

PARA PHARYNGEAL SPACE LIPOMA MIMICKING CYSTIC HYGROMA

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ABSTRACT

Para pharyngeal space tumors constitutes only 0.5-0.8 % of all tumors of which Lipoma is the rarest. Generally, lipoma is found just beneath the subcutaneous tissues. Here we are presenting a case of 9 year old child with lipoma of left parapharyngeal space extending into the neck mimicking cystic Hygroma on presentation.

Key Words: Parapharyngeal space tumors, Lipomas, Tumors in children, Soft tissue tumor.

INTRODUCTION

Lipoma is a benign neoplasm of adipose cells and constitutes 16% of all the soft tissue neoplasms.¹ It is mesenchymal in origin and rarely found in deep areas. It's presence in head and neck is rare and contribute to 0.5% of all Para pharyngeal space (PPS) tumors.²

Only 10 cases have been reported so far of lipoma in PPS.³⁻⁸ In a study Singh et al. reported female preponderance in Asian subcontinent and with highest occurrence in 20-40 years of age and with nil cases with less than 10 years of age.⁹

Lipomas are found over vast range of age but most commonly affect adults in 40-60 years of age.^{10,11} When found in children usually part of Bannayan Zonnana Syndrome (autosomal dominant trait although few sporadic cases) diagnosed on basis of¹²

- Macrocephaly
- Multiple lipomas
- Hemangiomas

Lipomas affect 1% of general population, with male and female in equal ratio.¹³ They may be in multiple in number showing male preponderance. They form as a result of trauma or due to familial multiple lipomatosis.¹⁴⁻¹⁶ Their size remains small but may reach huge size and weight over the years. Usually when small do not cause symptoms other than cosmetic concerns

but may cause compression of surrounding structures when attain large size. Its different variants include angiolipomas, neomorphic lipomas, spindle cell lipomas, and adenolipomas. Liposarcoma which is Malignant variant is hard to differentiate from a simple lipoma on basis of physical examination and radiology. That is why every lesion suspected as lipoma having greater than 5 cm size should raise suspicion and biopsy should be done to exclude Liposarcoma. Liposarcoma contribute to 15-20% of head and neck sarcoma with only 10-20% affecting children.

In parapharyngeal space it can cause oropharyngeal swelling, odynophagia, difficulty in breathing, cervical swelling, neurological deficit like hoarseness, slurring of speech, weakness of shoulder, blurring of vision and multiple cranial nerve palsy. CT scan and MRI and fine needle aspiration have enabled accurate assessment and approach to them. The area is tricky and approach is based on location, size, vascularity and malignant potential though rare.⁹



Fig 1: Extra Oral picture of Left side of Face showing swelling bulging from the submandibular region into the neck

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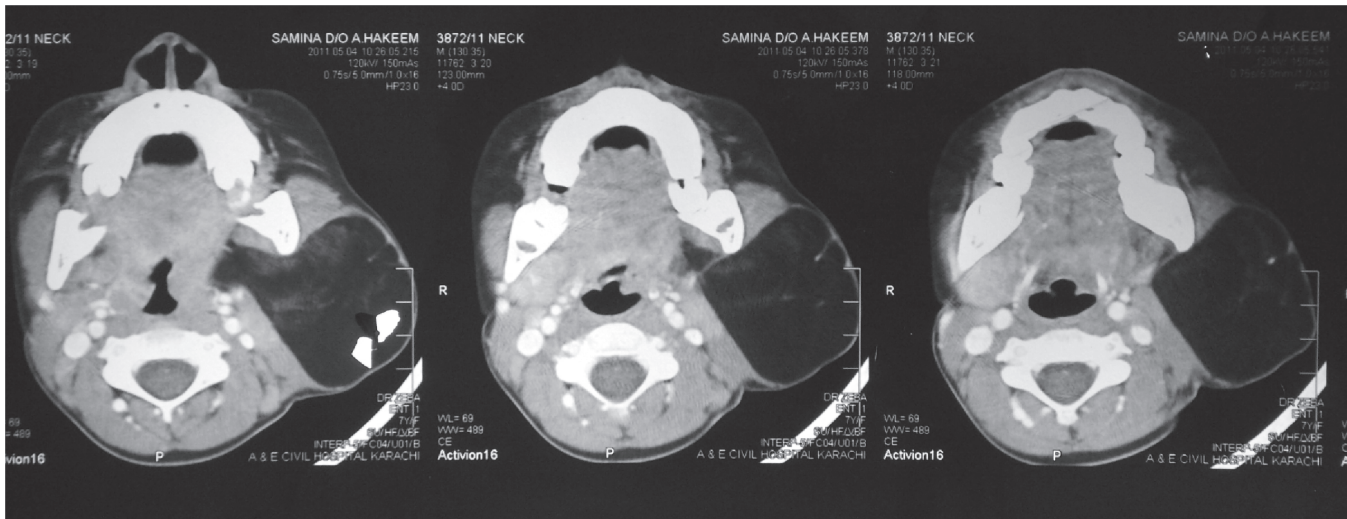


Fig 2: CT scan of the patient done 4 years back showed radiolucent area starting from the left parapharyngeal space.

