Hemolymphangioma of the waist: A case report and review of the literature

YONGCHAO LI[1,2*], XIAODONG PANG[2*], HONG YANG[2], CHUNHUA GAO[2] and BAOGAN PENG[2]

[1]Postgraduate Training Base, General Hospital of Armed Police Force, Liaoning Medical University, Jinzhou, Liaoning 121000; [2]Department of Spinal Surgery, Institute of Spinal Surgery of Armed Police Force, General Hospital of Armed Police Force, Beijing 100039, P.R. China

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Abstract. Hemolymphangioma is a malformation of the lymphatic and blood vessels. To the best of our knowledge, only a limited number of hemolymphangioma cases have been reported in the literature thus far, with no cases developed in the waist region. The present study reported the case of a 17-year-old male patient with hemolymphangioma growing on the waist, presented with back pain for four months. Upon physical examination, the lesion was identified to be oval in shape, soft and compressible, with mild tenderness. No abnormalities were detected in the results of laboratory examinations. However, a magnetic resonance imaging (MRI) scan revealed a tumor with low signal intensity on T1-weighted imaging (WI) and high signal intensity on T2-WI. The mass was successfully removed during surgery. During the seven-month follow-up period, the patient was asymptomatic with no evidence of recurrence. The present study discussed the imaging findings and pathological features of this uncommon case and reviewed the relevant literature.

Introduction

Lymphangioma, also known as angioma lymphaticum, is a congenital malformation of the vascular system, comprising newly-formed lymph spaces and channels (1). Landing and Farber (2) classified this benign malformation in four categories, including capillary, cavernous and cystic (hygroma) lymphangioma, and hemolymphangioma, which is a combination of hemangioma and lymphangioma.

Correspondence to: Professor Baogan Peng, Department of Spinal Surgery, Institute of Spinal Surgery of Armed Police Force, General Hospital of Armed Police Force, 69 Yongding Road, Beijing 100039, P.R. China
E-mail: pengbaogan76611@163.com

*Contributed equally

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Hemolymphangioma is a congenital malformation that may be asymptomatic for a long period of time (3). This lesion is typically considered to be a benign and noninvasive disorder, characterized by the presence of dilated lymphatic spaces, extravasation of red blood cells, hemosiderin deposition and fibrosis (4). Hemolymphangioma formation may be due to venolymphatic communication obstruction between the dysembrioplastic vascular tissue and systemic circulation (5).

The incidence of hemolymphangioma is 1.2-2.8 per 1,000 newborn infants (6). Hemolymphangioma has been previously detected at the pancreas (5,7-12), spleen (13-16), stomach (1,17), rectum (18), mediastinum (19-21), chest wall (22-25), small intestine (26), extremities (3,27,28), cervix (29,30), pericardium (31), oral region (32), esophagus (33), axilla (34), retroperitoneal space (35,36), adrenal gland (37), abdomen (38), duodenum (4) and hepatica (39), as well as on the tongue (40,41) and orbit (42,43). However, to the best of our knowledge, no studies have reported this type of tumor in the waist region, as determined by a review of the medical literature until June 2014 using the PubMed database (http://www.ncbi.nlm.nih.gov/pubmed; accessed on 9th June 2014). Complete excision is considered the optimal treatment for hemolymphangioma, which exhibits a low recurrence rate. Non-surgical treatments are also used, including aspiration and drainage, cryotherapy, injection of sclerotic agents, laser therapy and radiotherapy, however, to date, the outcomes of such treatments have been unsatisfactory (3,22). In cases of tumor recurrence, conservative treatment methods such as laser therapy, may be applied (3,44). Generally, the prognosis of hemolymphangioma is good (3,4,13,17,18), however, careful follow-up is required.

The present study reported the case of a 17-year-old male patient with hemolymphangioma on the waist and reviewed the characteristics of this disease based on the existing literature.

Case report

A 17-year-old male was admitted at the General Hospital of Armed Police Force (Beijing, China) in September 2013, complaining of a mass on the right side of the waist and back pain for approximately four months. The back pain was significantly increased when the patient was sedentary and was slightly alleviated by rest. On admission, the patient was

