Interdisciplinary Neurosurgery 18 (2019) 100503

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery

journal homepage: www.elsevier.com/locate/inat

Case Reports & Case Series

Paraspinal lipoblastoma with multidirectional spread occurring in a pre-school child

Paseka Seroto (FC Neurosurgery (SA))[a],
Adrian Kelly (MMed Neurosurgery cum laude (SMU), FC Neurosurgery (SA))[a,*],
Tshilidzi Sidiki (MBChB)[a], Aftab Younus (FC Orthopedics (SA))[b]

[a] Department of Neurosurgery, Dr George Mukhari Academic Hospital, Sefako Makgatho Health Sciences University, Pretoria, South Africa
[b] Department of Orthopedics, Helen Joseph Hospital, University of the Witwatersrand, Johannesburg, South Africa

A B S T R A C T

Lipoblastoma is a rare neoplasm arising from embryonic mesodermal cell rests and occurs most commonly in male infants in the first decade of life. While demonstrating the exponential growth of a true neoplasm these lesions do not typically invade surrounding structures but rather present with compressive symptoms. We report a case of a three year old male pre-school child whom presented to our unit with an asymptomatic paraspinal subcutaneous mass on his upper back. Clinical examination revealed no signs of spinal dysraphism and his neurological examination was normal. MRI imaging revealed the typical signal characteristics of a lipomatous mass with a subcutaneous component, extension into and enlargement of the adjacent T5/T6 foramen with thecal impingement, and anterior intrathoracic spread. A gross total resection was achieved through a posterolateral extra-pleural approach. Post-operative histology reported the lesion to be a lipoblastoma. According to our literature review this is the second case of a paraspinal lipoblastoma with demonstrated radiological and intra-operatively confirmed multi-directional spread.

1. Introduction

While lipomas comprise 6% of all soft tissue tumors, lipoblastomas account for only 4.7% of these. These tumors characteristically occur in the first decade of life with a peak incidence between 5 and 6 years of age and demonstrate a male to female ratio of 4:1 [1]. Common sites include the extremities, trunk, head and neck, and mediastinum. Vertebral and spinal cord involvement are rare [2,3]. We present a case report of a three year old male pre-school child whom presented with a T5/T6 lipoblastoma that on imaging demonstrated multi-directional spread.

2. Case description

A three year old male pre-school child presented to our unit, accompanied by his mother, whom complained of a progressively enlarging painless paraspinal subcutaneous mass on his upper back since infancy. He had no family history of spinal dysraphism and had reached developmental milestones within normal parameters.

Clinical examination revealed no stigmata of spinal dysraphism and his neurological examination was normal. The mass was 4 cm by 5 cm in size in a left paraspinal location adjacent to the spinal process of T5/T6. The mass was soft, non-fluctuant, smooth edged, non-tender with no overlying skin changes. Due to the proximity of the mass to the adjacent spinal column thoracic X-rays were requested which revealed enlargement of the T5/T6 foramen. A MRI was subsequently done which revealed the lesion to demonstrate hyper intensity on T1W and T2W imaging. On fat suppression MRI the lesion demonstrated hypo intensity in keeping with lipomatous tissue. The lesion demonstrated multidirectional spread with a subcutaneous component insinuating between the paraspinal muscles, a dumbbell component extending into the T5/T6 intervertebral foramen abutting the thecal sac, as well as anterior intrathoracic extension limited by the parietal pleura [Figs. 1–2].

The patient was taken to surgery where a posterolateral approach was utilized. Upon opening the deep fascia the tumor was immediately visible, yellowish in color, and glistening with fibrous bands [Fig. 3]. The dorsal extra spinal portion was resected and demonstrated minimal attachment to the surrounding tissues. A costotransversectomy was not performed as the tumor had widened the space between the superior and inferior costotransverse joints secondary to chronicity with generous accessibility to the foraminal component. Within the tumor the lesion demonstrated infiltration into the exiting T5 nerve root which was diffusely enlarged necessitating its sacrifice to ensure total

* Corresponding author at: Department of Neurosurgery, 3rd floor Sciences Building; Sefako Makgatho Health Sciences University, Garankuwa, Pretoria, South Africa.
E-mail address: adriankelly1000@yahoo.co.uk (A. Kelly).

https://doi.org/10.1016/j.inat.2019.100503
Received 16 April 2019; Received in revised form 3 June 2019; Accepted 4 June 2019

