Case Report: Retro-auricular Lipoblastoma a Rare Tumor at an Uncommon Site

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Abstract

Lipoblastoma is a rare benign tumor present commonly in the first three years of life. Here, we present a 6-month-old baby who presented with a left retro-auricular mass since birth. The mass was gradually increasing in size. Ultrasound was performed which raised the differential diagnosis of cystic hygroma versus a lipomatous tumor. Magnetic resonance imaging (MRI) showed a large subcutaneous fat intensity mass in the left post-auricular region. The lesion was excised with clear margins and histopathology was reported as lipoblastoma. To the best of our knowledge, this is the first case of head and neck lipoblastoma from Oman.

Keywords: Rare; Lipoblastoma; Retro-auricular.

Introduction

Lipoblastoma is a rare benign tumor that typically occurs in children under the age of three. It arises from immature fat cells and is commonly found in the extremities or trunk. The exact cause of lipoblastoma is unknown, and it is not thought to be hereditary.[1,2] Symptoms of lipoblastoma can include a painless, slow-growing mass or lump, usually found under the skin. The tumor may be soft and easily moveable and can sometimes be mistaken for a cyst or other type of benign growth. Rarely, lipoblastomas can cause pain, compression of nearby tissues or organs, or other symptoms depending on the location of the tumor. The diagnosis of lipoblastoma is typically made through imaging studies, such as ultrasound, CT, or MRI. A biopsy may also be performed to confirm the diagnosis.[3] Treatment of lipoblastoma usually involves surgical removal of the tumor. In some cases, radiation therapy may be used after surgery to reduce the risk of recurrence. With appropriate treatment, the prognosis for lipoblastoma is generally good, and most patients experience a full recovery. However, in some rare cases, the tumor may recur or progress to a more aggressive form of cancer.[4]

Case Report

A 6-month-old female infant was referred from a secondary care hospital who had developed left posterior auricular swelling. She has no previous medical or surgical background. Delivered at term with uneventful Antenatal, natal, and postnatal periods. Notably, prenatal imaging did not reveal any evidence of the observed mass.

Following the appearance of the mass at the age of 2 weeks, it displayed progressive enlargement since then. The mass has remained asymptomatic and has not shown any skin abnormalities. However, it caused anterior displacement of the left ear's pinna due to its size. The child was thriving well and gaining her milestones as for

her corresponding age. There was no history of constitutional symptoms. Also, her parents have not reported any additional masses in other areas, nor is there a history of cutaneous hyperpigmentation or hypopigmentation. The child was vaccinated as per Oman's Ministry of Health's national extended vaccination program. The mother denied any medication usage during pregnancy. Unremarkable family history.

On Assessment: No dysmorphic features, nor skin abnormalities including edema or lymphadenopathy. The mass examination indicated a localised swelling in the left retro-auricular region displacing the pinna of the left ear and going medially to the occipital area with no overlying skin changes [Figure 1]. Hair was growing normally above the mass. On palpation, the mass was soft and puffy in consistency characterized by a smooth surface. Its dimensions measured around 10 x 5x 4 cm, with no notable changes in temperature, and it was attached to underlying tissue without involvement of the overlying skin. Both auscultation and transillumination tests did not yield any relevant findings. A neurological examination of the face revealed no abnormalities with intact functions of facial and trigeminal nerves.

Figure 1: Swelling in the retro-auricular region.

An ultrasound examination was done to rule out cystic hygroma but it showed an adipocytic tumor. MRI of the mass showed a large abnormal signal intensity mass in the scalp soft tissues of the left temporo-occipital region. T1 weighted sequences show that the lesion is showing hyper-intense signals [Figure 2 A]. T2 weighted sequences show that this lesion was also hyperintense [Figure 2 B]. T1-fat sat sequences show that this lesion is showing fat suppression representing that it has macroscopic fat [Figure 2 C]. On post-contrast sequences, this lesion does not show any significant enhancement representing that this is likely of benign aetiology [Figure 2 D]. The overall finding is a large fatty lesion in the soft tissue of the scalp on the left side with benign features.

