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ENTCase

CERVICAL LIPOBLASTOMA: SURGICAL INTERVENTION AND CLINICAL OUTCOMES IN THE MANAGEMENT OF A RARE ADIPOCYTIC TUMOR IN INFANCY AND EARLY CHILDHOOD

SERVİKAL LİPOBLASTOMA: BEBEKLİK VE ERKEN ÇOCUKLUK DÖNEMİNDE GÖRÜLEN NADİR BİR ADİPOZ TÜMÖRÜN TEDAVİSİNDE CERRAHİ MÜDAHALE VE KLİNİK SONUÇLAR

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Baş Boyun Cerrahisi

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Özet

Lipoblastoma (LB), çoğunlukla bebeklik ve çocukluk döneminde ortaya çıkan, embriyonik beyaz yağ hücrelerinden kaynaklanan, nadir görülen iyi huylu bir mezenkimal tümördür. LB genellikle üç yaş altı çocuklarda görülür. Sıklıkla gövde ve ekstremitelerde yerleşim gösterse de vücudun herhangi bir bölgesinde gelişebilmektedir. Baş ve boyun bölgesinde son derece nadir rastlanan bir kitledir. Hikaye ve fizik muayene ayırıcı tanıda oldukça önem arz etmekle birlikte boyun kitlelerinin olası nedenlerinin ve özelliklerinin belirlenmesinde görüntüleme çok önemlidir. Boyun bölgesine özgü cerrahi zorlukların ve cerrahi anatominin bilinmesi, hayati anatomik oluşumların bu bölgede yer alması nedeniyle zorunluluk arz etmektedir. Bu olguda servikal lipoblastom tanısı alan 18 aylık bir çocuğun sunumu, tanısı ve tedavisi tartışılacaktır.

Anahtar kelimeler: Lipoblastom, boyun benign neoplazm infant

Abstract

Lipoblastoma (LB) is an uncommon benign mesenchymal tumour that predominantly manifests during infancy and childhood. It is commonly seen in children under the age of three and originates from embryonic white fat cells. Although it has the potential to manifest in any part of the body, is frequently observed in the thoracoabdominal region and extremities. These lesions are exceedingly uncommon in the cervical region. The history and physical examination can narrow the potential diagnoses for the cervical masses and imaging is important for identifying the probable cause and characteristics of the mass. Knowing the surgical difficulties spesific to the head and neck region is crucial because of the anatomic relationships. In this case, the presentation, diagnosis and the treatment of an 18 months child with cervical lipoblastoma will be discussed.

Keywords: Lipoblastoma, neck benign neoplasm infant

Introduction

Lipoblastoma (LB) is an uncommon benign mesenchymal tumour that predominantly manifests in infancy and childhood [1], typically observed in children under the age of three [2]. It originates from embryonic white fat cells. [1]. These benign tumours exhibit rapid expansion and their symptoms vary based on their specific location. They have the potential to cause deformities or exert pressure on nearby structures. [3, 4]. It has the potential to manifest in any part of the body. However, it is frequently observed in the trunk and extremities. LB is extremely uncommon in the cervical region [2]. It can be categorised into two discrete types: circumscribed LB (CLB), which is enclosed within a capsule, and diffuse lipoblastomatosis (DLB). DLB is a lesion that is deeply placed and does not have clear boundaries and its growth pattern is infiltrative that could potentially affect nearby muscle components. [5, 6]. LB in the neck manifests as a fast growing mass without pain. However, in certain cases, it may exert pressure on nearby structures, leading to breathing difficulties, Horner syndrome, and hemiparesis. [7, 8]. In this case, the clinical presentation, imaging, pathological examination findings, and treatment choices of a

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LB that developed in the cervical region of a 1.5-year-old child will be discussed.

Case Report

An 18-month-old female applied to ENT-outpatient clinic with a six-month history of a mass on the right cervical region, which had grown by the time (Figure 1.).

