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Case Report

Lipoblastoma nose: a rare case report

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ABSTRACT

Lipoblastoma is a rare exclusively benign encapsulated tumor arising from embryonic white fat. Tumor occurs primarily in infancy and early childhood. It often presents as painless swelling in extremities and trunk, and rarely develops in head and neck and other sites. We report case of a 15 months old boy with a gradually enlarging, painless swelling over the left side of the dorsum of nose. Computed tomography scan revealed a subdermal soft tissue mass. Treatment remains complete surgical excision. Diagnosis is determined by histopathologic findings. Histologically, lipoblastoma reveals immature adipose cells in varying stages of maturity arranged into lobules separated by septa. No recurrence of tumor has been noted over 6 months of follow-up of this patient.

Keywords: Lipoblastoma, Benign tumor, Pediatric, External nasal mass

INTRODUCTION

Lipoblastoma is a rare, benign, well circumscribed tumor arising from embryonic adipose tissue and is characterized by a 3:1 male predominance. The tumor occurs primarily in children younger than 3 years old.[1]

Most of them present as superficial, circumscribed, slow growing mass in limbs.[2] It most commonly occurs in extremities (30-70%) and trunk (20-50%), and rarely develop in head and neck, mediastinum and retro peritoneum. Even rarer locations include cardiac sites and the labia[3]. It typically presents as rapidly enlarging, painless mass. Symptoms can be just a swelling or can occur from compression of adjacent structures. Diagnosis is suggested by CT and MRI findings, but can only be confirmed by microscopic examination.

This exclusively benign tumor consists of fat lobules of variable maturity, multivacuolated lipoblasts, fibro capillary network and myxoid stroma.[4] We hereby report an exceptional case of lipoblastoma of nose, and to the best of our knowledge this is the first case of lipoblastoma of nose reported in literature.

CASE REPORT

A 15-months old male was brought to ENT OPD with a swelling over the dorsum of nose on left side that had been there for four months. It gradually increased in size to the present size. No other complaints like pain, tenderness, nasal blockage or watering of eyes were noticed by his parents. He had no discomfort or any associated symptoms like irritability, fever or weight loss. There was no history of trauma or nose piercing. Physical examination was normal except for a swelling of size 1x1 cm, which was nodular and mobile, skin overlying was normal, it was not increasing on crying, non-tender, non-transilluminating, Furstenberg test was negative. Computed tomography scan was done which demonstrated limits of the mass and was showing a well-defined soft tissue density lesion (20-35 HU) in subdermal aspect of left nostril with no luminal narrowing. Ultrasound would have helped to outline the mass but wouldn't have supplied any additional information. All laboratory investigations were normal. Complete surgical resection done under general anesthesia through an incision along the relaxed skin tension line (RSTL), as shown in Figure 1. At the

operation 1×1×1 cm well circumscribed, yellowish white mass was excised with a combination of sharp dissection and electrocautery (Figure 2).

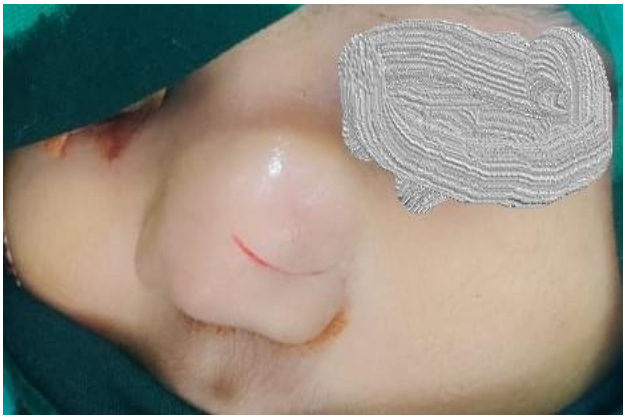

Figure 1: Curvilinear incision along RSTL.

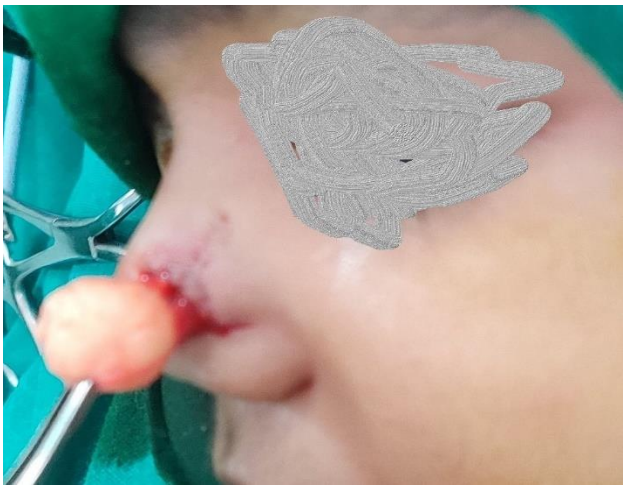

Figure 2: Tumor before cutting final attachments.