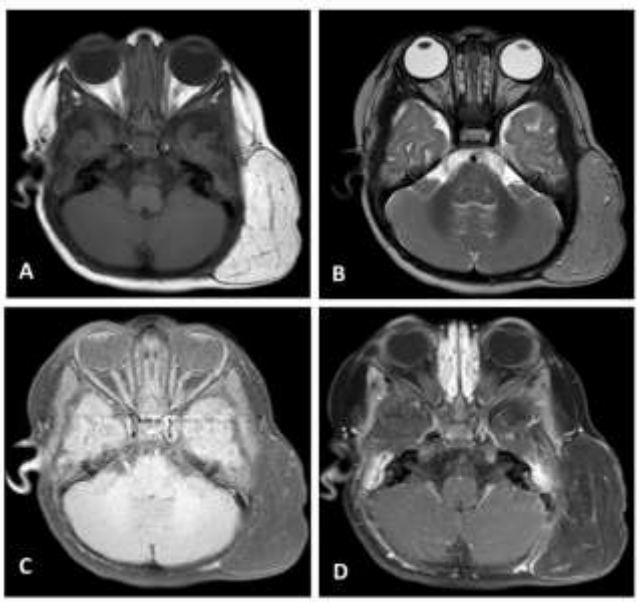
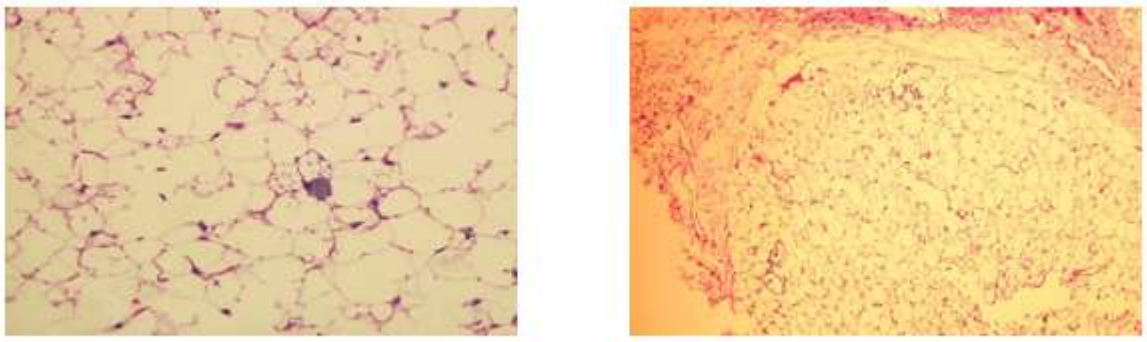


Figure 2: MRI of the mass showed a large abnormal signal intensity mass in the scalp soft tissues of the left temporo-occipital region.

At this point, the surgical team decided to proceed with total excision of the mass and to be evaluated by histopathology. Surgery revealed a mass measured 6 x 6 inches: soft in consistency, and not adherent to underlying vital structures (nerves or blood vessels). There was only one blood vessel at the base supplying the mass. Before amputation or tying it, a nerve stimulation is done to confirm that it is not attached to the nerves, which came as negative.

The lesion was completely excised with clear margins. Histopathological examination revealed a thinly encapsulated lobular neoplasm. The lobules were composed of variable-sized adipocytes [Figure 3 A]. Individual cells showed multi-vacuolated cytoplasm with hyperchromatic indented nuclei (lipoblasts) [Figure 3 B]. Morphology with age and clinical presentation favoured the diagnosis of lipoblastoma.



A B

Figure 3: Histopathological examination revealed a thinly encapsulated lobular neoplasm.

The patient then was followed after 3 months, The wound was healing well with mild seroma collection. Neurological examination was unremarkable.

Discussion

Lipoblastoma is a rare tumor, and there is limited data on its incidence and prevalence. However, there have been many case reports and case series describing individual cases of lipoblastoma in the literature.[5] Lipoblastoma is derived from embryonic fat, with only a limited number of cases reported globally.[6] According to a review published by Pediatr Int. in 2017, lipoblastoma accounts for only 0.3% to 2.0% of all soft tissue tumors in children. with the majority of cases occurring in children under the age of 5. The review also found that lipoblastoma was more common in boys than

girls, with a male-to-female ratio of approximately 2:1.[7,8] In general, adipose tumors are rare in children. Among soft tissue tumors present in the first two decades of life, they represent almost 6%, and approximately 30% of them are lipoblastomas.[9]

Case reports about lipoblastoma worldwide are variable in terms of body parts origin, mostly found in extremities, representing 50-60%. Followed by 20-30% presenting in the superficial parts of the body (axilla, groin, trunk, labia, etc.), whereas 15-20% are present in the head and neck region, and less than 10% present in retroperitoneal and mediastinal regions as in a case reported in Royal Hospital in Oman.[10,11]

Two cases of lipoblastoma have been reported from Oman, both presented in children less than 3 years old. One presented with an extremely rare location, posterior mediastinal mass with spinal epidural extension, and the other one presented with right-sided neck swelling. This exemplifies the rarity of such conditions.[6,10]

It may present as a progressively enlarging fatty, soft, and painless soft tissue mass, identical to benign lipomas, or as compressive symptoms to adjacent structures.[11]

