LETTER TO THE EDITOR

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Lymphangioma Presenting as Juxta-Articular Swelling In Children: A Case Series

Deepak Jain,* Harpal Singh Selhi, Mohd Yamin

Dear Sir,

Lymphangiomas are rare congenital malformations arising due to the failure of development of communications between lymphatic channels with subsequent sequestration and dilatation of lymphatic tissue. Majority of these lesions are seen in the neck and facial region while other sites such as axilla, mediastinum, retroperitoneum, mesenteric and groin have also been reported [1]. The occurrence of lymphangiomas in upper or lower limbs has been published in a few case reports [2, 3]. We report our experience of treating lymphangiomas presenting as isolated juxta-articular swellings in paediatric population. These lesions can be confused with intra-articular swellings / effusions and need to be differentiated from them. The clinical features, diagnostic workup and operative findings of the cases in our series are given in Table 1. All the patients underwent surgical excision for these swellings under regional or general anaesthesia. The average time of follow up after surgery was 10 months (range 6 to 14 months). There was no post operative infection, recurrence or any other complication in any patient.

Table 1: Clinical details of patients with diagnostic workup and operative findings

No	Age/ Gender	Clinical examination	Radiological investigations	Operative findings
1	2 year/ Male	• Swelling right elbow lateral aspect -12 cm × 5 cm, margins indistinguishable from elbow joint. • Soft, compressible and transillumination test positive.	• Ultrasound: Multiple hypoechoic areas filled with clear fluid separated by septae. • MRI: Multilocular swelling with hyperintense signal on T2 images and showing no communication with elbow joint.	Surgical excision done. Tumor adherent to joint capsule.
2	6 month/ Male	• Swelling right ankle – 3 cm × 5 cm, anterolateral aspect. • Firm, non compressible, transillumination test negative.	MRI: Single loculated, with heterogeneous solid material.	Excision biopsy done. Tumour adherent to joint capsule.
3	1.5 year/ Female	• Swelling left knee – 3 cm × 3 cm, inferolateral to patella anteriorly. • Firm, non compressible.	Ultrasound: Single loculated lesion filled with heterogeneous material.	Excision biopsy done. No adhesions found.
4	2 year/ Female	• Swelling left knee – 4 cm × 4 cm, in popliteal fossa. • Soft, compressible.	Ultrasound: Single cavity with clear fluid.	Excision biopsy done. No adhesions found.
5	13 year/ Male	• Swelling right wrist – 3 cm × 4 cm on dorsal aspect of the wrist. • Soft, non compressible.	FNAC: Clear hypocellular fluid.	Excision biopsy done. Tumor adherent to joint capsule.

The clinical presentation of the cases in our series is different from those reported in literature. All the cases presented with isolated swelling of the affected joints noted either at birth or within subsequent three months. On examination, they were eccentrically located in proximity of the joints which helped differentiate them from joint effusion. They were soft on palpation except in cases 2 and 3 which were firm and in variance with the typical presentation of lymphangiomas. Transillumination which is characteristic of lymphangioma was noted in only case 1 as the other lesions were small or had solid contents. Ultrasonography was done to find out the relation of the swellings with the joints. MRI was done in cases 1 and 2 to ascertain whether the lesions communicated with the joints and their relationship with neurovascular structures.

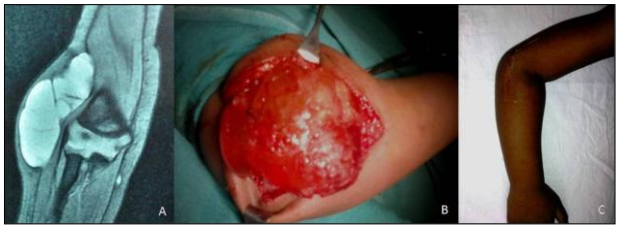

Figure 1 (A, B, C) A- MRI of elbow showing multilocular swelling with hyperintense clear signal, B-. Operative photograph showing typical findings of multilocular swelling with thin walls, C- photograph showing follow up at one year.

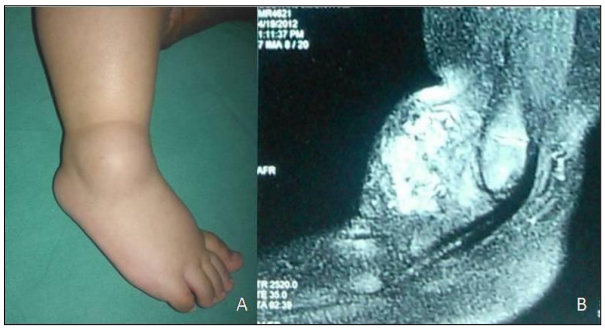

Figure 2: (Case 2) Clinical photograph of ankle swelling (A) and MRI of ankle showing solid tumour with heterogeneous signal (B).

In case 1, it was possible to make a pre operative diagnosis of lymphangioma based on clinical findings and diagnostic workup (Fig. 1). In the rest of the cases, excision biopsy was done and the diagnosis was established after histopathological examination. In three cases (Cases 1, 2 and 6) lymphangioma was adherent to the joint capsule in some areas and partial excision of joint capsule and synovium was required to ensure complete excision of the tumour. The typical finding of thin tumour sheaths filled with clear fluid was not present in cases 2 and 3 which were solid tumours filled with heterogeneous cellular material (Fig. 2). This is possible if spontaneous haemorrhage occurs in the cystic cavity with subsequent fibrosis. In our series, all the cases were macrocystic type of lymphangiomas except case 1 which was of mixed type.

Surgical excision is the mainstay of treatment of lymphangiomas. Sclerotherapy is also used to treat such lesions and agents like bleomycin, doxycycline and OK 432 have been used particularly in large lesions and those located in inaccessible areas.[2,4]. Ozeki et al have advocated the use of propranolol (a beta blocker) as an adjunct in treating intractable lymphatic malformations. [5] Vaccum assisted closure has also been used to assist the closure of wounds after surgical excision.[6] However, in our series all the lesions were isolated with well defined margins and showed excellent results with surgical excision.

REFERENCES

1. Masood SN, Masood MF. Case report of fetal axillo-thoraco-abdominal cystic hygroma. Arch Gynecol Obstet. 2010; 281:111-5.
2. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic hygroma: An overview. J Cutan Aesthet Surg. 2010; 3:139-44.
3. Dhrif AS, El Euch D, Daghfous M, Cherif F, Mokni M, Dhahri AB. Macrocystic lymphatic lymphangioma (cystic lymphangioma) of upper extremity: A case report. Arch Pediatr. 2008;15:1416-9.
4. Mirza B, Ijaz L, Iqbal S, Mustafa G, Saleem M, Sheikh A. Cystic hygroma of unusual sites: Report of two cases. Afr J Paediatr Surg. 2011; 8:85-8.
5. Ozeki M, Kanda K, Kawamoto N, Ohnishi H, Fujino A, Hirayama M, et al. Propranolol as an alternative treatment option for pediatric lymphatic malformation. Tohoku J Exp Med. 2013;229:61-6.
6. Katz MS, Finck CM, Schwartz MZ, Moront ML, Prasad R, Timmapuri SJ, et al. Vacuum-assisted closure in the treatment of extensive lymphangiomas in children. J Pediatr Surg. 2012; 47:367-70.

AFFILIATION:

Dayanand Medical College and Hospital, Ludhiana, India

CORRESPONDENCE:*

Deepak Jain,

Dayanand Medical College and Hospital, Ludhiana, India.

Email:* deepakjainorth@rediffmail.com

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