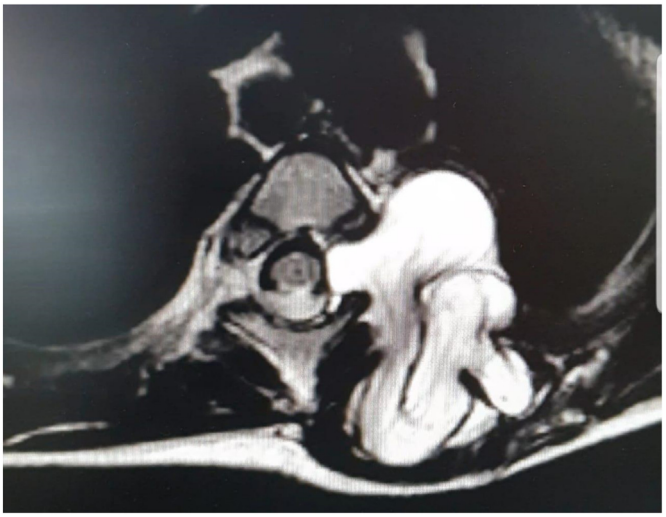

Fig. 1. Pre-operative MRI T2W axial: showing multi-directional spread of lipoblastoma with subcutaneous component, intraspinal component through enlarged T5/T6 foramen demonstrating thecal impingement, and anterior intrathoracic component.

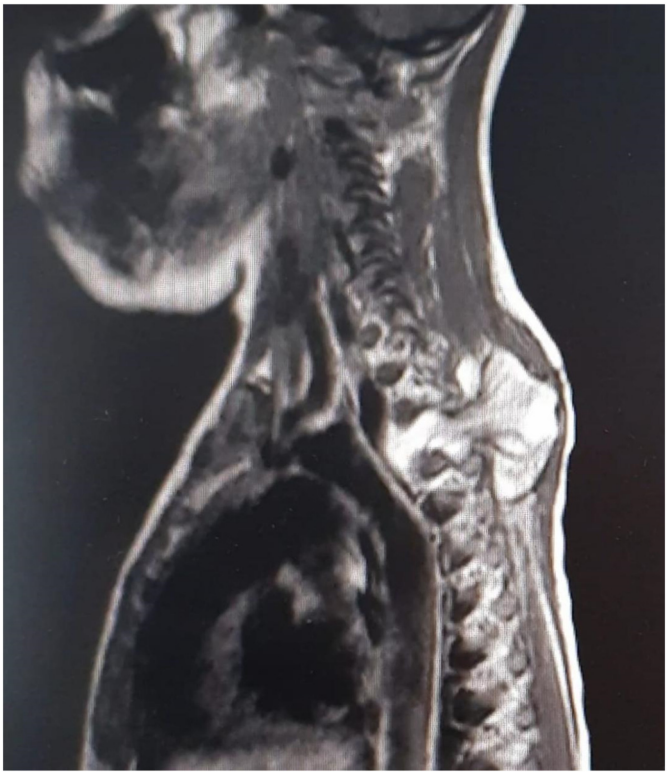

Fig. 2. Pre-operative MRI T2W sagittal: showing localization of intra-spinal component of lesion to T5/T6 level with a more extensive subcutaneous component.

resection [Fig. 4]. By following the lesion into the enlarged foramen the intra spinal component was resected from its abutment with the thecal sac. The intrathoracic component was removed last under direct visualization of the parietal pleura which was pushed forward but not infiltrated. An intra-operative gross total removal was achieved [Fig. 5], which was confirmed on post-operative MRI [Fig. 6].

Histologically the lesion was characterized by lobules of adipocytes. There were scattered myxoid zones that contained bland spindle cells and lipoblasts. The fibrous septa were hypo cellular and contained thick

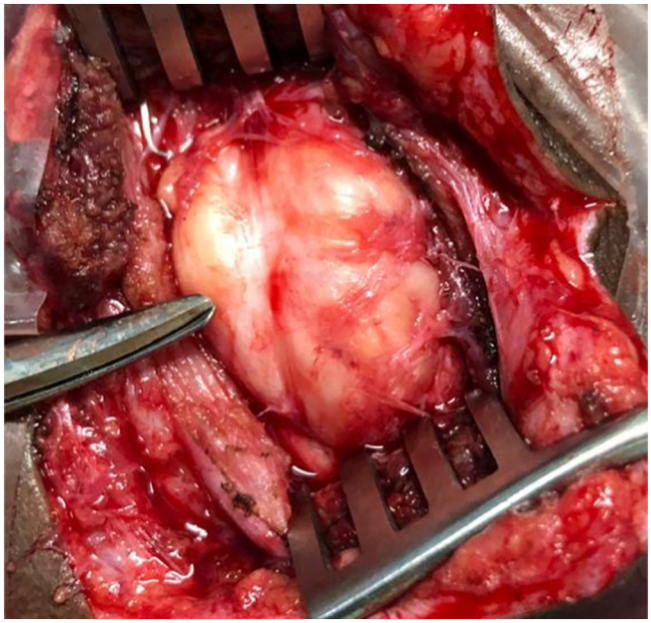

Fig. 3. Intra-operative photograph: subcutaneous component immediately visible upon opening of fascia.