Lipoblastomas have been categorized into two types, 70% of them are encapsulated, superficial, and well-circumscribed lipoblastomas, and 30% are lipoblastomatosis, a diffuse, infiltrative, non-capsulated, deeply located, and multicentric lesions.[11,12] Although it may cause local aggression, lipoblastoma usually has an excellent prognosis, especially if resected completely.[11] Local recurrence mostly occurs with patients presenting

with diffuse lipoblastomas, occurring in 9-22% of the cases.[13] Lipoblastoma has no potential malignant transformation or dedifferentiation.[13]

The differential diagnosis for a mass with fatty content includes lipoma, lipoblastoma, teratoma, and liposarcoma.[6] Teratoma was excluded from this case due to the absence of calcifications, ossifications, or any other germ cell tissue. Also, it was less likely to be liposarcoma as it is very rare in children less than 10 years old and the benign-looking mass in imaging. However, it was difficult to differentiate between lipoma and lipoblastoma in this case, and total surgical excision with histopathological examination was required.

Although the diagnosis of lipoblastoma is not usually taken into consideration preoperatively as it is very similar in the presentation of lipoma, and excessive work-up is rarely done unless malignancy is suspected, it is usually diagnosed with histopathological examination post-op. Ultrasound and MRI play an important role in characterizing the internal features, local growth, and invasion patterns that are specific to this rare benign neoplasm.[6] Some US findings might suggest the diagnoses of lipoblastoma such as mass size 5>cm, lobulation in shape, and heterogeneous echotexture. Lipoblastoma can be differentiated from other soft tissue malignancies by sonoelastography as it is increased in superficial malignancies.[14] Some studies suggest the use of Contrast-enhanced ultrasound (CEUS) which is one of the new modalities to study vascularization of liver and renal masses, however, it is poorly studied in cases of soft tissue masses, and the use in the head and neck area. CT was determined to be of limited utility in distinguishing lipoblastoma, but it may be of use for adjacent structure evaluation like bones.[15] Furthermore, MRI is the best imaging modality and more helpful than CT, as well to avoid extra radiation at a younger age. MRI Findings that suggest Lipoblastoma are found to be: solid appearance, encapsulation, and lobulation with heterogeneously hyperintense signals both in T1 and T2.[16] More valuable in MRI, is the fat suppression sequence which confirms the presence of a lipomatous component that is suggestive of Lipoblastoma.[16,17] Some studies suggest using the Apparent Diffusion Coefficient (ADC) in the diagnosis of soft tissue masses to differentiate malignant vs benign tumors, however, not widely used due to the overlapping values between different causes, and false negative rate. [18,19]

Histopathology serves as the definitive diagnostic tool for Lipoblastoma.[10] Histopathology shows different stages of adipocytes (white fat) variable in maturations, ranging from primitive cells with spindles or stellate to multivaculated lipoblasts, small univacuolated signet ring-type lipoblasts, and large mature adipocytes.[20] The cells usually are lobulated and septated with either Cells, blood vessels, or myxoid foci.[21] Immunocytochemical studies have been conducted in some literature which was immunoreactive for S100 and CD34; Mib-1, along with Abnormalities in Cytogenetics in Chromosome 8 (8q24.1.), 6, 7 & 17, however, it is worth noting that these findings have been somewhat non-specific and lack comprehensive investigation.[25)]

Surgical resection is the mainstay of the management of lipoblastoma, especially due to the need for histopathology for diagnosis confirmation. In many cases, the tumor was treated with surgical excision alone, and most patients had a favorable outcome with no recurrence of the tumor.[22,23] However, there have been rare reports of lipoblastoma recurring or progressing to a more aggressive form of cancer, such as liposarcoma. These cases highlight the importance of long-term follow-up and monitoring of patients with lipoblastoma, even after successful treatment.[24,25]

Conclusion

Lipoblastoma is a rare tumor that is more rare in the head and neck. Excision with clear margins is the treatment of choice.

References

1. Chun YS, Kim WK, Park KW, Lee SC, Jung SE. Lipoblastoma. J Pediatr Surg 2001 Jun;36(6):905-907.

2. Kerkeni Y, Sahnoun L, Ksia A, Hidouri S, Chahed J, Krichen I, et al. Lipoblastoma in childhood: about 10 cases. Afr J Paediatr Surg 2014;11(1):32-34.

3. Gilbert TJ, Goswitz JJ, Teynor JT, Griffiths HJ. Lipoblastoma of the foot. Skeletal Radiol 1996 Apr;25(3):283-286.

4. McVay MR, Keller JE, Wagner CW, Jackson RJ, Smith SD. Surgical management of lipoblastoma. J Pediatr Surg 2006 Jun;41(6):1067-1071.

5. Choi HJ, Lee YM, Lee JH, Kim JW, Tark MS. Pediatric lipoblastoma of the neck. J Craniofac Surg 2013;24(5):e507-e510.

6. Ambusaidi FM, Al-Mammari L, Al-Brashdi Y, Al-Shamsi RM. Imaging features of Lipoblastoma. Int J Pediatr Adolesc Med 2022 Mar;9(1):69-72.

