Cavernous Lymphangioma of the Thigh in a 4-Year Old Boy Treated by Excision: A Case report

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INTRODUCTION

Lymphangiomas are uncommon, hamartomatous, congenital malformations of the lymphatic system that involve the skin and subcutaneous tissues. They usually occur in children before the age of two years. We are reporting here a rare case of a gradually enlarging cavernous lymphangioma that arose at birth in a Filipino boy.

The objective of this paper is to create increased awareness of the clinical and histological characteristics of cavernous lymphangioma in order to enhance its diagnosis, thereby avoiding confusion with other disease entities. It is also its objective to discuss potential treatment options.

CASE REPORT

This is the case of a 4-year-old Filipino boy who presented with a gradually enlarging right thigh since birth with no accompanying symptoms. The patient's right thigh at birth was noted to be slightly enlarged as compared to the left thigh. X-ray and ultrasound were done at Tondo General Hospital, which revealed a huge tissue mass, probably an inflammatory process. The right thigh gradually enlarged as the child grew. At the age of one year, the patient was brought to the Pediatric Surgery Service of Philippine General Hospital where a diagnosis of lymphangioma was made. Complete blood count and X-ray of the right thigh were done. Surgical excision was advised but due to unavailability of bed, the patient was not admitted. He was then lost to follow-up.

Thirty-six months prior to referral, the patient's right thigh was noted to be erythematous and continuously enlarging. This time, there were intermittent bouts of high-grade fever. He was brought to the Pediatric Service of this institution and was admitted with a diagnosis of lymphangioma with cellulitis. The patient was given intravenous oxacillin 100 mg/kg/day, chloramphenicol 100 mg/kg/day and paracetamol 10 mg/kg/dose which provided relief of the fever and improvement of the cellulitis. He was then referred to the Surgery Service whose assessment was a tissue mass of the right thigh. Ultrasound and x-ray were requested which were not done. He was discharged improved and was lost to follow-up.

Fifteen months prior to referral, the patient had low to moderate grade fever accompanied by cough and colds. His enlarged right thigh was noted again to be erythematous. He was brought to the Pediatric Service of this institution and was admitted with the diagnosis of infected lymphangioma. He was given a course of antibiotic and was referred to the Pediatric Surgery Service. Ultrasound of the right thigh was done which revealed an inflammatory process; differential diagnoses were cellulitis, abscess, lipoma and lymphangioma. The Pediatric Surgery Service advised follow-up on an out-patient basis. He was discharged after 11 days with resolution of fever, cough, colds and improvement of the cellulitis.

Two weeks prior to referral, the patient's mother noted multiple minute vesicles on the affected thigh which was erythematous and tender and associated with moderate grade fever. The boy was again admitted at this institution with a diagnosis of infected lymphangioma. He was then given a course of antibiotics. Repeat ultrasound was done which

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revealed a huge tissue mass. Considerations included hemangioma and lymphangioma. A malignant soft tissue neoplasm could not be excluded. He was also referred to the Otolaryngology Service for evaluation of decreased hearing ability. Play Audiometry was recommended. He was then referred to the Dermatology Service for further evaluation and management.

Skin examination revealed an enlarged, fluctuant right thigh with girth at mid thigh measuring 52 cm, with erythematous to hyperpigmented verrucous plaques on the surface. The girth of the left thigh was 27 cm.

The skin punch biopsy of the hyperpigmented verrucous plaques of the right thigh showed a hyperplastic epidermis with focal thinning and marked spongiosis. In the upper and lower dermis were numerous dilated cystic spaces, lymphatic channels lined with flat endothelial cells, with proteinaceous material seen inside some of the cystic spaces. There were moderate superficial and deep infiltrates of lymphocytes and histiocytes and numerous neutrophils and plasma cells. The report was signed out as cavernous lymphangioma.

The patient was referred to the Vascular and Plastic Surgery Service of this institution for possible resection of the mass. He was also referred to Pediatric Service for complete immunization and to the Otolaryngology Service for evaluation of his hearing impairment.

