CASE REPORT

LYMPHANGIOMA OF AN UNUSUAL SITE:
A CASE REPORT

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Abstract

Lymphangiomas are hamartomatous congenital malformations of the lymphatic system that usually involve subcutaneous tissues of cervico-facial region. Rarely, it can be found in subcutaneous tissue of proximal extremities, the buttocks and the trunk. Magnetic Resonance Imaging (MRI) is the best modality to assess the tumor specification and extension. We report a case of lymphangioma at a rare site with its radiological features and patient responded to the sclerosant therapy.

Keywords: Lymphangioma, Benign Vascular Abnormality, OK-432, Children

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Introduction

Lymphangiomas are hamartomatous congenital malformations of the lymphatic system that usually involve subcutaneous soft tissues. They are rare and accounting for 4% of all vascular tumors and approximately 5.6% of all benign vascular tumors in children [1]. Near 80% of lymphangiomas developed at cervico-facial region followed by the proximal extremities, the buttocks and the trunk. Less commonly, they can be seen in the intestines, the pancreas and the mesentery. We report a case of lymphangioma occurring at a rare site which is at the buttock, with literature review on lymphangioma.

Case Report

A one-year-old girl presented with history of progressive left buttock swelling for 3 months duration. There was no tenderness or skin discoloration. On clinical examination, there was an isolated cystic swelling at the left gluteal region. It was soft in nature, did not attach to underlying bone and without any signs of inflammation. On ultrasound examination, there were elongated anechoic tubular structures in the subcutaneous tissue and they were fully compressible. No color Doppler flow was detected within the lesions (Figure 1). The findings of tubular structures without Doppler signals favor for lymphatic vessels.
Figure 1. Ultrasound of subcutaneous area at left buttock region. There were tubular structures without color Doppler signals (arrow)

Magnetic Resonance Imaging (MRI) was done at the age of 16 months and showed a lobulated, multicystic lesion located superficially within the subcutaneous fat of left gluteal region. It extended inferiorly to the proximal upper third of the thigh and measures 5.5cm x 3.5cm (width x craniocaudal) at the largest area (Figure 4(a)). The lesion was hypointense to muscle on T1-weighted image (T1WI), heterogeneously hyperintense on T2-weighted image (T2WI) and peripherally enhanced after gadolinium administration (Figure 2).
Figure 2. The first MRI performed before treatment. Axial image on TI (a), T2 (b) and post-contrast (c). Multiple cystic structures with septal contrast enhancement.
The diagnosis of lymphangioma was made and the patient was advised for aspiration and sclerotherapy. Patient underwent sclerotherapy at the age of 2 years and 5 months old. On aspiration, chylous fluid was obtained, confirming the diagnosis.

Sclerotherapy (Figure 3) was given with Picibanil (OK-432) for three times at 8 weeks and 17 weeks intervals from the last treatment. MRI was repeated at the age of 3-year-old, that is 1 year after the last therapy which showed reduction in size (Fig.4 (b)).

Figure 3. Left buttock lymphangioma sclerotherapy procedure with OK-432
Figure 4. MRI images before and after therapy. (a) Coronal T2 images before therapy. (b, c) Coronal and axial T2 image of the same lesion after therapy showing reduced in overall size.
Discussion

Lymphangiomas are developmental malformations of the lymphatic system, which is the network of vessels responsible for transporting fluid away from tissues before returning it to the blood. Lymphangiomas are benign multilobular cystic masses lined by endothelial cells. They occur as a result of maldevelopment and obstruction of the lymphatic system. Eventually, there is no communication between sequestrated lymphatic tissue and normal lymphatic system [2].

Lymphangioma can occur anywhere in the developing lymphatic system but mostly, about 75% of the cases are located in posterior cervical triangle [1]. About 20% of cases is located at the axilla and uncommonly, cases of lymphangioma has been seen in the chest wall, retroperitoneum, abdominal organs, groin and bone. The lesions size is variable and margins can be either well-defined to ill-defined. They tend to diffuse with unclear borders. The majority of lymphangioma are presented at birth and the rest usually manifest before the age of 2 years. They can appear suddenly in childhood and occasionally in adolescence or adulthood [3].