7. Susam-Sen H, Yalçın B, Kutluk T, Cahit Tanyel F, Haliloğlu M, Orhan D, et al. Lipoblastoma in children: Review of 12 cases. Pediatr Int 2017 May;59(5):545-550.

8. Rajput S, Alsubaie SA, Singh K, Huang TC, Moran SL. Lipoblastoma of the extremities. J Plast Reconstr Aesthet Surg 2022 Oct;75(10):3761-3767.

9. George HL, Unnikrishnan PN, James LA, Perry DC, Garg NK, Bruce CE. Lipoblastoma–an unfamiliar but important diagnosis. A case series and literature review. Acta Orthop Belg 2009 Aug;75(4):533-536.

10. Raman Sharma R, Mahapatra AK, Pawar SJ, Sousa J, Musa MM. An unusual posterior mediastinal lipoblastoma with spinal epidural extension presenting as a painful suprascapular swelling: case report and a brief review of the literature. J Clin Neurosci 2002 Mar;9(2):204-207.

11. Alaa Ghallab, Elmahmoud M, M.A. Mohammed Alhadad, Khalil Albatniji, Ameen Alsaggaf, A. Gawad Elhendy, et al. Cervicothoracic lipoblastoma: three case reports and review of literature. Ann Pediatr Surg 2021 Nov;17(1).

12. Chung EB, Enzinger FM. Benign lipoblastomatosis. An analysis of 35 cases. Cancer 1973 Aug;32(2):482-492. doi:10.1002/1097-0142(197308)32:2<482:aid-cncr2820320229>3.0.co;2-e.

13. Gnepp DR. Diagnostic Surgical Pathology of the Head and Neck. 2nd ed. Saunders; 2009.

14. Catalano O, Varelli C, Sbordone C, Corvino A, De Rosa D, Vallone G, et al. A bump: what to do next? Ultrasound imaging of superficial soft-tissue palpable lesions. J Ultrasound 2019.

15. Pham NS, Poirier B, Fuller SC, Dublin AB, Tollefson TT. Pediatric lipoblastoma in the head and neck: a systematic review of 48 reported cases. Int J Pediatr Otorhinolaryngol 2010 Jul;74(7):723-728.

16. Gupta P, Potti TA, Wuertzer SD, Lenchik L, Pacholke DA. Spectrum of Fat-containing Soft-Tissue Masses at MR Imaging: The Common, the Uncommon, the Characteristic, and the Sometimes Confusing. Radiographics 2016;36(3):753-766.

17. Sidhu PS, Cantisani V, Dietrich CF, Gilja OH, Saftoiu A, Bartels E, et al. The EFSUMB Guidelines and Recommendations for the Clinical Practice of Contrast-Enhanced Ultrasound (CEUS) in Non-Hepatic Applications: Update 2017 (Long Version). Ultraschall Med 2018 Apr;39(2):e2-e44.

18. Corvino A, Sandomenico F, Setola SV, Corvino F, Tafuri D, Catalano O. Morphological and dynamic evaluation of complex cystic focal liver lesions by contrast-enhanced ultrasound: current state of the art. J Ultrasound 2019 Sep;22(3):251-259.

19. Corvino A, Catalano O, Setola SV, Sandomenico F, Corvino F, Petrillo A. Contrast-enhanced ultrasound in the characterization of complex cystic focal liver lesions. Ultrasound Med Biol 2015 May;41(5):1301-1310.

20. Coffin CM, Lowichik A, Putnam A. Lipoblastoma (LPB): a clinicopathologic and immunohistochemical analysis of 59 cases. Am J Surg Pathol 2009 Nov;33(11):1705-1712.

21. Collins MH, Chatten J. Lipoblastoma/lipoblastomatosis: a clinicopathologic study of 25 tumors. Am J Surg Pathol 1997 Oct;21(10):1131-1137.

22. Sheckter CC, Francis CS, Block V, Lypka M, Rizvi M, Urata MM. Multifocal lipoblastoma of the face. J Craniofac Surg 2012 Nov;23(6):e585-e587.

23. Alperovich M, Ayo D, Staffenberg DA, Sharma S. Lipoblastoma of the hand and cleft palate: is there a genetic association? J Craniofac Surg 2014;25(2):e189-e191.

24. Demir F, Öztürk S, Balta H, Erol FS. Lipoblastoma of scalp in a 4-month-old patient: an extremely rare case report and review of the literature. Childs Nerv Syst 2021.

25. Merzouqi B, Laachoubi M, Oukessou Y, Mahtar M. Head and neck lipoblastomas: Report of 3 cases and review of the literature. Int J Surg Case Rep 2021 Jul;84:106050-0.