Fibrous hamartoma of infancy (FHI) of the parotid region in a 3-year-old female presenting as a huge parapharyngeal tumor: A very rare case presentation

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ABSTRACT

Introduction: Fibrous hamartoma of infancy (FHI) is a rare, benign tumor arising from the lower dermis and subcutaneous tissue. It usually occurs in children below two years of age and is often misdiagnosed as being of malignant nature. It most commonly effects the lower extremities, very rarely involves the head and neck region, and is commonly found in males. Microscopically, it is a poorly circumscribed and non-capsulated tumor comprising of interlacing fascicles of pale eosinophilic myofibroblasts and stellate shaped fibroblasts with island of mature adipocytes. Case Report: We report a very rare case of FHI of parotid gland in a 3-year-old female child, posing a diagnostic difficulty for management. The site, size, and age of presentation of the tumor were unique. This tumor is common in males but our case was a female child. Our patient presented with involvement of the parotid, submandibular and upper neck area with a huge size of around 7 × 8 cm which was causing mild swallowing difficulty due to intraoral swelling in the child. Cytopathology from the lesion demonstrated spindle cell lesion, the differential diagnosis of which is often malignant in pediatric age group. We took a biopsy for confirmatory diagnosis and after histopathological diagnosis of FHI proceeded with complete surgical excision. Conclusion: Treatment of choice for these benign tumors is complete surgical excision, with excellent prognosis.

Keywords: Fibrous hamartoma of infancy, Myofibroblasts, Parapharyngeal tumor, Parotid

INTRODUCTION

Fibrous hamartoma of infancy (FHI) is a rare benign tumor of subcutis and lower dermis which usually occurs within first two years of life [1, 2]. About 25% of them are congenital and 91% occur within first year of age. Only about 200 cases have been reported so far, emphasizing its rarity and diagnostic ambiguity. These lesions occur commonly in the superficial soft tissues of axilla, upper arm, trunk, or external genital area [3]. Fibrous tumor of infancy very rarely occurs in the parotid region and primary FHI of the parotid region has not been reported in such young age group making this case a unique one.
to report. One case of hamartoma of the parotid has been reported in a 16-year-old boy [4]. Histologically, a poorly circumscribed tissue with triphasic elements of fibrous trabeculae, mature adipose cells, and immature mesenchymal tissues is specific for this diagnosis. The treatment of choice is complete surgical excision of tumor. Its clinical course is typically benign and slow growing with excellent prognosis.

CASE REPORT

A 3-year-old female child presented to our outpatient department with chief complaint of a swelling in the right parotid region. It was of insidious onset which gradually progressed over one year to its present size. It was associated with compressive symptom of mild dysphagia though no breathing difficulty was present. The child was otherwise healthy with the parents’ only concern being suspicion of malignancy and cosmetic deformity. On clinical examination the child was found to have a swelling approximately $7 \times 8$ cm in size arising from the right parotid region. It had a smooth surface, which extended inferiorly up to thyroid gland, medially reaching up to midline and posteriorly encroaching the posterior triangle of neck (Figure 1). On palpation, the swelling had a multilobulated surface with overlying skin being normal in appearance. It was firm in consistency, and seemed adherent to surrounding deeper structures. It pushed the right tonsil medially. The swelling was non-pulsatile in nature. The facial nerve function was intact.

We performed a contrast enhanced computed tomography (CT) scan, which revealed a large heterogenous mass lesion in the right side of neck with soft tissue enhancement and fatty component within, measuring $7.0 \times 7.2 \times 8.3$ cm. The mass lesion was seen to be arising from right parotid region extending medially into parapharyngeal space, abutting nasopharynx and oropharynx. The lesion was also extending superiorly from the infra-auricular region to inferiorly up to upper level of thyroid gland. Parotid gland was displaced and stretched out laterally. Posteriorly the lesion was abutting the carotid space. Anteriorly muscles of mastication were displaced with loss of fat plane. Anteroinferiorly it extended into the floor of mouth displacing mylohyoid muscle medially with remodeling of adjacent body of mandible. Submandibular gland was displaced posteroinferiorly. Facial and temporal arteries were encased by the lesion (Figures 2 and 3). On obtaining a fine needle aspiration cytology, a spindle cell lesion was suspected. Spindle cell lesion of the head and neck region is a rare diagnosis. The differential diagnosis of spindle cell lesion in pediatric age group includes both benign and malignant lesion. It includes lesions of neural, fibroblastic, myofibroblastic, myogenic, and epithelial tumor.