After the patient was assessed as having cavernous lymphangioma, the Vascular and Plastic Surgery Service suggested a CT scan of the thigh to determine the extent of the lesion prior to excision of the mass. The gross operational findings were a 17 cm mass in its widest diameter on the antero-lateral aspect of the right thigh, extending from the inguinal area up to the knee. On opening, the mass was noted to be fibromuscular with small vacuoles containing serous fluid. The mass was adjacent to the muscle and fascia. Neuromuscular structures were not involved. The microscopic pathological finding was lymphangioma with no evidence of malignancy.

The patient was given intravenous ampicillin 100 mg/kg/day and ketorolac 0.5 mg/kg/day. He was discharged improved after 10 days and was given cefalexin 50 mg/kg/day and paracetamol 10 mg/kg/dose as home medications.

Figure 1

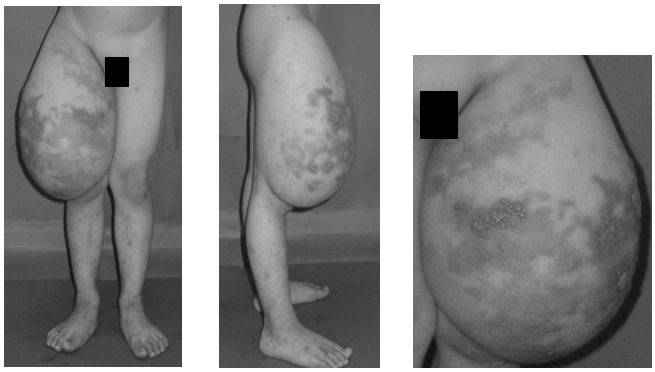

Figure 1A *Figure 1B* *Figure 1C*

Figure 1: Enlarged, fluctuant right thigh with girth at mid thigh measuring 52 cm (A) front view (B) side view (C)erythematous to hyperpigmented verrucous plaques on the surface.

Figure 2

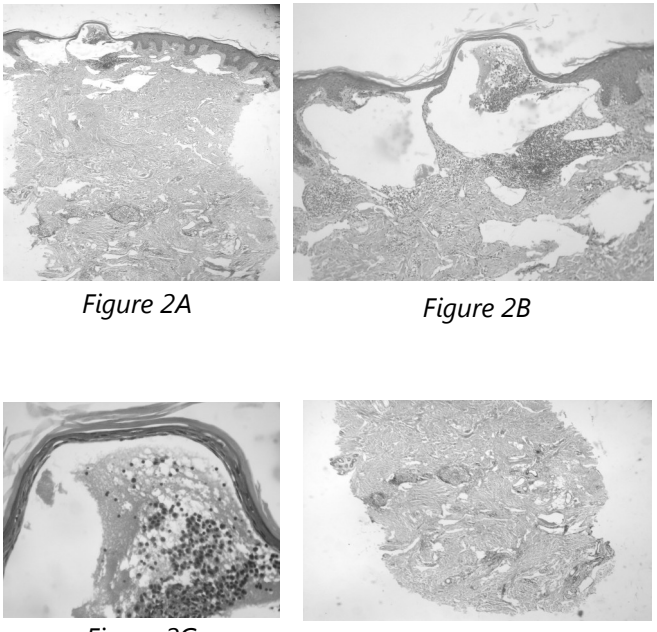

Figure 2A *Figure 2B*

Figure 2C *Figure 2D*

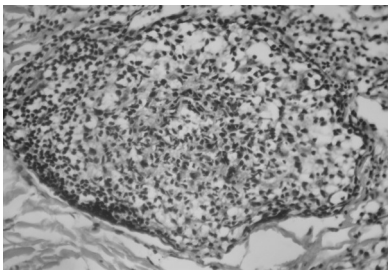

Figure 2E

Figure 2: (A) Scanning view: hyperplastic epidermis with focal thinning and marked spongiosis. (B)Low power: In the upper and lower dermis were numerous dilated cystic spaces, lymphatic channels lined with flat endothelial cells, with proteinaceous material seen inside some of the cystic spaces (C) High power (D) Scanning view: There were moderate superficial and deep infiltrates of lymphocytes and histiocytes and numerous neutrophils and plasma cells (E) High power