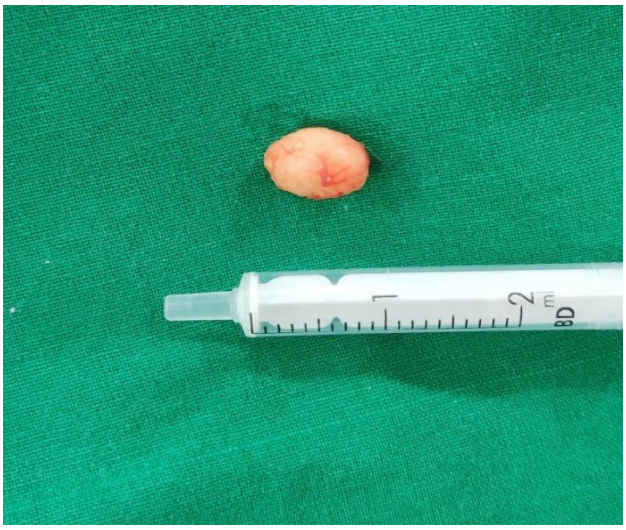

Figure 3: Final specimen sent for histopathological examination.

Histopathological examination

Macroscopically shows yellowish white mass with glistening surface.

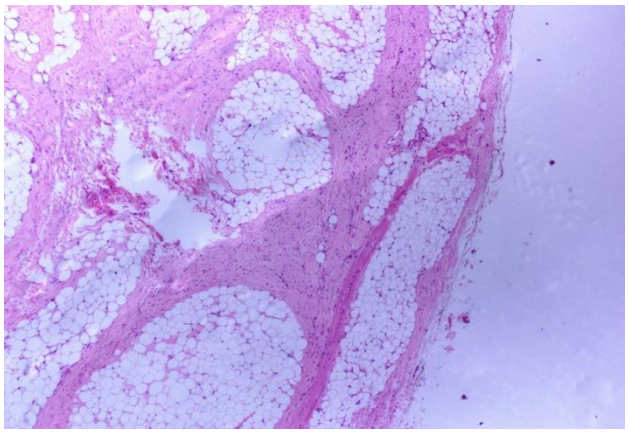

Figure 4: Low power microscopy suggestive of both mature and immature adipocytes.

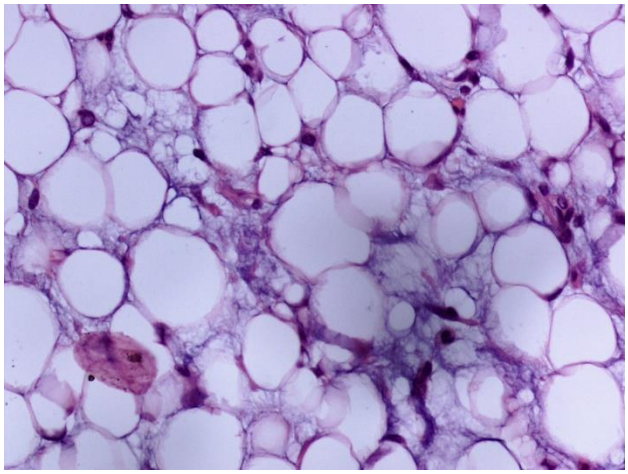

Figure 5: High power microscopy (200X) large vacuolated adipocytes with stellate cells.

DISCUSSION

Lipoblastoma, benign tumor of embryonic adipose tissue, affecting principally males before the age of 3 years, although cases up to age 7 or 8 years are encountered rarely. Age ranges from 5days to 18 years and more than 80% cases are below 3 years of age. In previous reports, lipoblastoma have been noted to have a predilection for sites that possess the most primitive adipose tissue in the newborn; axilla, neck, chest wall, however 70% occurred in extremities.

Here, we present a case of lipoblastoma of nose, evaluated clinically, investigated and completely excised. Histopathological examination disclosed the final diagnosis.

From all pediatric soft tissue tumors, <10% are adipose tumors of which 5-30% are lipoblastomatous type. This tumor presents in two forms, localized well circumscribed lesion (lipoblastoma) or multicentric type (lipoblastomatosis).[4]

Axial, Sagittal and coronal views of computed tomography scan are helpful to determine accurately the size and extent of the lesion as well as the integrity of adjacent bony structures. CT scan shows it as a fat dense mass containing enhanced septa. Magnetic resonance imaging can suggest histology of some components i.e., can help identifying the lipomatous nature of the mass, but the findings can be inconsistent due to variable maturity of fat cells and the mesenchymal content of the tumor. But these findings are not diagnostic. So, the definitive procedure to diagnose is excision biopsy and histopathological examination. Principle histologic clues for diagnosis of lipoblastoma are striking lobulation, maturation towards the center of the lobules and absence of nuclear atypical or atypical mitosis.

Cytogenetic analysis has shown similar chromosomal abnormalities in adipose tissue tumors, lipoblastoma more consistently shows 8q11-q13 abnormalities. Lipoma shows translocation in chromosome 12, mucoid liposarcoma shows t (12; 16) (q13; p11), hibernoma shows rearrangement in chromosome 11q13.

The differential diagnosis of lipoblastoma must rule out myxolipoma, myxoid liposarcoma, hibernoma, infantile fibromatosis and lipofibromatosis.[5] Some authors had considered lipoblastoma as a tumor of intermediate malignancy and postulated that it may progress to liposarcoma, although there has been no evidence to support this viewpoint. Treatment of choice remains to be Complete surgical excision. Recurrence has been reported in 14-25% of cases, have been attributable to incomplete removal. Recurrent lesions quite often show maturation towards a simple lipoma but no metastasis or malignant transformation has been seen, which signifies a favorable prognosis.

CONCLUSION

In day-to-day clinical practice evaluation of a nasal mass is a routine, a thorough examination and pathological diagnosis plays a key-role in clinching the diagnosis and effective management. Henceforth owing to the rarity of presentation the case has been documented.

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