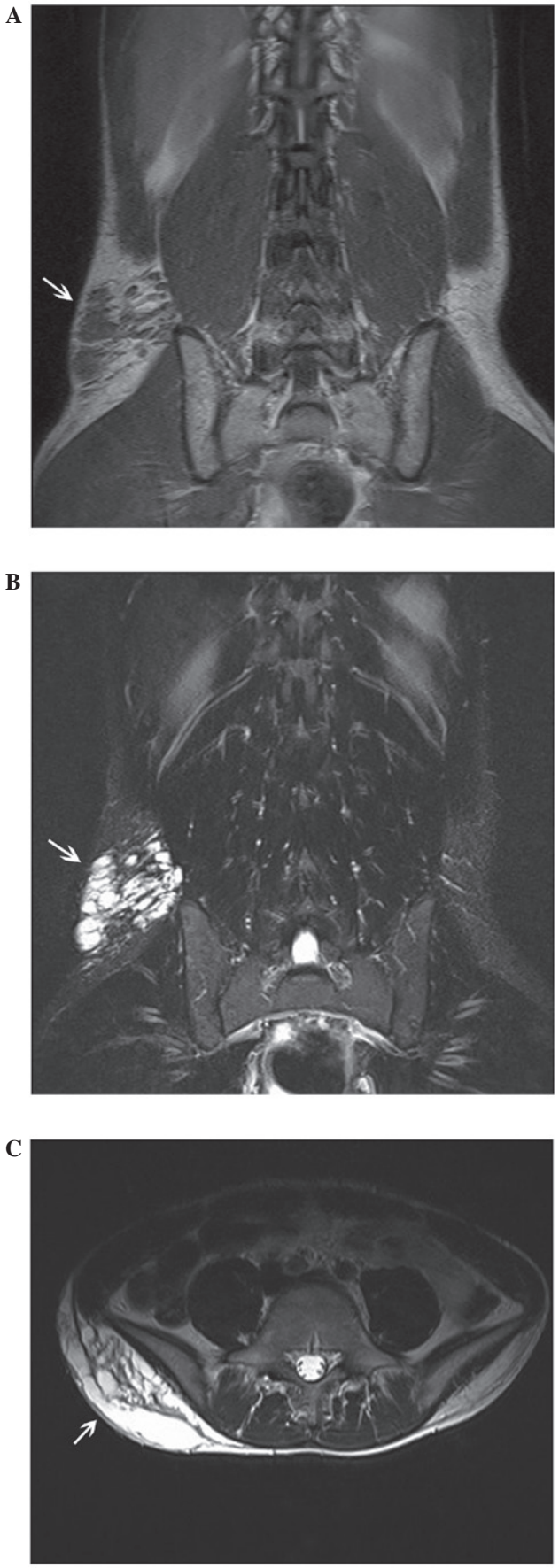

Figure 1. Magnetic resonance imaging (MRI) findings. The scans demonstrate the (A) coronal T1-weighted imaging (WI), (B) coronal T2-WI and (C) axial T2-WI. A mass (arrows) was identified, with low signal intensity on T1-WI and high signal intensity on T2-WI in the right side of the waist subcutaneous tissue with the fifth lumbar level parallel.

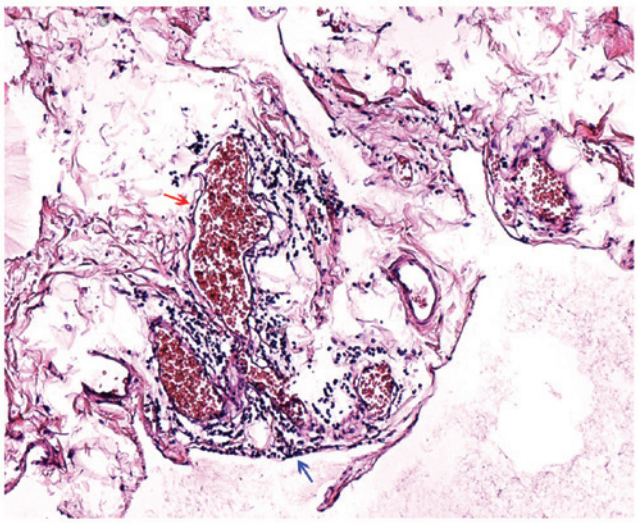

Figure 2. Histological analysis of biopsy specimen (hematoxylin and eosin stain; magnification, x100). Blood vessels (red arrow) and lymphatic vessels (blue arrow) can be observed in the tumor.

well, with no symptoms of lower extremity numbness and pain, or abdominal pain. Upon physical examination, the lesion was identified to be oval in shape, soft and compressible, with mild tenderness. No abnormalities were detected in the results of laboratory examinations. An ultrasound (Doppler sonography) detected a cystic lesion (15.0x10.0 cm) with blood flow, and revealed multiple echo and irregular cavities in the subcutaneous fat layer of the right waist. The most likely diagnosis was hemolymphangioma.

