plasma cells infiltrated in the marrow were not seen. The HE pictures were showed in Figure (5).

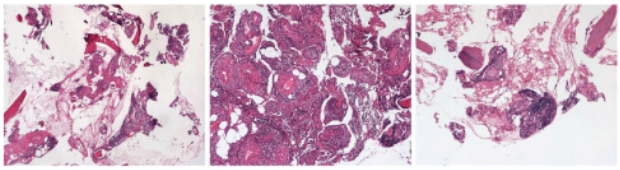

Figure 5: The bone marrow tissue and the lymphocytes and plasma cells infiltrated in the marrow were not seen

The MRI enhanced examination in our hospital. At the same time, the patient went on second MRI in his lesion, and On February 13, plain and enhanced MRI of the hip joint in our hospital showed patchy iso-T1 long T2 signal shadow in the upper part of the right femoral neck-femur, erosion of the bone shaft, and uneven enhancement in lacy shape. The lesion did not involve the epiphysis of the femoral head. The upper and lower range of the lesion was about 14 cm, adjacent to the medial femur and iliac muscle, rectus femoris, iliopsoas muscle, pubis muscle and interfemoral muscle. No obvious enhancement was found. The nature of these changes remains to be determined, taking into account the right femoral neck-upper femoral lesions and surrounding soft tissue changes. In addition, a small amount of hydrocephalus of the hip joint, narrowing of the gap between the ischial tubercle and the femoral trochanter, and high signal intensity of quadratus femoris muscle are suspected to be diagnosed as femoral-ischial impingement syndrome. The MRI enhance imaging were showed in Figure 6.

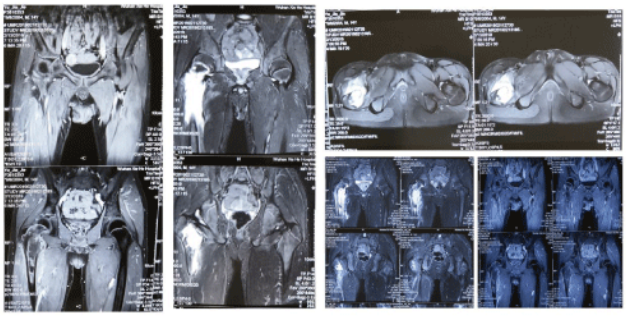

Figure 6: The nature of these changes remains to be determined, taking into account the right femoral neck-upper femoral lesions and surrounding soft tissue changes

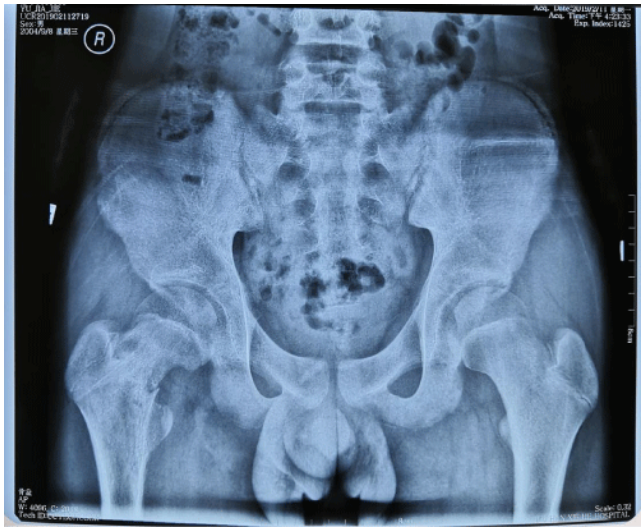

Figure 7: The upper part of right femur occupies space, considering bone tumors or bone tumour-like lesions, accompanied by pathological fracture.

X-ray examination before surgery. What's more, the X-ray was carried out for the preparation of surgery. The X-ray reported: in the upper part of right femur, there was osteolytic bone destruction with a range of 8.7cm × 5.0 cm (upper and lower diameters × left and right diameters). Accumulated femoral neck and intertrochanteric bone was dense, local cortex thinned and discontinuous, femoral neck was obvious and surrounding soft tissue swelling. The upper part of right femur occupies space, considering bone tumors or bone tumour-like lesions, accompanied by pathological fracture. The X-ray imaging was showed in Figure 7.