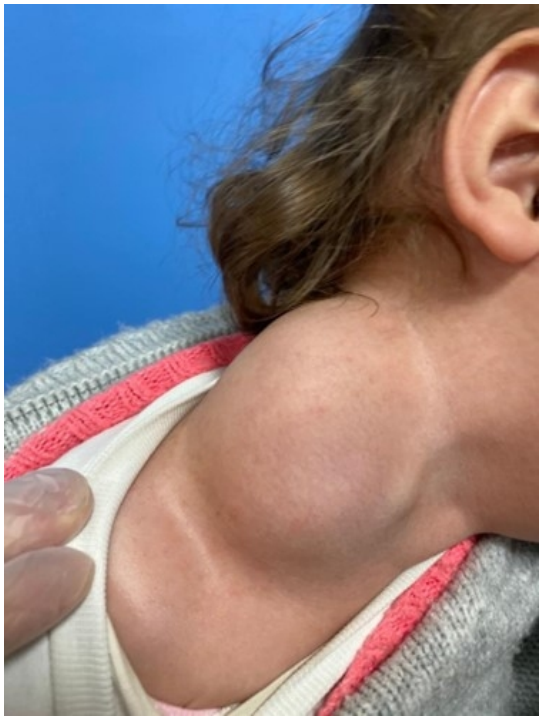

Figure 1

The preoperative apperance of the right neck mass

Her neck and extremity movements were not restricted by the mass. The physical examination revealed an approximately 9 cm diameter cervical mass on the right cervical region, located in the supraclavicular and posterior cervical triangles. The mass was solid, not painful, mobile in relation to the skin and did not demonstrate any signs of inflammation. No lymph node enlargement was detected in the neck region. Her brachial plexus and cranial nerve examinations were normal. Her medical records showed no indication of any physical injury, infection, contact with animals that transmit diseases, or tuberculosis. The patient did not exhibit any symptoms of losing weight and breathing problems. She was otherwise healthy, as indicated by normal findings from routine total blood count and blood biochemical parameters. Neck ultrasonography revealed a 48x50 mm encapsulated solid tumour in the right neck. A preoperative MRI revealed a 43x50x70 mm (Anteroposterior x Transverse x Coronal) tumour with a hyperintense signal in T1 imaging. It was found beneath the deep cervical fascia - superficial layer, between the Sternocleidomastoid and the trapezius muscle, extending to the superior axilla - the superior-posterior neighbourhood of the pectoral muscles in the inferior and to the submandibular gland level in the superior. The septae are contrasted. The mass was in fat-intensity (Figure 2a, 2b, 2c).

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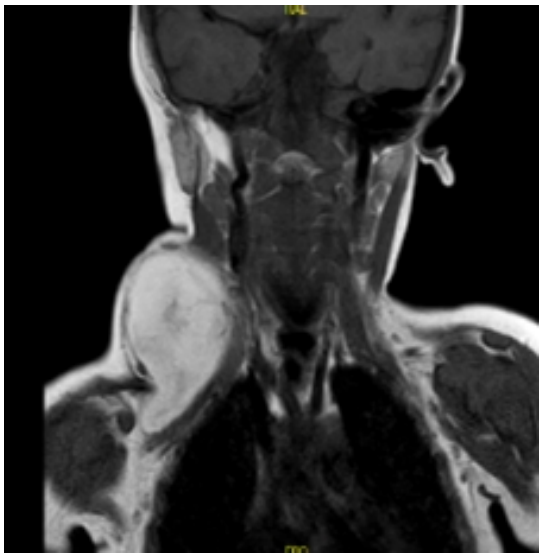

