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Case Report

Lymphatic Malformation of Inguinal region in an adult: A Case Report.

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Abstract

Lymphatic malformation are most commonly located in regions of confluence of major lymphatic channels, including the neck (75%), axilla (20%), mediastinum, retroperitoneum, pelvis and groin. We are reporting here a rare case in which a 35year old female presented with swelling over right groin and thigh. A provisional diagnosis was made on radiological investigations. Wide Local Excision with Tensor Fascia Lata Flap Reconstruction with Split Skin Graft was done and the diagnosis was confirmed on histopathology and immunohistochemistry.

Keywords: Lymphatic malformation, Acquired Progressive Lymphangioma, Benign lymphangioendothelioma.

Introduction

Lymphatic malformations represents the morphogenetic abnormality rather than a neoplasm. They are commonly located in regions of confluence of major lymphatic channels with predilection for head, neck and axilla. The other sites are mediastinum, retroperitoneum, pelvis and groin.[1,2] Lymphatic malformation accounts for approximately 4% of vascular malformations and 26% of benign vascular tumors.[3]Cutaneous Lymphatic malformation is classified as superficial and deep forms. Superficial forms, also referred as lymphangioma circumscriptum in which lymphatic vessels are localised and restricted to superficial dermis. Deep forms involve the deeper dermis and subcutaneous tissue. Acquired progressive lymphangioma is a slow growing cutaneous lymphatic malformation that deserves special mention because of its mimicry of well-differentiated angiosarcoma.[2]

Although these vascular anomalies are benign, they can cause morbidity because of its size, location and secondary infection. The mainstay of treatment is a combination of surgery and sclerotherapy.[1,3]

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Case Report

A 35 year old female patient presented to us with complaints of swelling over right groin region and right thigh since 2years. History of Non-foul smelling sero-sanguinous discharge present from area. There was past history of previous surgery 4years back in right groin region. No history of fever, pain, trauma, burning micturition and regional lymph node enlargement.

On General examination she was conscious, co-operative, oriented to time, place and person. Vital signs were within normal limits. No abnormality detected on systemic examination of Respiratory system, cardiovascular system and central nervous system. On local examination, there was a swelling

measuring about 30x25Cms involving the anterior lower aspect of Right thigh, lateral aspect of vulva and inguinal region associated with serosanguinous discharge. There was No local rise of temperature or tenderness present. Surgical scar over Right inguinal region was seen.

Ultrasonography showed an ill-defined heterogenous predominantly hyperechoic space occupying lesion, in deep subcutaneous plane of right inguinal region 7.7x2.4x6.4cm with a diagnosis of Hemangioma was made. MRI showed a diffuse heterogenous multilobulated soft tissue lesion of 30x15x15cm seen in the subcutaneous soft tissue of antero-lateral aspect of thigh not invading muscles of thigh and adjacent bony cortex shows no destructive changes with a diagnosis of lymphovascular malformation/hemangioma was made.Core biopsy of the lesion was performed and sent for histopathological examination. Vascular tumor of Intermediate grade was given in the diagnosis with a possibility of Epitheloid hemangioendothelioma (Figure 1A & B).

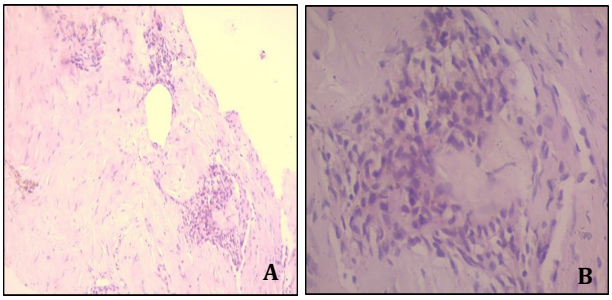

Figure 1A :(40x, H&E) & 1B:(100x, H&E): Core biopsy showing presence of islands of atypical cells amidst the fibro collagenous tissue.

Patient underwent Wide local excision and tensor fascia lata flap reconstruction with split skin graft.

Pathological examination:

On gross examination, received specimen of skin covered soft tissue mass from right thigh with right vulval region measuring 25x17x6 cms. External surface of skin shows scar measuring 7.5 cms (Figure 2). Cut surface of mass showed greyish brown lesion with spongy appearance measuring 17x8x2 cms (Figure3 & 4). The lesion was 3 cms from superior resected margin and medial resected margin; 7.5cm, 10cm and 0.2cm from inferior, lateral and deep resected margins respectively. Also identified two greyish brown lesions on medial aspect near to vulval region, one measuring 3x2x1.5 cms and other measuring 3x2x2cm.