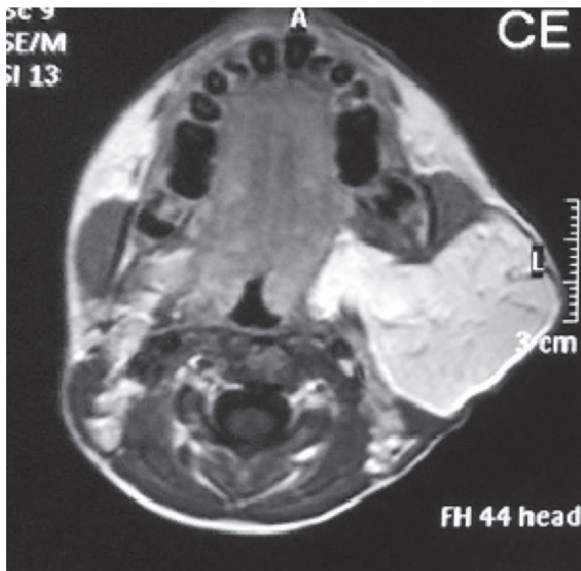


Fig 3: MRI scan of the patient prior to surgery, showing the soft tissue growth from the left parapharyngeal space extending upto the base of the skull



Fig 4: Pre-operative marking of trans-cervical approach

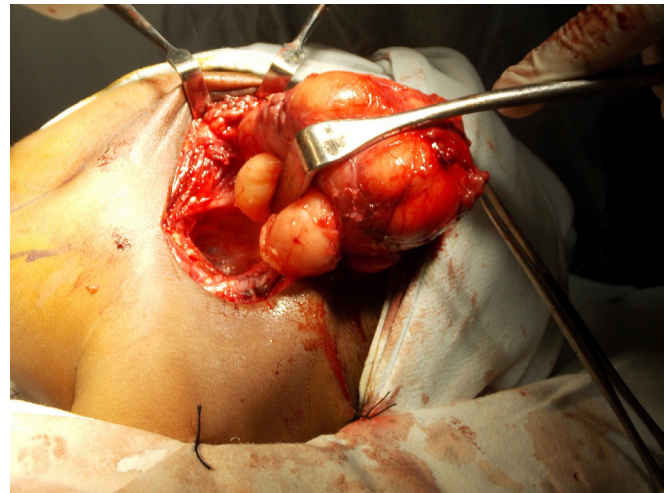


Fig 5: Intra-operative picture showing the fatty mass



Fig 6: Excised lesion from the parapharyngeal space

CASE REPORT

A 9 year old female presented with four year history of slowly progressive swelling of the left side of the neck causing dysphagia. No history of pain, pus/blood discharge was present. Patient had visited another



Fig 7: Post-operative picture showing closed incision and drain in place

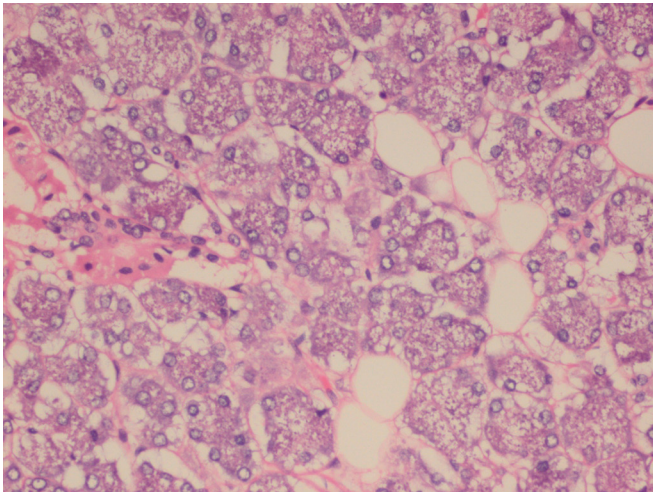


Fig 8: Histopathological picture showing fat cells surrounded by benign ducts

hospital based facility where she was diagnosed to have cystic Hygroma on basis of appearance. She didn't undergo treatment due to financial problems.

Overall patient appeared lean and thin. On examination soft, mobile, painless, non-tender mass having rubbery consistency was observed on left side of neck. Mass was extending superiorly up to the parotid and inferiorly upto level II of neck having approximately 7.2*6 cm size (Fig 1). The overlying skin had a scar otherwise it was normal without any signs of discoloration or abnormal vascularisation. Facial nerve function was intact. Submandibular lymph nodes were palpable.

Initial differential diagnosis on basis of clinical appearance and examination consisted of cystic hygroma and lipoma. After the MRI scan, differential included lipoma and liposarcoma. The ultrasound was advised which revealed hypoechoic area in left submandibular region measuring approximately 6.2/4.8cm showing linear echogenic striation, no fluid seen inside, no

vascularity noted on doppler. No evidence of invasion into surrounding tissues. Features are suggestive of benign lipoma. Previous CT scan (4 years back) of the patient showed radiolucent area in the parapharyngeal space displacing surrounding tissues (Fig 2).

