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CASE REPORT

Lipomatosis of the neck: Case report and literature review☆

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Summary Lipomas are common benign tumors of the head and neck that are discrete and encapsulated. Lipomatosis, however, is a more diffuse fatty tissue growth rarely found in the head and neck. A 2.5-year-old female presented with a painless, right supraclavicular mass. Radiologic evaluation revealed extension into the anterior mediastinum. Incisional biopsy and histology were consistent with lipomatosis. The lesion was not excised and at 2 years follow-up the child remains asymptomatic with no enlargement on imaging. Conservative nonoperative management should be considered with asymptomatic disease in which the morbidity and mortality of excision may be high.

1. Introduction

Lipomas are benign tumors of mature fat cell origins and represent the most common mesenchymal lesion in adults. Approximately 13% of lipomas occur in the head and neck region and have an overall 5% recurrence rate [1]. They are usually discrete and often encapsulated. Lipomatosis, however, is an excessive, poorly circumscribed proliferation of mature adipose tissue [2]. Histologically, lipomatosis is indistinguishable from lipoma except for the lack of circumscription [3].

Lipomatosis is typically either neoplastic or hamartomatous in nature. It is often one component in a constellation of developmental or structural abnormalities. Diagnosis can be challenging since many of these disease entities have overlapping features. The differential diagnosis of lipomatosis involving the head and neck in children and adolescents includes steroid-induced mediastinal lipomatosis, congenital infiltrating lipomatosis of the face, lipoblastomatosis, diffuse lipomatosis, Bannayan-Zonana syndrome, and encephalocraniocutaneous lipomatosis [3].

Lipomas and lipomatosis with mediastinal involvement were reviewed by Nguyen et al. [4]. We report only the second case of isolated, idiopathic

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mediastinal lipomatosis presenting as a neck mass in the pediatric population.

2. Case report

A 2.5-year-old female presented with a painless right supraclavicular mass of less than 6 months duration. No associated symptoms of airway compromise or swallowing difficulty were reported. Past medical history was remarkable for asthma and recurrent otitis media. Birth history was unremarkable. On physical exam, the neck mass was 2 cm × 3 cm in size, soft to palpation, and transilluminated upon examination with a light source. The remainder of the head and neck examination was normal. Computerized tomography (CT) revealed a mass of fat density extending from the level of the cricoid cartilage inferiorly into the mediastinum adjacent to the trachea and esophagus, without enveloping these structures (see Fig. 1). There was no evidence of invasion or bony destruction.

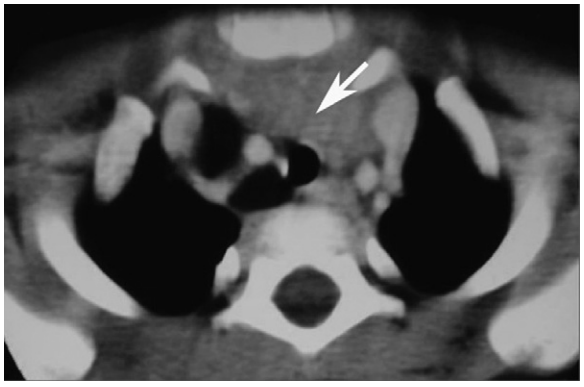

Fig. 1 CT scan of mediastinum showing mass (arrow) adjacent to trachea and esophagus without enveloping them.

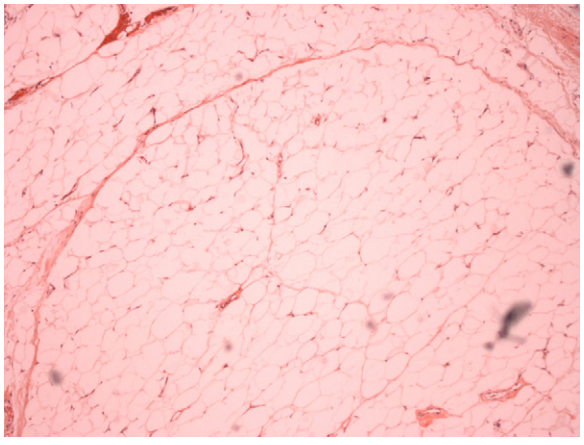

Fig. 2 Low magnification view of adipocytes with focally increased fibroconnective tissue.

Incisional biopsy was taken in the operating room using a transverse incision anterior to the sternocleidomastoid muscle border. A fatty mass was noted without evidence of fluid or unusual vascularity. The biopsy was taken and the wound was closed in layers. Histologic evaluation was consistent with lipomatosis (see Fig. 2).