Figure 3

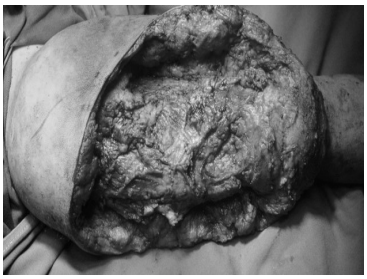

Figure 3A

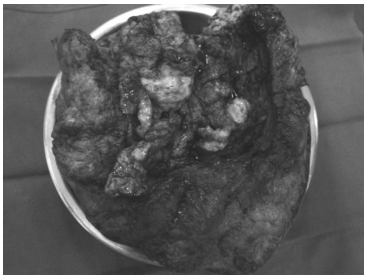

Figure 3B

Figure 3 (A) gross operational findings were a 17 cm mass in its widest diameter on the antero-lateral aspect of the right thigh, extending from the inguinal area up to the knee (B) specimen for pathology

Figure 4

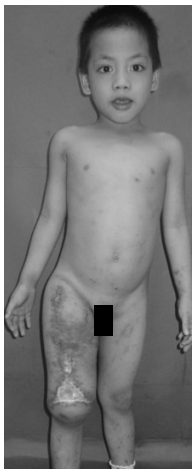

Figure 4A

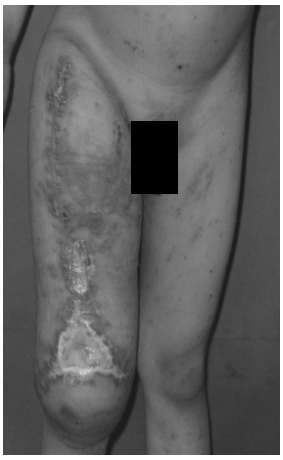

Figure 4B

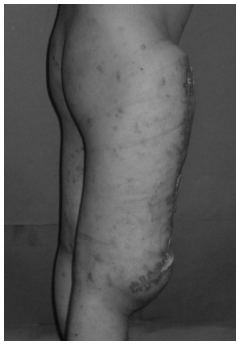

Figure 4C

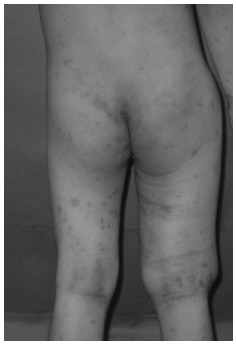

Figure 4D

Figure 4:Post-operational pictures(A) full view (B) front view (C) side view (D) back view

DISCUSSION

The lymphatic system is an extensive unidirectional system of blunt-ending vessels that retrieve excess fluid from the interstitium, transport it to regional lymph nodes, and return it to the venous system by way of the thoracic duct. The lymphatics absorb protein and lipid from the liver and small intestine.

Redenbacher was cited as first describing lymphatic malformation in the European literature in 1828.[1] The lesion is thought to represent a failure of the primitive lymph sac to connect to the venous system during embryogenesis. In 1938 Goetsch noted

that this sequestered lymphatic tissue forms cysts and that endothelial sprouts projected from the walls of the cysts which produce spaces.[1] The enlargement of the cyst is due to the accumulation of lymph. These sprouts may surround and destroy tissue and force the lesion into areas of least resistance between muscles and vessels, invading tissue planes. This invasion of local structures causes atrophy, fibrosis and hyalinization of the engulfed tissue.

Lymphangiomas, also known as lymphatic malformations are categorized into four types of lymphangiomas (lymphangioma circumscriptum, cavernous lymphangioma, cystic hygroma and diffuse lymphangiectasia.)

Cavernous lymphangioma is a rare benign proliferation of the lymphatic vessels that usually arises at birth in 50-60% of cases with equal sex indices. It usually involves the limbs. These lesions are deep seated in the dermis, forming a painless swelling or thickening of the skin, mucous membranes and subcutaneous tissue.