The overlying skin is often normal or a bluish hue. Lymphangioma in extremities may associate with diffuse or localized swelling or gigantism due to soft tissue and skeletal overgrowth. There was a rare type of spongiform lymphangioma in the lower extremity with a large proximal cystic lymphatic reservoir in the groin [4]. Our patient did not have skin discoloration, bone asymmetry or bone over growth.

Lymphangiomas are classified into three subtypes based on microscopic features, which are capillary and cavernous lymphangiomas and cystic hygromas [5]. Our case was a cavernous lymphangioma, which composed of large dilated lymphatic channels. This type of lymphangiomas characteristically invades surrounding tissues.

Ultrasound is the usual preliminary study, which will reveal subcutaneous soft tissue mass with tubular structures. The absence of color Doppler signals implies that the lesion is arising from lymphatic vessels and is not pulsatile like arterial system. This ultrasound feature is useful to differentiate lymphangioma from vascular hemangioma.

MRI is the best imaging modality for lymphangioma. The lesion will have variable signal intensity on T1WI and showed hyperintense on T2WI due to high water content. They can have multiple cystic lesions. Large cysts may have fluid-fluid levels due to protein or blood components. However, fluid-fluid level was not seen in our case suggestive of absences of the components. Contrast administration usually gives rim enhancement around large cysts, within septa and intervening soft tissue as seen in our case.

Teratoma and infantile fibrosarcoma are two differential diagnoses that can appear as cystic on radiologic imaging and be confused with cystic lymphangioma.

Other diagnostic imaging that can be performed are conventional and Magnetic Resonance Lymphangiography (MRL). However, both are rarely performed nowadays except for conventional lymphangiography, which still has a roll to determine the precise location of lymphatic or chylous leakage in a patient with a thoracic lymphatic anomaly [4].

Although 10% of lymphangioma will
resolved spontaneously [6], most of lymphangioma cases need treatment due to painful swelling, recurrent cellulitis or for cosmetic reasons [7]. There are two main treatments for lymphatic anomalies, which are interventional treatment of sclerotherapy and surgical resection. Due to the nature of lymphangioma growth, which is infiltrative in nature, only two thirds of lymphatic malformations can be completely excised. Neural and vascular structures must be carefully dissected to achieve a good outcome [4, 8]. The recurrence rate is 40% after incomplete excision and 17% after macroscopically complete excision. Regrowth and reexpansion from microcystic channels are responsible for postoperative recurrence [4].

For sclerosing therapy, cystic lesion should be large enough to permit injection of sclerosant. Pure ethanol, sodium tetradecyl sulfate and doxycycline were used as sclerosing agents previously, which have been replaced with newer sclerosant agent such as bleomycin and OK-432. We used OK-432 as sclerosant agent for our case. OK-432 is produced by incubation and interaction of Streptococcus pyogenes with penicillin G potassium [9]. Three cycles of injection of OK-432 into the largest cysts were done in our patient (Figure 2). The interval of 1\textsuperscript{st} and 2\textsuperscript{nd} treatment was about 9 weeks and for 2\textsuperscript{nd} and 3\textsuperscript{rd} treatment was about 28 weeks. Significant reduction in the size of the lesion has been noted after 3 sclerosant therapy session which was about 57.8% in reduction (from 5.55 cm x 3.53 cm to 3.21 cm x 2.57 cm). There are number of case reports and series that show total lesion shrinkage after OK-432 administration and some of the authors have proposed OK-432 as the first line treatment for lymphangiomas [10]. Currently, our patient is planned for another OK-432 sclerosant therapy session.

There are other types of treatment such as cryotherapy or diathermy. However it shows marginal success and may exacerbate infection. Aspirations of fluid from a larger cyst only provide temporary decompression and rarely yield a positive culture. It is no longer recommended except for urgent decompression at a specific site [8]. Radiation treatment has no benefit in the treatment of lymphatic malformations and it will cause significant morbidity in the growing child [8].

Depends on the site of the lesion, patients can present with complication such as respiratory obstruction, feeding and speech difficulties, skeletal overgrowth and maxillary malocclusion [11]. In cervical lymphangioma, the incidence of infection can occur up to 71% of the cases and most of the cases would need parenteral antibiotic administration. Ulceration can also occur due to pressure necrosis, which would be managed by dressing and course of antibiotic [11].

Conclusion

Lymphangiomas are benign vascular lesions that are commonly occur at the head and neck region, however it may occur anywhere in the body. Radiological procedure can assist in the diagnosis as well as management of the lesions.

References