Surgery

For we have finished all the examination, although the pathology is still not clearly diagnosed, we orthopaedics, pathology, imaging all agree that this patients is more like a benign tumor, we decided to perform surgery on the treatment of benign tumors. The methods of this surgery included tumor scraping, artificial bone implantation, internal fixation and the most important, iliac bone flap transplantation. This patient was just 13 years old and his fracture is in a special position, the femoral neck, which is more difficult to heal, the iliac bone flap transplantation will help the process of fracture healing, also help to reduce the possibility of femoral head necrosis. We found this tumor is more like a fluid mixed with lymphatic and vascular tissue. Picture of tumor exposure is showed in Figure 8.A, and the picture of musculoskeletal transplantation is showed in Figure 8.B. The operation is smooth, and the patient recovers well after anesthesia wakes up.

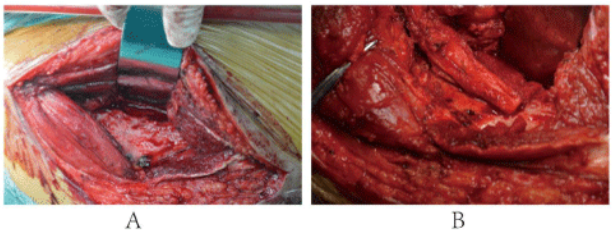

Figure 8: The tumor exposure (A) and musculoskeletal transplantation (B).

The finial diagnosis

After the surgery, this tumor specimen was sent to pathology for the final diagnosis. 7 days after the surgery, the pathology in our hospital reported about this tumor: (Right femur) vascular tumor (hemangioma with lymphangioma) with pathological fractures and traumatic microscopic structure, immunohistochemical staining showed hemangioma components: CD34(+), ERG(+), S1000(-), Langerin(-); Lymphangioma composition: D2-40(+); Fibroblasts: SMA(+). The HE stain is showed in Figure 9. A and the immunohistochemistry is showed in Figure 9. B. After the final diagnosis, we discussed the reasons why the first two biopsies did not lead to pathological findings. We believe that biopsies cannot reach the tumor wall, which is the most important structure in the diagnosis process, only by the lymph or bloody fluid cannot get the right diagnosis. 3 days after surgery, X-ray was carried out to ensure the internal fixed position, which were in a right position and the artificial bone were in a good condition (Figure 9. C).

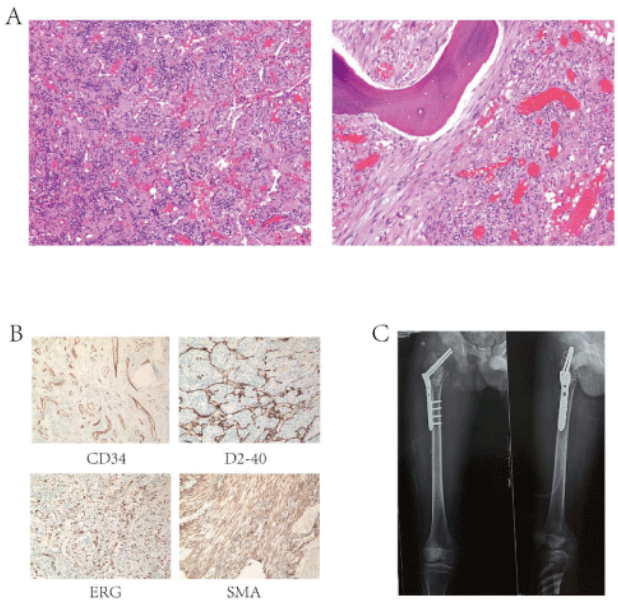

Figure 9: The HE stain (A), immunohistochemistry (B) and the postoperative X-ray imaging (C).

Discussion

Angiolymphangioma is a benign tumor of mesenchymal origin, which contains both the pathological components of hemangioma and the components of lymphangioma. The disease often occurs in infants and children which is often located in the body surface and other parts. The clinical incidence of angiolymphangioma is low about 0.12% ~ 0.28% [6]. Angiolymphangiomas of the bone are much less common. Bone angiolymphangiomas have been reported mainly in the spine [7]. It is an accepted fact that there