The diagnosis of lymphangiomas rests mainly on the clinical history and findings from physical examination and conventional light microscopy. In some infants with atypical lesions, observation of change over time or radiologic investigation is necessary to confirm the diagnosis. Ultrasonography is a useful and cost-effective diagnostic method but does not portray the extent of the lesion or its relation to adjacent structures. MRI can define the degree of involvement and the entire anatomy of the lesion. It can also help prevent unnecessary extensive and incomplete surgical resection.

Histological examination reveals large, irregular channels in the reticular dermis and subcutaneous tissue that are lined by a single layer of endothelial cells. An incomplete layer of smooth muscle often lines the walls of these malformed channels. The surrounding stroma consists of loose or fibrotic connective tissue with a number of inflammatory cells. As previously described, the histopathological result of the patient was consistent with cavernous lymphangioma.

Immunohistochemical studies may be useful in differentiating lymphangiomas from hemangiomas in difficult cases. Immunohistochemical studies for laminin may differentiate a normal blood vessel from a lymphangioma. High endothelial reactivity for the erythrocyte-type glucose transporter protein GLUT 1 is observed in hemangiomas and may provide a diagnostic tool to differentiate it from vascular malformation.

Typically, cavernous lymphangiomas appear as subcutaneous nodules with a rubbery consistency. They may have large dimensions. The area of involvement varies, ranging from lesions smaller than 1 cm in diameter to larger lesions that involve an entire limb.

The patient's clinical presentation as an enlarging fluctuant mass on the right thigh at birth is consistent with cavernous lymphangioma. This is further supported by the CT-scan showing a bulging, slightly hypodense soft tissue mass with cleavage between the mass and the muscular compartment seen except at its inferior portion. Haziness and slight decrease in density were also noted in the vastus lateralis and a portion of the vastus intermedius with no evidence of neurovascular or osseus involvement.

The major indications for treatment include life or function threatening lesions, those that tend to leave disfiguring scars or deformities, facial lesions, and those lesions that are exposed such as the extremities. Therapeutic decisions should be tailor-made to each individual lesion, and should always take into account psychosocial factors, including the emotional well being of both the child and parents. Because no treatment is without risks, all potential complications should be detailed prior to initiation of therapy.

Treatment of lymphatic malformations can be challenging and various approaches have been attempted. The most appropriate management of cavernous lymphangioma at any site is conservative surgical therapy with preservation of all important neurovascular structures.

Another option for cavernous lymphangioma is injection of sclerosing agents such as alcohol, steroids, bleomycin sulfate, tetracycline and OK-432 (picibanil).

Qin et al[2] described 200 patients with various types of head and neck lymphangiomas who were treated by inratumorous injection of bleomycin-A5. All 200 cases were followed from 2 to 10 years; 95 cases were followed for more than 5 years. The effective rate was 97% and the curative rate was 86.5%. No pulmofibrosis or other serious complications were found.

In a study made by Kobayashi et al[3], three patients with cavernous lymphangioma of the legs were treated. The suggested guidelines for treatment are extensive surgical resection of the mass and approximately two weeks after the operation, aspiration of serous fluid which has accumulated at the operation site. This is then followed by injection of OK-432 at the resected area.

Side effects include pyrexia and focal inflammation for several days, but the accumulation of serous fluid disappeared after the injection. There were no complications with any recurrence, joint contracture, or pain during a mean follow-up of 48 months.

Recurrence of larger lymphomas after surgical excision is common because the tumor closely involves important nerves and vessels. Saijo et al[4] reported that tumor resection recurred in one third of operations that were only partially successful. Patients who have grossly or microscopically infiltrative lesions should be counseled about the increased likelihood of recurrence, even if resection is thought to be complete. In our patient's case, the neurovascular structures were not involved and the likehood of recurrence is less.

The prognosis of cavernous lymphangioma heavily depends on early diagnosis with definitive surgical treatment. Full recovery is possible in patients whose lesions are completely resected.

CONCLUSION

Cavernous lymphangioma is a rare clinical entity that is defined as a congenital malformation of the lymphatic system that involves the skin and subcutaneous tissues.

A keen awareness of the clinical and histological features of cavernous lymphangioma can alert physicians to early detection and proper management. Several treatment options are available but the patient should be aware of the risk of recurrence with this disease.

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