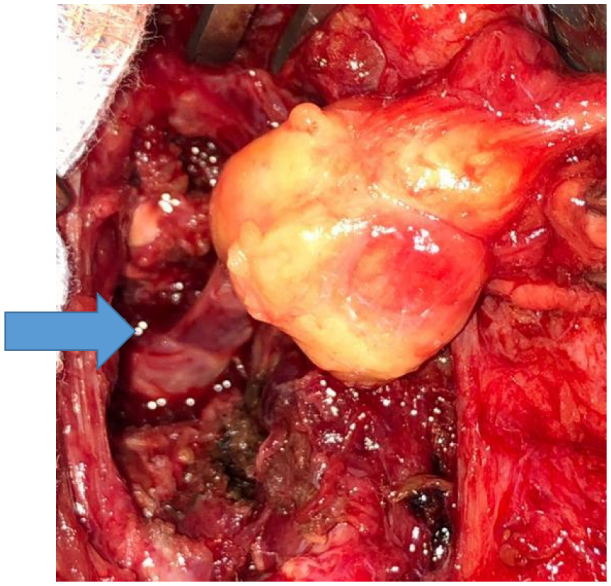

Fig. 4. Intra-operative photograph: The lipoblastoma was able to be mobilized from the enlarged foramen (arrow) and resected by dividing the involved T5 exiting nerve root.

eosinophilic collagen bundles with no necrosis. Histological diagnosis confirmed a lipoblastoma.

Post-operatively the child's wound healed well with normal post-operative neurology. Adjuvant therapy was deferred for recurrence.

3. Discussion

Lipoblastoma's were first described by Jaffe as early as 1926 as a tumor comprised of white fat occurring mostly in the first decade of life with a male to female ratio of 4:1 [1]. The morphologic criteria for the diagnosis of this tumor was however only described in 1973 by Chung and Enzinger who differentiated two subtypes, one being a diffuse lipoblastomatosis and the other being a more well circumscribed lipoblastoma [3]. Radiologically differentiating the diffuse

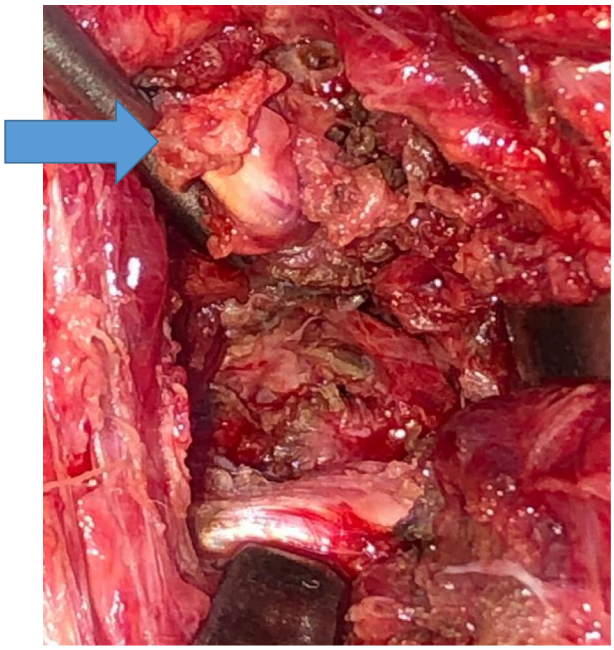

Fig. 5. Intra-operative photograph: showing the extra pleural resection cavity and the transected exiting T5 nerve root (arrow).

Fig. 6. Post-operative T2W axial MRI: showing gross total resection was achieved. The enlarged intervertebral foramen secondary to the chronicity of the lesion is also visible.

lipoblastomatosis from the well circumscribed lipoblastoma relies on MRI imaging where the former demonstrates a relative hypo intensity on T1W imaging and hyper intensity on T2W imaging, while lipoblastoma's demonstrate hyper intensity on T1W and T2W imaging identical to adipose tissue [4]. High cellularity with myxoid infiltration may however result in a lipoblastoma demonstrating hypo intensity on T1W imaging making radiological differentiation difficult [6]. Newer genetic insights have demonstrated lipoblastomas to demonstrate rearrangement in chromosome 8q11-13 [5].

