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Case Report

Lipoblastoma of the Scalp: A Case Report and Literature Review

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Abstract Lipoblastoma is a rare benign mesenchymal tumours arising from embryonal white fat and most often encountered on trunk and extremities. We report a case of lipoblastoma of the scalp in a 3-month-old boy with review of the literature. This is to our knowledge the second case of lipoblastoma of the scalp reported in published literature in English. Here we illustrate the diagnostic challenge of lipoblastoma of infancy.

Key words Lipoblastoma; Scalp; Tumour

Introduction

Lipoblastoma is a rare, rapidly growing though benign neoplasm arising from embryonic adipose tissue. It occurs almost exclusively in infants and children less than 3 years of age. They most commonly presented as painless progressively growing subcutaneous mass on the trunk or extremities.[1-4] There is lack of characteristic clinical feature to differentiate from other soft tissue tumours clinically. Rapid growth and lack of timely diagnosis often render surgery mutilating. We present a child with scalp lipoblastoma but initially misdiagnose as infantile haemangioma. Our case illustrates the diagnostic dilemma of this rare tumour.

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A 3-month-old boy first presented to general paediatrician with a progressively enlarging mass over right parietal scalp since 4 weeks of life. There is no history of trauma and no family history of congenital anomalies. Physical examination revealed a 1.5 cm mobile mass over right parietal scalp with no skin changes or regional alopecia. Ultrasonography (USG) has been arranged afterward with the provisional diagnosis of non-specific scalp nodule. USG showed 1.7 cm oval-shaped soft tissue mass in the hypodermis of the scalp with venous flow present intra-lesionally which is suspicious of scalp haemangioma. It was thus managed as infantile haemangioma with conservative management provided. However, there was progressive increase in the size during subsequent follow-up: 3 cm at 4 months and 4 cm at 7 months of age respectively. Propranolol was started after assessment by paediatric dermatologist. There was further interval growth of the mass to 6 cm at 8 months of age despite escalating dose of propanolol. He was referred to our department for the diagnosis of atypical haemangioma at 13 months of age. Magnetic resonance imaging (MRI) was then arranged in our institution and showed a predominantly fatty tumour with internal septae arising from the right parietal scalp (Figure 1). At that juncture, diagnosis was redefined as scalp lipoblastoma. Surgical excision with primary skin closure was performed at 17

months of age and the immediate postoperative recovery was uneventful (Figure 2). The patient has been followed up at regular interval for 1 year with no evidence of recurrence.

Histologically, the tumour comprised of multilobulated adipose tissue transversed by thin fibrous septa and centered in subcutis. The constituent cells are mostly mature adipocytes, some variation in size and with occasional lipoblasts. Mild patchy myxoid change is noted. Resection margin is clear. Pleomorphic adenoma gene 1 (PLAG1) translocation is positive. The histological features are compatible with lipoblastoma.

Discussion

Lipoblastoma is a rare benign mesenchymal tumours arising from embryonal white fat. It occurs almost exclusively in infants and children less than 3 years of age.[1-4] Median age at presentation in literature was 22.6 months with slight male preponderance.[3] It most commonly presents as painless progressively enlarging mass and they are most commonly found in trunk or extremities.

With regard to the histology, there were 2 variants reported: an encapsulated localised form (lipoblastoma) and an infiltrative diffuse form (lipoblastomatosis). Histologically, both forms composed of lipoblasts, plexiform vessels, focal myxoid stroma which overlaps with lipoma, myxoid liposacroma and hibernoma.[3-6] Recent cytogenetic studies identifies association of lipoblastoma with a rearrangement of a region on the proximal part of

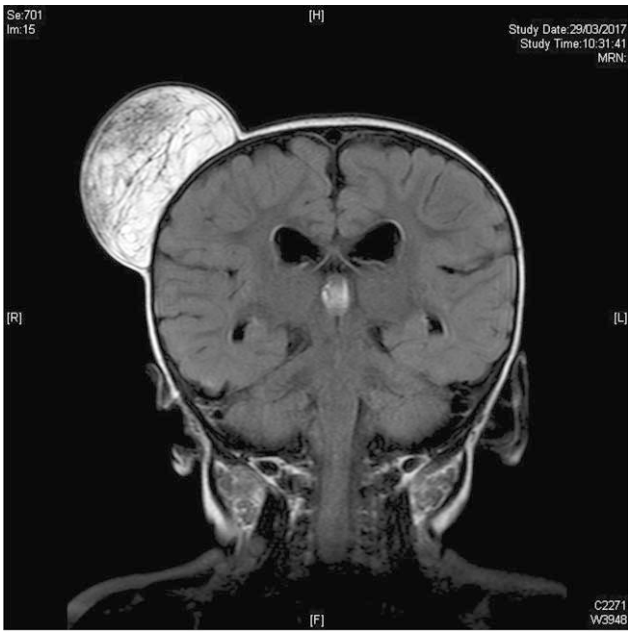

Figure 1 Coronal T1-weighted FLAIR MRI image of scalp demonstrating a large and well-circumscribed lesion at right parietal scalp.