are abundant blood vessels and sinuses in bones. And the fact that there are lymphatic vessels in the bone has also been confirmed by Nixon with lymphangiography, which is the pathologic source of angiangioma of bone [8]. The pathological manifestations of angiolymphangioma are cystic or cavernous lesions, composed of dilated veins and lymphatic vessels, interspersed with normal matrix tissue and vascular system, and dilated vessels may contain thrombosis [9]. There have no previous research about the hemolymphangiomas which happened in long bones, maybe for the reason of less lymphatic and blood drainage. As for this case, hemolymphangiomas happened in a more specific position, femoral neck and femoral shaft, a special anatomy structure. Especially for children, femoral neck fractures in children are very rare and account for about 1% of all paediatric fractures. The aim of this retrospective study was to analyse the clinical and radiographic outcome in paediatric femoral neck fracture and to review the role of early decompression of the hip in the final outcome. Femoral neck fracture in children is one of the refractory fractures [10-12]. Due to the poor blood supply of the femoral head and the presence of the epiphyseal plate, the incidence of complications after femoral neck fracture is high, and the treatment of complications is difficult, and the long-term efficacy is poor [13,14]. For this case, the patient suffered not only the fracture, more specific, it's the pathological fracture, which means it's more difficult for him to heal. A systematic review and meta-analysis [15] analysis the quadratus femoris muscle pedicle bone flap transplantation in the treatment of femoral neck fracture for Chinese young and middle-aged patients and found that for the treatment of femoral neck fractures, transplantation of quadratus femoris muscle pedicle bone flap combined with hollow compression screw; fixation is superior to the AO hollow compression screw fixation in terms of the rate; of fracture healing, the rate of good function of hip joint, the rate of femoral head; necrosis and the time of fracture healing. So we decided to use a myocutaneous flap to reduce the possibility of femoral head necrosis.

On immunohistochemistry analysis, hemolymphangiomas are CD31 [16-18] and D2-40 [19-21] positive [22], which represent blood vessels and lymphatic vessels origin. From the point of view of disease occurrence, hemangiolymphangioma is divided into congenital and acquired. Congenital hemolymphangiomas mainly originate from an obstruction of the venolymphatic communication between the systemic circulation and the dysembryoplastic vascular tissue [23]. The occurrence of acquired hemangiolymphangioma is due to insufficient lymphatic drainage and lymphatic injury caused by surgery or trauma [7]. In this case, the patient was acquired angiolymphangioma, which may be related to the above factors. What's more, we analysis the reason why the first and second biopsy cannot reach the final diagnosis. For hemangiolymphangioma is more like a liquid tumor, puncture not really reach the specific lesion. Only after the surgery can get the wall of tumor, which is the most important section to get the right pathological diagnosis.

Accurate diagnosis of angiolymphangioma of bone is very important. At present, imaging diagnosis and pathological biopsy are the main methods for the diagnosis of bone angiolymphangioma. Imaging diagnosis is one of the most important preoperative diagnoses of this disease. Ultrasound, computed tomography (CT) and MRI are all useful in the diagnosis of bone angiolymphangioma, and the accuracy of their results depends on the number and water content of blood vessels in the lesion [24]. For this case, MRI showed a huge abnormal signal range, however, this abnormal section was not all the tumor, some edema of the surrounding tissues and bone marrow edema also contribute to the abnormal performance. This patient had done MRI exanimation for two times and through the comparison of two MRI imaging, we found the abnormal signal range were smaller, that's why we decided to treat this tumor as a benign tumor treatment. The final diagnosis confirmed we had done a right decision.

The diagnosis of angiolymphangioma in bone and the compared pathological fracture both need more and deeper research; we hope this case report can give some evidence of this disease.

Source of Funding

This work was supported by the National Natural Science Foundation of China (No. 81472134) and the grants 2016YFC1100100 from The National Key Research and Development Program of China.

References

1. Donnelly LF, Adams DM, Bisset GS. Vascular malformations and hemangiomas: a practical approach in a multidisciplinary clinic. AJR Am J Roentgenol. 2000; 174: 597-608.
2. Mentzel T, Kutzner H. Tumors of the lymphatic vessel of the skin and soft tissue. Pathologe. 2002; 23: 118-127.
3. Merritt AJ, Wilkins BS, Williams MS, Hay C, Byers RJ. Synchronous splenic and bone marrow haemangiolymphangioma: a novel entity. J Clin Pathol. 2014; 67: 645-647.