Figure 2A
The coronal MRI section

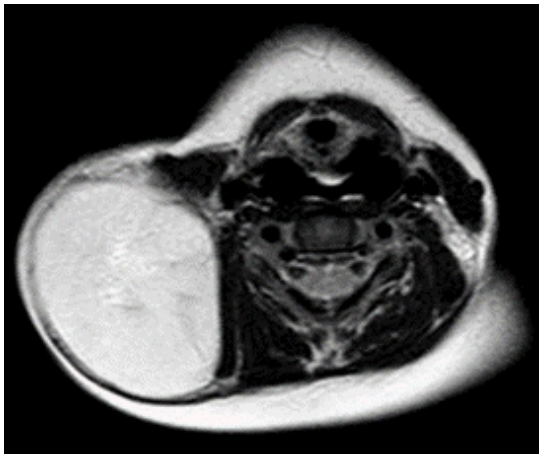

Figure 2B
The axial MRI sections

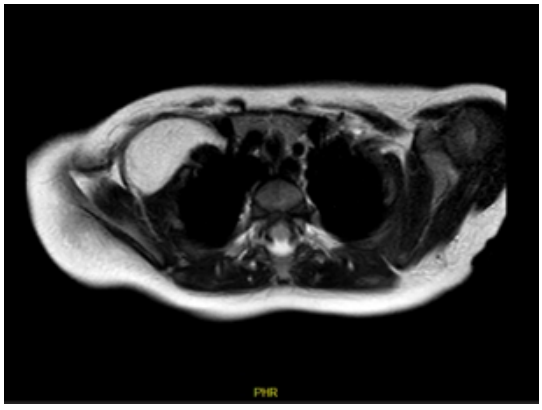

Figure 2C
The axial MRI sections

She had already underwent a fine needle aspiration biopsy at another clinic. The diagnosis was confirmed by referring it to the pathology department. The histopathological examination revealed an immature lipomatous lesion with no obvious signs of malignancy, indicating lipoblastoma. Due to the relationship of the mass with the

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brachial plexus and upper extremity, surgery was planned in collaboration with the orthopedics and traumatology department.

The patient was given general anaesthesia, and after minimal neck extension, a transverse incision was performed (Figure 3, 4).

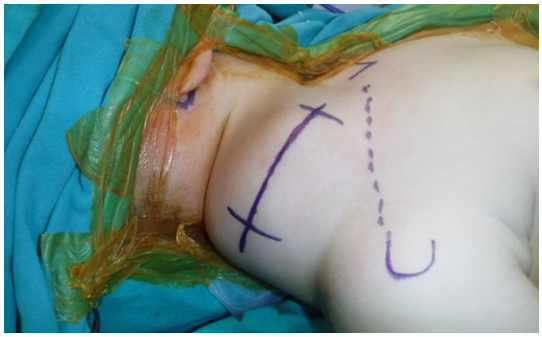

Figure 3
The incision, planned

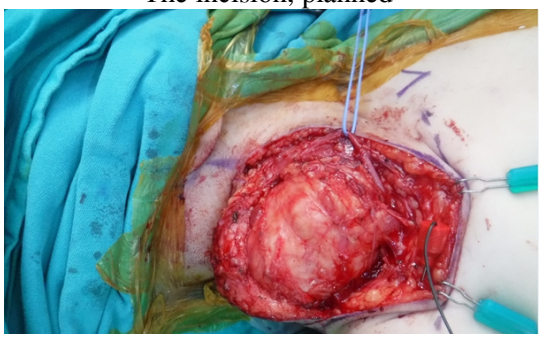

Figure 4
The intraoperative apperance of the tumour

The dissection route was the subplatysmal plane, preserving the all the surrounding structures including sternocleidomastoid muscle, internal jugular vein, brachial plexus and accessory nerve. It appeared to be enclosed, which made it easier to remove. Meticulous dissection was carried out and no adjacent neck structures were invaded. The tumour moved from the posterior triangle's floor (Figure 5, 6).

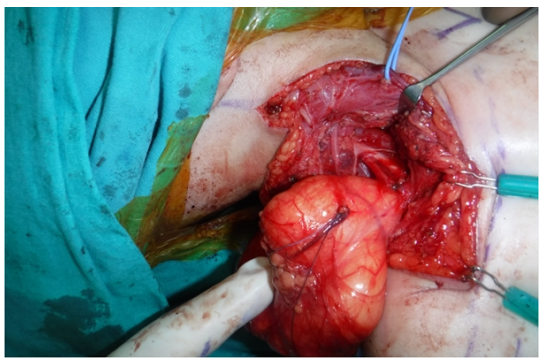

Figure 5
The relationship of the tumour with the brachial plexus

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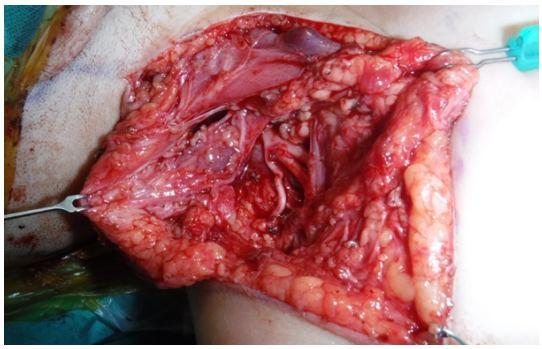

Figure 6

The surgical area, after the tumour removal, the brachial plexus is intact

A complete surgical excision revealed 9.5x5.5x4.6 cm yellowish, encapsulated adipose tissue (Figure 7).