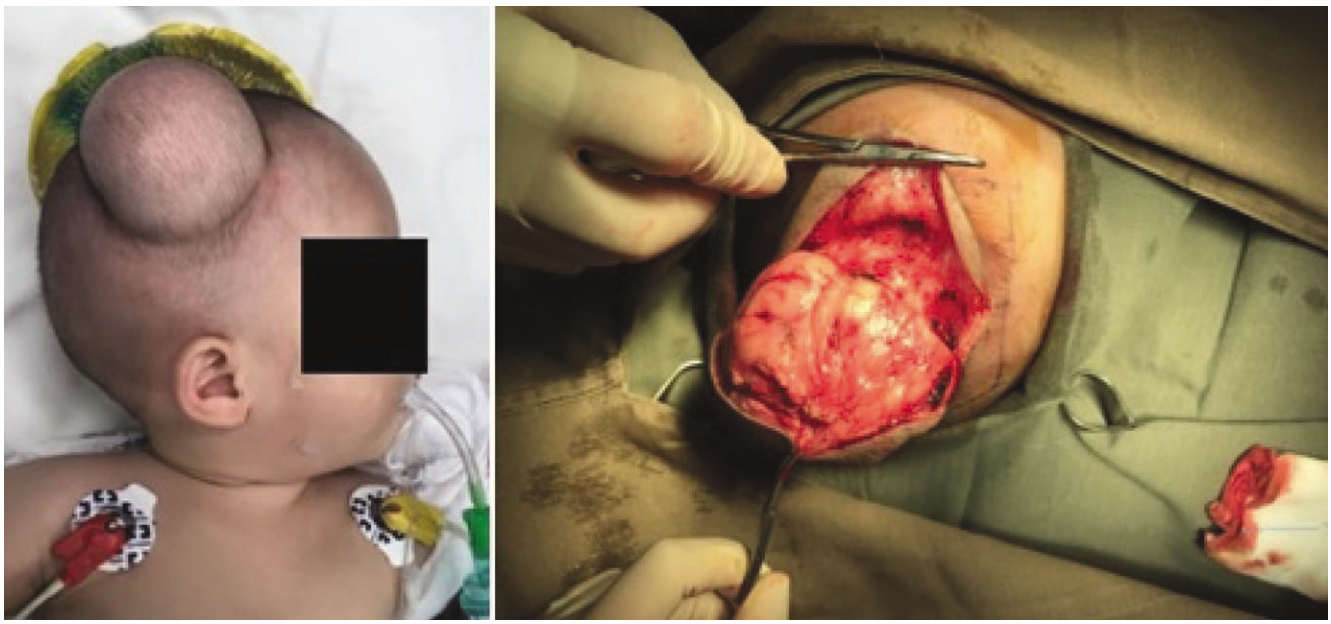

Figure 2 Pre-operative photo of the scalp lipoblastoma (left); Intra-operative photo of surgical excision of the scalp lipoblastoma (right).

the long arm of chromosome 8, 8q11-13 which upregulates PLAG1 and molecular analysis is important for accurate diagnosis.[7,8]

Local excision with complete resection is the standard of care.[4,6] However, total excision may not be feasible in certain cases due to close proximity to vital structures or in the setting of lipoblastomatosis. Despite the capability of rapid growth, the prognosis is excellent. The reported local recurrence rate ranges from 5-25% in different series due to incomplete excision.[3,4] They are believed to occur in the setting of lipoblastomatosis. Thus, close follow-up is suggested for early detection of recurrence, especially in cases with subtotal resection.

The natural history of unresected lipoblastoma is still largely unknown. Mognato et al[6] reported a case of spontaneous resolution of biopsy-proven lipoblastomatosis and he proposed the 'wait-and-see' approach in managing lipoblastoma. This was the only case of spontaneous resolution reported in the literature. However, expectant management is suggested given the rapid growth observed in most of the patients. Given the lack of diagnostic radiological features, excision is warranted to exclude sinister pathology and preventing pressure symptoms from further growth.[4,5]

Our case report highlights the clinical challenge in making accurate and early diagnosis of lipoblastoma as it mimicks an infantile haemangioma clinically and sonographically at the beginning. Prompt detection of atypical features with early radiological investigation is helpful to downscale the operation. Clinician should aware of the differential diagnosis of lipoblastoma in patients presenting with rapid-growing masses in children under the age of 3. The benign nature of this rare tumour warrants conservative surgical approach to avoid mutilating or extensive area of excision; but at the same time regular follow-up is essential due to the likelihood of local recurrence. To conclude, scalp lipoblastoma is a rare lesion in clinical practice and a high clinical suspicious is essential for a correct diagnosis and subsequent management. Despite the large tumour size, families can be reassured that tumour is benign in nature and low recurrence rate is expected with complete surgical excision.

Conflict of Interest

No conflict of interest to declare

References

1. Hicks J, Dilley A, Patel D, Barrish J, Zhu SH, Brandt M. Lipoblastoma and lipoblastomatosis in infancy and childhood: histopathologic, ultrastructural, and cytogenetic features. Ultrastruct Pathol 2001;25:321-3.
2. Singh V, Raju R, Singh M, Singh TH. Congenital Lipoblastoma of the Scalp. Amer J Perinatol 2004;21:377-81.
3. Shen LY, Amin SM, Chamlin SL, Mancini AJ. Varied Presentations of Pediatric Lipoblastoma: Case Series and Review of the Literature. Pediatr Dermatol. 2017;34:180-6.
4. Speer AL, Schofield DE, Wang KS, et al. Contemporary management of lipoblastoma. J Pediatr Surg 2008;43:1295-300.
5. Perlis CS, Collins MH, Honig PJ, Low DW. Forehead lipoblastoma mimicking a hemangioma. Pediatrics 2000;105:123-8.
6. Mognato G, Cecchetto G, Carli M, et al. Is surgical treatment of lipoblastoma always necessary? J Pediatr Surg 2000;35:1511-3.
7. Gisselsson D, Hibbard MK, Dal Cin P, et al. PLAG1 alterations in lipoblastoma: involvement in varied mesenchymal cell types and evidence for alternative oncogenic mechanisms. Am J Pathol 2001; 159:955-62.
8. Choi J, Bouron Dal Soglio D, Fortier A, et al. Diagnostic utility of molecular and cytogenetic analysis in lipoblastoma: a study of two cases and review of the literature. Histopathology 2014;64:731-40.