4. Copeland J, Müller KM, Müller AM. Pulmonary haemangiolymphangioma--a new entity of pulmonary vascular tumours. Histopathology. 2008; 52: 527-529.
5. Sapountzis S, Singhal D, Chen HC. Radical resection and reconstruction with bilateral gluteal fold perforator flaps for vulvar hemangiolymphangioma. Int J Gynaecol Obstet. 2013; 121: 179-180.
6. □□□, □□, □□□. □□□□□□□□□□□1□. □□□□□□□□□. 2017; 33: 1245.
7. Pan X, Dong Y, Yuan T, Yan Y, Tong D. Two cases of hemolymphangioma in the thoracic spinal canal and spinal epidural space on MRI: The first report in the literature. Medicine (Baltimore). 2017; 96: e9524.
8. Nixon GW. Lymphangiomatosis of bone demonstrated by lymphangiography. Am J Roentgenol Radium Ther Nucl Med. 1970; 110: 582-586.
9. Pandey S, Fan M, Chang D, Zhu J, Zhu Y, Li Z. Hemolymphangioma of Greater Omentum: A Rare Case Report. Medicine (Baltimore). 2016; 95: e3508.
10. Caldwell L, Chan CM, Sanders JO, Gorczyca JT. Detection of Femoral Neck Fractures in Pediatric Patients with Femoral Shaft Fractures. J Pediatr Orthop. 2017; 37: e164-e167.
11. Spence D, DiMauro JP, Miller PE, Glotzbecker MP, Hedequist DJ, Shore BJ. Osteonecrosis After Femoral Neck Fractures in Children and Adolescents: Analysis of Risk Factors. J Pediatr Orthop. 2016; 36: 111-116.
12. Fitschen-Oestern S, Lippross S, Klüter T, Behrend P, Weuster M, Seekamp A. Femoral neck fractures in young patients. Unfallchirurg. 2016; 119: 763-780.
13. Bukva B, Abramović D, Vrgoč G, Marinović M, Bakota B, Dučić S, et al. Femoral neck fractures in children and the role of early hip decompression in final outcome. Injury. 2015; 46: S44-47.
14. Al-Ani AN, Neander G, Samuelsson B, Blomfeldt R, Ekström W, Hedström M. Risk factors for osteoporosis are common in young and middle-aged patients with femoral neck fractures regardless of trauma mechanism. Acta Orthop. 2013; 84: 54-59.
15. Wang XJ, Zhang ZH, Li L, Su YX, Wei L. Quadratus femoris muscle pedicle bone flap transplantation in the treatment of femoral neck fracture for Chinese young and middle-aged patients: A systematic review and meta-analysis. Chin J Traumatol. 2017; 20: 347-351.
16. Gallay N, Anani L, Lopez A, Colombat P, Binet C, Domenech J, et al. The role of platelet/endothelial cell adhesion molecule 1 (CD31) and CD38 antigens in marrow microenvironmental retention of acute myelogenous leukemia cells. Cancer Res. 2007; 67: 8624-8632.
17. Yildirim A, Akkus M, Nergiz Y, Yuruker S. Immunohistochemical analysis of CD31, CD36, and CD44 antigens in human omentum. Saudi Med J. 2004; 25: 308-312.
18. Balduini CL, Noris P, Giorgiani G, Martinetti M, Klersy C, Spedini P, et al. Incompatibility for CD31 and human platelet antigens and acute graft-versus-host disease after bone marrow transplantation. Br J Haematol. 1999; 106: 723-729.
19. de Vilhena AF, das NPJC, Parra ER, Balancin ML, Saber A, Martins V, et al. Histomorphometric evaluation of the Ki-67 proliferation rate and CD34 microvascular and D2-40 lymphovascular densities drives the pulmonary typical carcinoid outcome. Hum Pathol. 2018; 81: 201-210.
20. Al-Rohil RN, Milton DR, Nagarajan P, Curry JL, Feldmeyer L, Torres-Cabala CA, et al. Intratumoral and peritumoral lymphovascular invasion detected by D2-40 immunohistochemistry correlates with metastasis in primary cutaneous Merkel cell carcinoma. Hum Pathol. 2018; 77: 98-107.
21. Sadullahoğlu C, Dere Y, Atasever TR, Öztop MT, Karaaslan Ö. The Role of CD34 and D2-40 in the Differentiation of Dermatofibroma and Dermatofibrosarcoma Protuberans. Turk Patoloji Derg. 2017; 1: 223-227.
22. Handra-Luca A, Montgomery E. Vascular malformations and hemangiolymphangiomas of the gastrointestinal tract: morphological features and clinical impact. Int J Clin Exp Pathol. 2011; 4: 430-443.
23. Li Y, Pang X, Yang H, Gao C, Peng B. Hemolymphangioma of the waist: A case report and review of the literature. Oncol Lett. 2015; 9: 2629-2632.
24. Chanfi M. Hemolymphangioma of the orbit in a young girl: a clinical observation. J Fr Ophtalmol. 2004; 27: 1047-1049.