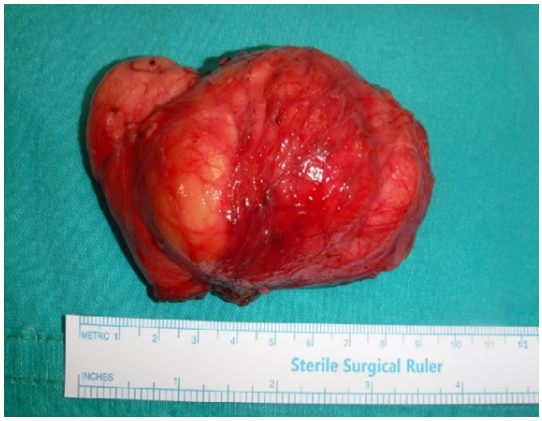

Figure 7

The tumour, removed

The mass had an uniform appearance. The final histopathological examination confirmed the diagnosis of LB.

Discussion

Lipoblastoma is an uncommon, noncancerous tumour that arises from embryonic adipose cells. Ninety percent of LB occurs under the age of three years. [9, 10]. It can be categorised into two discrete types: CLB, which is enclosed within a capsule, and diffuse lipoblastomatosis. Diffuse lipoblastomatosis is a lesion that is deeply placed and does not have clear boundaries. DLB's growth pattern is infiltrative that could potentially affect nearby muscle components [5, 6, 11]. Although they have been in the head and neck, shoulder, inguinal region, axilla, back, and abdominal cavity, these tumours typically appear in the extremities [12, 13]. A rapid growing, painless neck mass is the typical presentation of cervical lipoblastoma; but, on occasion, it can compress nearby structures and cause hemiparesis, Horner syndrome, and respiratory issues. [7, 8]. The differential diagnosis for the neck masses can be narrowed with the help of the history and examination. Additionally, imaging is crucial for identifying the features and most likely aetiology of neck masses [14]. The imaging technique that can identify these lesions the best is magnetic resonance imaging (MRI) [15]. According to a comprehensive analysis, MRI was the most commonly used method for evaluating LB, accounting for 49% of cases. This was followed by computed tomography (CT) at 36%, ultrasound (US) at 28%, X-ray at 8%, and airway endoscopy at 2%. In 11 cases (21%), preoperative biopsy was carried out in this systematic study [16]. Myxoid liposarcoma is the differential diagnosis that matters the most. Imaging does not differentiate lipoblastoma from myxoid liposarcoma. But the patient's age is crucial. Liposarcomas are incredibly uncommon in those under the age of ten. [5]. Therefore, in the case of a young child (under 2 years old), if an imaging scan shows a lesion that contains fat, it is most likely to be a lipoblastoma [17, 18].

When possible, doing a complete surgical removal is curative for these lesions [15]. Understanding the anatomical links is essential for recognising the specific surgical challenges associated with the head and neck region [15]. One case was the misidentification of a lipoblastoma as a neurofibroma, leading to the removal of the spinal accessory nerve [19]. The tumor's relationship with the brachial plexus, adjacent vessels like subclavian vessels, and mediastinum in multiple cases caused significant difficulties and challenges during the surgical removal. Sun and Hilsinger (2003) documented a case which is a lipoblastoma in the neck in close proximity to the spinal cord. In this situation, subtotal resection was chosen to avoid any potential damage to the spinal cord [20]. In the presented case, there was no injury to the adjacent anatomical structures. Postoperative evaluations indicated that the neurological functions of the right upper extremity, where the surgery was performed, and the functions of the cranial nerves were preserved.

There have been no reports of metastases or malignant transformation. LB is a well-defined and localised lesion, often measuring less than 6 cm in diameter. However, there have been instances of larger lipoblastomas have been documented as in the presented case [12, 21].

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