Figure 2: Skin covered soft tissue mass.

Figure 3: Serial sections of the soft tissue mass.

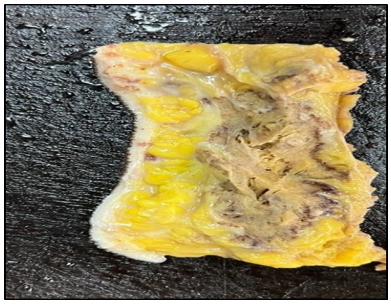

Figure 4: Cut section showed a greyish brown lesion with spongy appearance.

Microscopic examination of this lesion showed skin with epidermis, dermis and subcutaneous tissue. Subcutaneous tissue showed a diffuse lesion composed of lymphatic channels arranged in microcysts and macrocysts lined by flattened endothelial cells. Amidst these lymphatic channels were seen lymphoid aggregates forming lymphoid follicles and stroma showed areas of hemorrhage. (Figure 5A & B) Medial, lateral, superior, inferior, overlying skin and deep resected margin are free from the lesion and the two lesions nearer to the vulvar region also showed similar features as stated above. Diagnosis of Cutaneous lymphatic malformation of deep type was given. Immunohistochemistry showed the endothelial lining of the lesion positive for the endothelial marker CD31 and the lymphatic marker D2-40 (Figure 6A&B).

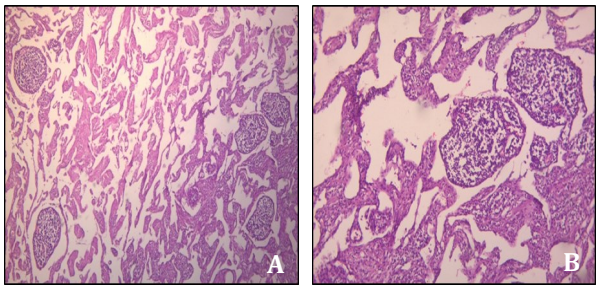

Figure 5A: (40x, H&E) & 5B:(100x, H&E): Microscopy of resection specimen showing microcysts and macrocysts lined by flattened endothelial cells with lymphoid aggregates.

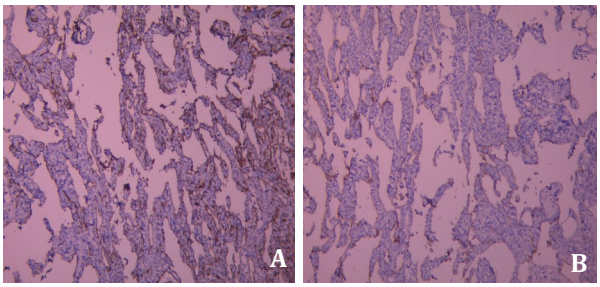

Figure 6: Endothelial cells expressing blood and lymph vessel markers. A) CD31, original magnification ×100.B) D2-40, original magnification ×100.

Discussion

Lymphatic malformation consists of dilated lymphatic vessels caused by occlusion of the lymphatic drainage system due to congenital malformations or acquired causes such as the effects of trauma, infection, or surgery.[4] Acquired progressive lymphangioma is a term used for a cutaneous slowly progressive lymphatic proliferation that occurs primarily in adults.[2] The other synonym of APL is Benign Lymphangioendothelioma. The incidence is equal in males and females. Most lesions are located on the extremities, especially the lower limb, but also occurs on the face, back and abdomen.[5] They typically arise as a solitary lesion on the trunk and limbs in both children and adults.[6]

Histological features that favour the diagnosis of lymphatic malformation over vascular malformation are lymphoid aggregates in the stroma and irregular lumens with widely spaced nuclei.[2] Immunohistochemically, markers like podoplanin (D2-40), LYVE-1, and PROX-1 can be used reliably for determining the lymphatic lineage.[2,7]

F. Messeguer et al described a case of acquired progressive lymphangioma which presented as a lesion in the hypogastrium characterized by nonspecific local discomfort ,intense pain and accompanied by mild edema of the scrotum and top of the right thigh that made walking difficult. In their case report they have summarized the clinical characteristics of acquired progressive lymphangioma described in the literature that Acquired progressive lymphangioma is more common in Head and neck (20%) followed by lower limbs (33%), upper limbs, shoulders, back, pectoral region and abdomen. The possible triggers for the lesion are Minor surgery, radiotherapy, arteriography and trauma.[8]