Magnetic resonance imaging (MRI) examination was performed preoperatively, in order to establish the extent of the tumor and define its association with the surrounding tissues. A waist MRI scan (Fig. 1) revealed a 12.6x9.7-cm mass, with low signal intensity on T1-weighted imaging (WI) and high signal intensity on T2-WI in the right side of the waist subcutaneous tissue with the fifth lumbar level parallel. Upon performing a tumor biopsy with a 5-ml syringe, 10-ml yellow, clear liquid was extracted and laboratory examinations were performed. Preoperatively, no abnormalities were revealed in the laboratory data, including the levels of tumor markers [α-fetoprotein, carcinoembryonic antigen, carbohydrate antigen (CA) 19-9, and CA-125] and concentrated mycobacterium tuberculosis.

Due to the patient experiencing back pain that was increased upon sitting, surgical excision was decided as the treatment strategy. During surgery, the boundary of the mass was unclear and a bloody yellow exudate was observed. Macroscopically, the mass measured approximately 12.0x6.0x6.0 cm, and was oval and soft. Multiloculated cystic masses filled with blood and yellow fluid were extracted. Histologically, the tumor was composed of lymphatic and blood vessels with polycystic spaces (Fig. 2). Considering these observations, the definitive histological diagnosis was hemolymphangioma of the waist. The postoperative course of the patient was uneventful. In the course of a seven-month follow-up period, no recurrence of hemolymphangioma was observed. This study was approved by the ethics committee of General Hospital of Armed Police Force (Beijing, China)

and written informed consent was obtained from the patient's family.

Discussion

Hemolymphangiomas, a congenital malformation of the vascular system, can be classified into primary and secondary lymphatic vascular tumors. Primary tumors are congenital malformations of the lymphatic vascular system, possibly formed due to obstruction of the venolymphatic communication between the dysembryoplastic vascular tissue and the systemic circulation. By contrast, secondary tumors are likely to be caused by poor lymph drainage and lymphatic damage resulting from surgery or trauma (43). Hemolymphangioma mainly presents as cystic or cavernous lesions. Histologically, hemolymphangioma is composed of dense fibrous tissue that develops in bands between the numerous vascular spaces, invading the subcutaneous fat and involving the blood or lymphatic vessels (3).

The incidence of hemolymphangioma is 1.2-2.8 per 1,000 newborns (45), and the two genders are equally affected. In the present study, a review of the literature up to June 2014 was performed using the PubMed database. The search strategy to identify all possible studies involved use of the word 'hemolymphangioma'. In total, 47 previous studies concerning this type of tumor were identified (1,3-42,44,46-50). However, to the best of our knowledge, no studies have reported hemolymphangioma of the waist. In the current case, the patient was a 17-year-old male, and the tumor occurred on the waist and appeared as a cystic lesion.

Clinically, the onset of hemolymphangioma can vary between a slow-growing cyst over a period of years and an aggressive enlarging tumor without invasive ability (3). The size of these tumors varies due to the different anatomical location and association with the neighboring tissues. In clinical examinations, they are usually described as soft and compressible masses, loculated in pattern. The most common complications are random or traumatic hemorrhage, rupture and infection (3). However, no abnormal laboratory findings were observed in the current patient, and the only symptom was back pain for four months.

In the present study, a waist MRI scan revealed a tumor with low signal intensity on T1-WI and high signal intensity on T2-WI on the right side of the waist subcutaneous tissue in the fifth lumbar level. These observations may indicate the presence of a lower number of tortuous blood vessels and water-based substance in the lesion, which was then confirmed during surgery. Imaging examinations, including ultrasound, computed tomography and MRI scans, are useful in order to confirm the diagnosis, identify the tumor nature, and observe its extension and association with the surrounding tissues, assisting the selection of the surgical strategy and follow-up treatment (51). However, a definitive diagnosis of the tumor in the present study was based on histological evidence.

Surgical resection appears to be the most effective treatment for hemolymphangioma. In order to prevent recurrence, thorough radical resection may be required during surgery. In the English literature, the reported recurrence rates were in the range of 10-27% upon complete removal of the lesions, while the recurrence rates were 50-100% in cases where the lesions were partially removed (4,52). The extend of surgical resection depends mainly on the anatomical location and complexity of the tumor (3,4,12,52).

In conclusion, hemolymphangioma of the waist is an uncommon vascular and lymphatic lesion, presenting mainly with back pain. Preoperative imaging examinations, including ultrasound and MRI, are important for a full evaluation of the tumor in order to confirm the diagnosis and plan the surgical strategy. Complete surgical resection is the most effective treatment with good prognosis.

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