The patient underwent MR imaging (Fig 3) T1,T2 W axial, sagittal and post contrast images of the neck were obtained. There was a abnormal signal intensity mass measuring 7.2+6.1+5.3cm in cranio- caudal, transverse and anteroposterior dimension seen in the subcutaneous tissues on left side of the neck. Superiorly it was extending up to the level of ramus of mandible, inferiorly extending up to the level of cricoid cartilage and medially extending into the left parapharyngeal space indenting the left lateral pharyngeal wall causing mild asymmetrical narrowing, laterally producing bulge in skin of neck on left side. It appears hyper intense on T1 and T2 images and show minimal contrast enhancement.

The mass was excised under general anesthesia via extended submandibular incision below the mandible in the submandibular region (Fig 4). Sub platysmal flap was raised and marginal mandibular branch of facial nerve was preserved. The mass had a fatty yellow appearance and was firmly adherent to the parotid salivary gland superiorly (Fig 5).

The posterior part of the mass was deeply extending towards the parapharyngeal space. While inferiorly lesion was extending up to the level II region of the neck. The tumour along with the gland was dissected free from the surrounding tissues and was excised completely along with the enlarged submandibular lymph nodes (Fig 6). The wound was primarily closed with silk 0/0 suture maintaining tight seal with drain placed (Fig 7).

The incision healed excellently. Patient was called on 7th day post-operatively to remove sutures. Overall outcome is good. Histopathological findings were consistent with lipoma showing partly encapsulated lesion composed of mature adipocytes with intervening thin capillaries (Fig 8). At places thin fibrous septae were seen imparting nodular configuration with focally entrapped benign ducts at periphery. Sections of Lymph node showed hyperplastic lymphoid follicles with pale germinal centers and intact mantle zones.

DISCUSSION

Lipoma is a benign tumor composed of mature white adipocytes and are the most common mesenchymal neoplasms in the adults and rarest in children. Known to affect the following sites in preference anterior neck, infratemporal fossa, oral cavity, hypopharynx, larynx, parotid gland and Parapharyngeal space.^{3,4}

Microscopically they are composed of lobules of mature adipocytes, identical to the surrounding adipose tissue except for slight variation in the size and shape of the cells in lipoma.¹ Depending on location, they may reach enormous size and weight many kilograms.¹⁷ Usually of little clinical concern and tend to grow insidiously large without causing any symptoms. PPS lipoma may cause few problems like dysphagia,

lower cranial nerve palsy, conductive hearing loss, sleep apnea and trismus^{3,9} which are due to localized mass compressing the surrounding structures and cosmetic concerns. Typically, these tumors are well circumscribed, are encapsulated (often by a fibrous shell), tend to be smooth or lobulated, are easy to remove, and rarely recur.⁸

Imaging evaluation of soft tissue tumors has undergone a dramatic evolution with the advent of CT scan and MR imaging, which are very useful for definite differential diagnosis. In a study CT scan proved to be 88% accurate in helping to reach a concurrent diagnosis. Comprised of mature adipose tissue, a classic lipoma has CT and MR imaging signal intensity characteristics similar to those of subcutaneous fat.¹⁸ The deep lesions tend to be variable in shape, likely because they tend to adopt according to the surrounding structures.¹⁹ It is important to differentiate benign lipoma from malignant liposarcoma as their appearance clinically are similar.

MR imaging provides better tumor delineation because it has superior soft tissue contrast resolution and clear definition of the location and longitudinal extent of the mass (giant infiltrating). It is particularly important in the oral facial region where the margins of lipoma are ill-defined because these lesions often are surrounded by normal fat tissue and have a very thin capsule.¹⁸

According to our review of the literature, our case is unique because of reaching this gigantic size in 4 years time and by far the second largest mass reported in PPS region.²⁰ Only mild dysphagia was reported by our patient despite of the huge size. MR imaging revealed that lipoma was medially extending into the left parapharyngeal space indenting the left lateral pharyngeal wall causing mild asymmetrical narrowing. The appearance was deceiving mimicking cystic hygroma which was the initial diagnosis. After the ultrasound and MRI differential diagnosis of lipoma and liposarcoma were made. Post-operatively lesion was sent for histopathology which confirmed the diagnosis of benign Lipoma.

CONCLUSION

Lipoma are rare lesions affecting the PPS in children. Though benign may attain huge size and may or may not cause symptoms. Approaching PPS lipoma is tricky territory specially in children and CT and MRI are very helpful in making the final diagnosis. Since, lipoma and liposarcoma are clinically similar, its important to make a final diagnosis either on basis of FNAC(Fine needle assisted cytology) or incisional

biopsy. Treatment is surgical removal of the complete lesion. Recurrences are rare.

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| 2 Noman Khan Niazi: | Helped in proof corrections/figs selection |
| 3 Zahid Ali: | Supervised the project. |