Jian-Wei Zhu et al reported a case of Acquired Progressive Lymphangioma presented in the Inguinal Area Mimicked Giant Condyloma Acuminatum.[3] Naveen Kumar Vittal et al in their case report described a case of Benign Lymphangioendothelioma which presented over left leg as a raised lesion in 25year old female and emphasized that trauma and hormonal stimulus, have been described in many pubescent and prepubescent patients because rapidly enlarging lesions.[7] Hwang et al reported a case of Acquired progressive lymphangioma in a 15-year-old boy who had a 10-year history of gradually enlarging lesions on his right foot.[9]

With the case presented here, the patient had a past history of surgery followed by swelling on the right groin region. On core biopsy based on the presence of islands of atypical cells seen amidst the fibro collagenous tissue, diagnosis of intermediate grade vascular tumor was made but later on resection specimen , Acquired Progressive Lymphangioma was made on histopathology and confirmed with immunohistochemistry. The differential diagnosis of APL should include malignant vascular tumors such as low-grade angiosarcoma and Kaposi sarcoma. The histologic finding of existence of cords of cells with minimal atypia projecting into the lumen enables low -grade angiosarcoma to be distinguished from APL.[8] Histologic features such as a lack of cellular atypia and mitotic figures or scanty inflammatory cells can exclude a diagnosis of an angiosarcoma.[3,10]

The local aggressive nature of APL with symptoms can affect the patient's quality of life. The factors that predict good response are the location of the lesion preferably head and neck, size less than 5cm and macrocystic architecture on microscopy and least response for infiltrating microcystic and combined microcystic-macrocystic lesions on histopathology.[2]

Conclusion

Acquired progressive lymphangioma, a benign lymphatic proliferation occurs as a response to prior trauma. The use of D2-40 is helpful to recognise the true lymphatic origin of the lesion. Awareness of this condition is important, so that misdiagnosis for malignant conditions such as Angiosarcoma and Kaposi Sarcoma can be avoided. Complications derived from subsequent growth can be minimized by thorough evaluation of the suspected cases by surgeons and pathologists to ensure an early diagnosis leading to resection.

References

1. Rahman M, Alim SA, Akhter M, Sultana S. Inguinal Lymphatic Malformation: A Rare Case Report. Chattagram Maa-O-Shishu Hosp Med College J 2013; 12(3):62-3.
2. Goldblum JR, Folpe AL, Weiss SW. Tumors and Malformations of Lymphatic vessels. In: Goldblum JR, Folpe AL, Weiss SW. Enzinger and Weiss's Soft Tissue Tumors.6th edition. Philadelphia: Elsevier 2014:735-42.
3. Zhu JW, Lu ZF, Zheng M. Acquired progressive lymphangioma in the inguinal area mimicking giant condyloma acuminatum. Cutis 2014 ;93 (6):316-9.
4. Sharma M, Mallya V, Khurana N, Kumar P, Duggal R. Lymphovascular Malformation - A Report of Two Cases. J Clin Diagn Res 2017 ;11(5):ED03-ED04.
5. Calonje JE, Fletcher CDM. Vasular tumors. In; Fletcher CDM Diagnostic Histopathology of Tumors. 4th edition. Philadelphia: Elsevier 2013:73-74.
6. Meunier L, Barneon G, Meynadier J. Acquired progressive lymphangioma. Br J Dermatol 1994;131 (5):706-8.
7. Naveen Kumar Vittal et al. Benign Lymphangioendothelioma- A Rare Entity. JCDR 2016;10(1): WD01-WD02
8. Messeguer F, Sanmartín O, Martorell-Calatayud A, Nagore E, Requena C, Guillén-Barona C. Linfangiomaprogresivo adquirido (linfangioendotelioma benigno) [Acquired progressive lymphangioma (benign lymphangioendothelioma)]. Actas Dermosifiliogr 2010;101(9):792 -7.
9. Hwang LY, Guill CK, Page RN, Hsu S. Acquired progressive lymphangioma. J Am Acad Dermatol 2003;49(5):250-1.
10. Guillou L, Fletcher CD. Benign lymphangioendothelioma (acquired progressive lymphangioma): a lesion not to be confused with well-differentiated angiosarcoma and patch stage Kaposi's sarcoma: clinicopathologic analysis of a series. Am J Surg Pathol 2000;24(8):1047-57.