Given the mediastinal extent of the tumor, surgical resection would have included thoracotomy or sternotomy with associated morbidity. For this reason, nonoperative management was recommended and agreed upon with the family. After 2 years, the child remained free of symptoms, and yearly imaging showed no enlargement of the mass.

3. Discussion

A review of the English literature for lipomatosis returned a surprisingly diverse subset of rare diseases. Multiple symmetrical lipomatosis (or Madelung's disease) was first described by Brodie in 1846 and is perhaps one of the more well-known causes of lipomatosis of the neck. This disorder is characterized by diffuse symmetric deposition of fat in the neck, face, shoulder, upper extremities, and trunk [5].

Lipomatosis secondary to extended corticosteroid use or Cushing's syndrome is also commonly reported. These lesions can manifest in the mediastinum, mimicking cardiomegaly on imaging with widening of the mediastinal silhouette [6]. This lesion can be seen in a child taking long-term steroids for asthma or graft versus host disease after bone marrow transplant [3].

Congenital infiltrating lipomatosis of the face is a rare disorder that presents at birth on the cheek. It has ill-defined borders and infiltrates muscle and soft tissue, making excision difficult and recurrence likely. There may be hypertrophy of underlying bone. Definitive excision and reconstruction are delayed until adolescence to allow regression of the buccal fat pad, minimize the risk of damage to the facial nerve, and to have a mature contralateral cheek contour to match [2].

Lipoblastomas are benign tumors of embryonic adipose tissue that occur in infancy and early childhood, with 90% occurring before 3 years of age [3]. They represent 2% of all pediatric soft tissue tumors [7]. While typically presenting as a rapidly growing soft tissue mass involving the extremities, these lesions have been reported in the head, neck, and mediastinum [3]. Lipoblastomatosis, as expected, can be defined as a locally invasive lipoblastoma. Sun et al. [7] reported that only 10 cases of lipoblastomatous tumors have been reported.

Several other rare pediatric disorders of the head and neck are characterized by lipomatosis. Diffuse lipomatosis presents as a rapidly enlarging mass of mature fat on the trunk or extremity, often associated with tuberous sclerosis, hyperostosis, and gigantism. Bannayan-Zonana syndrome features lipomatosis, lymphangioma, and hemangiomas of the trunk and extremities. Macroencephaly and other malformations can also be present. Finally, encephalocraniocutaneous lipomatosis involves epilepsy, mental retardation, and cerebral malformations in addition to lipomatosis of the scalp and neck [3].

The diagnosis of lipomatosis is made possible after a thorough history, physical exam, and routine imaging. A history of congenital malformations, extended steroid use, alcohol abuse, Cushing's syndrome, or other endocrinologic abnormalities are important in elucidating a specific cause of the lipomatosis. On physical exam, lipomatous tissue is mobile and soft. As in our patient, the lesion typically transilluminates on examination with a light source.

On computed tomography (CT), mature adipose tissue has low attenuation. Additionally, lipomatous tissue shows high intensity in both T1-weighted and T2-weighted magnetic resonance imaging (MRI) [8]. However, only biopsy and histopathologic examination can definitively differentiate lipomatosis from entities such as lipoblastomatosis and liposarcoma. The absence of lobule formation and embryonic fat rules out lipoblastomatosis. Unlike lipomas, liposarcomas have lipoblastic proliferation, pleomorphism, and mitosis on histologic examination [2].

Prognosis and treatment approach vary from case to case. For example, an infiltrating type of lipomatosis near a major vessel or the spinal cord will have a worse prognosis and surgical excision is more difficult. In our case, imaging revealed a relatively large lesion extending into the mediastinum. Despite the extensive nature of the lipomatosis, the patient was asymptomatic without evidence of airway obstruction or compression of vital structures, and histopathology ruled out malignancy. The decision was made to manage the lesion conservatively. An aggressive surgical approach could have subjected the young child to a thoracotomy or sternotomy and its associated morbidity and mortality. We felt that routine follow-up with imaging was warranted to monitor the extent and growth of the lesion.

The question remains as to what caused this instance of lipomatosis in a 2.5-year-old child with a past history of asthma and recurrent otitis media. Pungavkar et al. reported the only case of isolated mediastinal lipomatosis in a patient with no known risk factors [9]. Similarly, in our case, there is no history of congenital malformations or lipomatous abnormalities in the family. Prolonged steroid use, alcohol abuse, or Cushing's syndrome was not elicited in the history. We are reporting only the second case of isolated, idiopathic mediastinal *lipomatosis* in the pediatric population.

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