

Neck Lipoblastoma. Differential Diagnosis of Cervical Mass in Children

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ARTICLE INFO

Received: 📅 May 02, 2022

Published: 📅 May 11, 2022

Citation: Pablo Damian D'Alessandro, Javier Ruiz, Ramiro Carlos Perea, Camila Marossero, Yvonne Lenz, et al., Neck Lipoblastoma. Differential Diagnosis of Cervical Mass in Children. Biomed J Sci & Tech Res 43(5)-2022. BJSTR. MS.ID.006953.

Keywords: Lipoblastoma; Neck Mass in Children; Case Report

ABSTRACT

Background: When we find a head or neck mass in children we quickly think of a vascular malformation because of its frequency and location. Lipoblastoma is a rare, benign, rapidly growing tumor in infancy and early childhood. This tumor could be presented as a located painless mass in any part of the body, less frequently in head and neck, or as a diffuse and infiltrating behaviour called lipoblastomatosis.

Case Presentation: This series is about three patients all under 24-months-old with fickle, painless and rapidly growing neck mass. All patients had imaging including ultrasound. In only one case, who need a second intervention we did a magnetic resonance (MRI) to establish the extension of the residual mass. All microscopic analysis confirm lipoblastoma. Almost ten years follow-up did not show recurrences.

Conclusion: Lipoblastoma is an unusual soft tissue tumor. Surgical resection without collateral damage is the best option. Microscopic analysis confirm diagnosis. The purpose of this series is to describe an unusual presentation of a head and neck mass in children.

Background

Since 1926, Jaffe used the term lipoblastoma to describe a tumor of immature fat cells located in the groin area [1]. Thirty two years later, Vellios et al. [2] described an "unusual tumor resembling fetal adipose tissue located in anterior wall of the thorax in an 8-month-old-child"; since it differed from hibernoma and other tumors of adult adipose tissue clinically, grossly and microscopically named this as lipoblastomatosis. Chung and Enzinger used benign lipoblastoma for circumscribed types and benign lipoblastomatosis for diffuse multicentric type of these tumors [3]. Children Neck lipoblastoma is an infrequent diagnosis, if we stand beside a toddler with a mass in his neck our first diagnosis is lymphangioma.

The aim of this case series is showing our experience in the last ten years.

Case Presentation Section

Case 1

An Eight-months-old boy presented with a one-month rapidly growing painless, mobile and painless left-sided neck tumor. Ultrasound shows a solid, heterogeneous mass ahead of sternocleidomastoid muscle without contact with cervical large vessels. We decided surgical treatment performing a biopsy of this mass because it had run into the neck without a clear margin to do a

complete resection. In the patient's follow-up we noted a new mass bulge out from the left-side of the neck. Computed tomography (Figure 1) confirmed recurrence. After that, imaging specialists suggest a MRI to establish the extension of the residual mass. A new

surgery was performed with a complete wide resection without collateral damage. Findings were compatible with lipoblastoma (Figure 2). Seven years follow-up with clinical examination and imaging did not show any local recurrence.

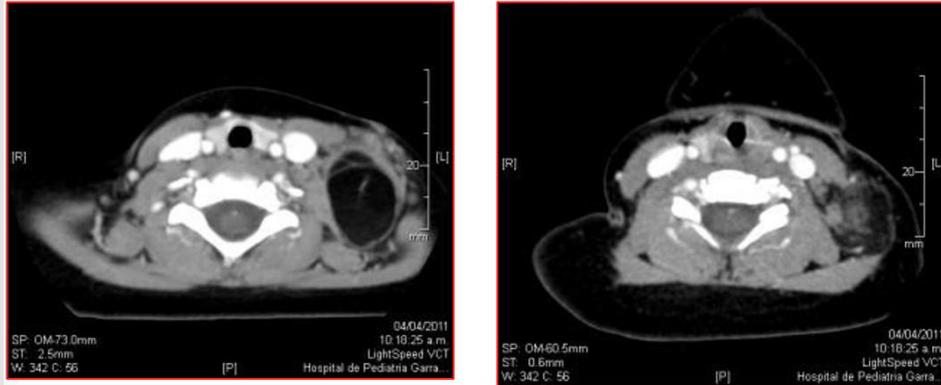


Figure 1: CT Scan showing a big, round, fat-Hounsfield-density, lobulated lesion.

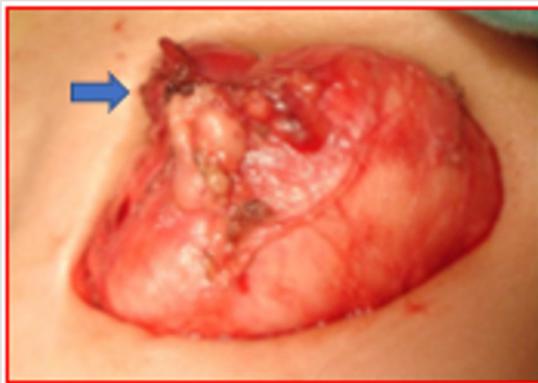


Figure 2: Macroscopic view of a neck lipoblastoma. The tumor is located over the skin in its anatomical position. Look at the root of the mass (blue arrow) that have origin in the neck and expands to the supraclavicular region and the upper chest.



Figure 3: Chest X-Ray of a toddler with a giant cervical mass (red arrow). The tumor doesn't involve bones only soft tissue without invasion of the thorax.