Macroscopically lipoblastomas are lobulated lesions that are yellow in color with a soft consistency, and hence differentiating them from lipomas without histological analysis is difficult. Histologically the lack of abundant vacuolation that characterizes lipomas is used to make the distinction. The differential diagnosis of lipoblastomas does however also include the malignant liposarcomas and the benign tumor of brown fat, the hibernoma. Liposarcomas are differentiated by the prominence of plexiform capillaries and from hibernomas which are comprised of histologically distinct brown fat with a central nucleus [7].

Lipoblastomas that are totally resected have recurrence rates of 13–20% [8]. Incomplete resection on the other hand is strongly associated with recurrence [1]. Lipoblastomatosis has a recurrence rate of 60% even when apparent gross total resection was achieved [9].

While chemotherapy has a defined role in the treatment of lipoblastomastosis its role in lipoblastomas is not defined [10]. The role of adjuvant radiotherapy has similarly not been defined in lipoblastomas and is best avoided in patients below 5 years of age due to long term concerns. As such the optimal management of lipoblastomas is gross total resection and as such this condition remains a largely surgical disease [11,12]. Post-operative surveillance of gross totally resected lipoblastomas is 2-yearly MRI scans with re-operation reserved as the first choice for recurrence [13].

4. Conclusion

According to our literature review this is the first case of a mid-thoracic paraspinal lipoblastoma demonstrating radiological and intra-operative multi-directional spread. Due to a favorable location limited by the surrounding structures a gross total resection was achieved. The child is currently under surveillance in our unit with planned 2-yearly MRI scans.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of Competing Interest

None of the authors listed below have any financial nor personal relationships with other people, or organizations, that could inappropriately influence (bias) their work, all within 3 years of the beginning the work submitted.

References

[1] G. Gupta, R. Garg, C. Wadhwa, H. Bansal, R.K. Kaushal, A rare primary dumbbell lipoblastoma, Asian J. Neurosurg. 13 (2018) 83–85.
[2] N.K. Majethia, Lipoblastoma in Paediatric tumor-report of two xcases, Med. Rep. Case Stud. 1 (2016) 103, https://doi.org/10.4172/2572-5130.1000103.
[3] P. Ghosh, R.N. Das, R. Ghosh, U. Chatterjee, C. Datta, P.K. Mishra, Lipoblastoma and lipoblastomatosis: a clinicopathological study of six cases, J. Cancer Res. Ther. 11 (2015) 1040.
[4] Hussein A.M. Kamel, Paul R. Brennan, Michael A. Farrell, Cervical epidural lipoblastomatosis: changing MR appearance after chemotherapy, AJNR Am. J. Neuroradiol. 20 (1999) 386–389 March.
[5] D.D. Rasalkar, W.C. Chu, Lipoblastomatous tumours, a rare entity: report of two cases in children Hong Kong, J. Radiol. 13 (2010) 209–212.
[6] P. Lopez-Ferrer, J.A. Jimenez-Heffernan, L. Yebenes, B. Vicandi, J.M. Viguer, Fine needle aspiration cytology of lipoblastoma: a report of 2 cases, Diagn. Cytopathol. 32 (2005) 32–34.
[7] J.R. Brodsky, D.Y. Kim, Z. Jiang, Cervical lipoblastoma: case report, review of literature, and genetic analysis, Head Neck 29 (2007) 1055–1060.
[8] G. Grant, M.D. Miller, L. Natalie, M.D. Yanchar, F. James, Lipoblastomas and liposarcoma in children: an analysis of 9 cases and review of the literature, Can. J. Surg. 41 (1998) 455–458.
[9] S.F. Wang, C.Y. Chang, H.D. Wu, Lipoblastomatosis of the shoulder: unusual MR appearance, Br. J. Radiol. 71 (1998) 884–885.
[10] R.J. Stewart, H. Martelli, O. Oberlin, Treatment of children with non-metastatic paratesticular rhabdomyosarcoma: results of the Malignant Mesenchymal Tumors studies (MMT 84 and MMT 89) of the International Society of Pediatric Oncology, J. Clin. Oncol. 21 (2003) 793–798.
[11] A.C. Dubaine, J. Chatten, L. Schut, L. Rorke, Cervical lipoblastomatiosis with intraspinal extension and transformation into fat in a child, Childs Nerv. Syst. 3 (1987) 304–306.
[12] T. Mentzel, E. Claionje, C.D.M. Fletcher, Lipoblastoma and lipoblastomatosis: a clinical pathological study of 14 cases, Histopathology 23 (1993) 527–533.
[13] J. Krishan, V. Hathiramani, M. Hastak, R.G. Redkar, Myxoid lipoblastoma, Indian Pediatr. 50 (2013) 603–605.