Case 2

A 20-months-old boy presented with an eighth-month progressive growing left-sided neck tumor (Figure 3). Ultrasound showed a solid, heterogeneous 4 inch longitudinal diameter mass in left supraclavicular space. Complete resection with clear margins was performed. Follow-up did not show any recurrence.

Case 3

A 14-months-old boy presented with an only-month rapidly growing left-sided neck tumor. Ultrasound (Figure 4) showed a superficial soft tissue mass with images features similar to lipoma. Complete resection was performed and pathology named it as a lipoblastoma. Margins were uncompromised and he did not show any recurrence in follow-up.



Figure 4.

Discussion

From 2011 to 2021, we described three cases, all younger than two years old, of located soft tissue mass in neck topography. All patients were male consisting with the male preponderance (3:1) reported in literature [4]. All patients have had markers in order to rule out the diagnosis of another cervical tumor like neuroblastoma or teratoma such as Alpha- fetoprotein, human chorionic gonadotrophin and urine catecholamines. This tumor is presented as a lateral cervical rapidly growing, soft mass in all cases. Ultrasound and Computer Tomography shows features similar to lipoma. Imaging is not helpful in diagnosis because it is very difficult to establish the difference between malignant and benign lesions. Preoperative imaging should be used to assess extent of disease and surgical planning [5]. In this series we do not think that was useful complete the patients study with a CT or MRI previous the surgery because the pathognomonic imaging in US. In case of recurrence we made more sensitive and specific imaging studies in order to know the extent of the disease and compromise of other structures to improve the surgical result trying to be the patient last surgery [6]. One patient of the series had a local recurrence successfully treated with a second surgery reaching complete resection of the tumor. The goal is always to perform a complete resection with negative margins, but this can be difficult to obtain with lipoblastomatosis. When negative margins are unobtainable, close clinical follow-up with examination and imaging studies is advised. Regardless of margin status, postoperative follow-up in light to document recurrence rate should be done. In our series, we have a seven-year follow up period according to the five-years follow up recommendation [4]. Patients with lipoblastoma have an excellent prognosis despite off the tumor potential for local invasion and rapidly grows up. Metastases have never been reported.

The recurrence rate is 14%, it is more frequent in lipoblastomatosis. There are slender descriptions about children's head and neck lipoblastoma. In 2006, Sinha BK [7] reported 100 cases in literature with only 10 patients involving cervical region [7]. Other reports described similar numbers ranging between 11 to 30%. In imaging lipoblastoma cannot be distinguished from myxoid liposarcoma but thinking on patients age, liposarcoma is extraordinarily rare in children younger than 10 years old. Fat-containing lesions of the head and neck are commonly encountered in daily practice. Lipomas, liposarcomas, lipoblastomas, dermoids or teratomas are different entities with similar behavior in imaging methods [8]. Cross sectional imaging using MRI or CT are the mainstay, they provide exquisite detail in head and neck pathologies diagnosis quest (Table 1) Identifying, localizing

and characterizing the tumors, imaging helps to establish the extent of local involvement, bone invasion, nodal disease and distant metastasis. Imaging provides critical information for biopsy, surgical resection, chemotherapy or radiation. It plays an important role in the assessment of treatment response and disease recurrence [9]. Microscopically differentiating between this two entities propose a challenge and as a result of that, genetic analysis is necessary [10]: Lipoblastomas may have break-points in the long arm of chromosome 8 (8q 11-13) and liposarcomas typically show either the translocation t(12;16)(q13;p11) with the FUS/TLS-CHOP chimeric fusion protein or the translocation t(12;22)(q13;q12) with the EWS-CHOP chimeric fusion protein [11]. No atypical lesions were described [12].

Table 1: Imaging differential diagnosis of head and neck masses.

LIPOMA	LIPOSARCOMA	LIPOBLASTOMA
Well encapsulated mass with internal septations. -50 to -140 Hounsfield units (HU) on CT and a High signal on MRI	Heterogeneous fat-containing masses, septations, calcification and enhancing nodules	Well circumscribed fatty lesions with septations

Conclusion

When we talk about head and neck tumors in children, lipoblastoma is a rare condition that usually confused with lipoma in imaging. We cannot reach a differential diagnosis of a cervical mass showing a rapidly painless growing behaviour within imaging. Therefore, surgical treatment is the best option after ruling out other malignant conditions. Imaging and clinical follow-up is also requested despite off benign condition and the feasibility of the surgical resection.

Declarations

Ethics Approval and Consent to Participate

Not Applicable.

Consent for Publication

Written informed consent were signed by parents of the study participants

Availability of Data and Material

Not Applicable.

Competing Interests

The authors declare that they have no competing interests.

Funding

Not Applicable.

Authors' Contributions

- DPD: Corresponding Author (conception, desing, acquisition and drafted)
- RJ, PRC, LY, QH and BM take part in surgeries and following of the patients (acquisiton in case series)
- FPS contributed to work desing and analysis in discussion.
- OA is on a fellowship in imaging and interventional radiology. He contributed to the manuscript writting differential diagnosis of head and neck fat-containing masses.
- MC revised the manuscript until sended.
- All Authors read and approved the final manuscript.

Acknowledgement

Special thanks to Prof. Mónica Laporte who provided professional services making possible the correct writing of this case series.

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ISSN: 2574-1241

DOI: 10.26717/BJSTR.2022.43.006953

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