The case was discussed in the institutional tumor board among departments of radiotherapy, medical oncology, surgical oncology, and ENT (ear, nose, and throat). It was decided to perform a diagnostic tissue biopsy and to proceed further only after the histopathological report. Routine hematological and biochemical investigations were done, which were within normal limits. Pre-anesthetic checkup was done. Tissue biopsy was done under general anesthesia and the histopathological report provided a differential diagnosis of embryonal rhabdomyosarcoma or FHI. Immunohistochemistry (IHC) was further obtained which concluded the lesion to be FHI. Pathological picture revealed areas of disorderly arranged collagenous fibrous tissue with delicate blood vessels and mild inflammatory cells. Islands of mature adipocytes were found between fibrocollagenous stroma. At places mature skeletal muscle fibers were also noted.
There was no atypia/mitosis/necrosis seen in the sections. The attached salivary gland was of unremarkable histology and all the six lymph nodes which were attached to the salivary tissue showed only reactive changes. A histological diagnosis of FHI was given and IHC was advised for further confirmation. Immunohistochemistry revealed positive stains for CD 34 in fibrous and myxoid areas, Vimentin in all areas, S-100 in adipocytes, Desmin in some cells in fibrous areas. It was negative for spinal muscular atrophy (SMA), h-Caldesmon, Myogenin, Myo D1, MUC-4, Ki-67 and was just 0.4%. The final diagnosis of FHI was confirmed after IHC (Figure 4).

Following this, surgical excision with the aim of complete removal of the tumor was planned with a possible tracheostomy and mandibulotomy, if required. Airway management was of paramount important in this patient. Due to the sheer size of this tumor and its difficult location we had planned elective tracheostomy if fiberoptic guided intubation would fail. Nasal intubation could be performed using the pediatric fiberoptic bronchoscope and tracheostomy could be avoided. The external carotid artery was ligated beforehand to reduce vascularity. However, intraoperatively, the tumor was not found to be densely adherent to the surrounding structures and hence could be completely dissected off from adjacent structures without sacrificing facial nerve (Figure 5). Complete surgical excision of the tumor was done and defect was closed primarily (Figure 6). The patient did well postoperatively and was discharged from the hospital after an uneventful recovery. Her facial nerve function was intact in the postoperative period. Histopathological examination (HPE) of the complete specimen confirmed it to be a case of FHI (description of which is enumerated above).
DISCUSSION

Fibrous hamartoma of infancy is poorly circumscribed superficial soft tissue mass. It is a rare benign tumor mostly diagnosed before two years of life. It is most commonly found in axilla, shoulder, upper arm, inguinal region, and chest wall, while rarely occurring in head and neck region. The case which we describe here had this lesion affecting the parotid and upper neck region making it a very rare site of involvement. Fibrous hamartoma of infancy was first described by Reye in 1954 [1]. Enzinger studied 30 cases and coined the term fibrous hamartoma of infancy [2]. These are usually firm in consistency, and may be affixed to the underlying tissue, thus causing a concern for malignancy [5]. Radiological investigations usually are nonspecific, and reveal well delineated soft tissue mass with varying intensities suggestive of fat and muscle tissues [3]. Imaging studies are useful for surgical planning to look for its attachment to surrounding structures. Initially, we were in dilemma due to the cytopathological report of “spindle cell neoplasm" the differential diagnosis of which ranges from benign to malignant lesion of mesenchymal. For confirmatory diagnosis we had to go ahead with incisional biopsy as management of these lesions may differ depending on the final HPE diagnosis. Incisional biopsy revealed FHI. Microscopically, FHI usually shows a poorly circumscribed unencapsulated tumor composed of interlacing fascicles of pale eosinophilic myofibroblasts and stellate shaped fibroblasts against a myxoid stroma. Areas of irregularly arranged collagenous fibrous tissue with delicate blood vessels and mild inflammatory cells are present. Islands of mature adipocytes are found in between fibro collagenous stroma. At places mature skeletal muscle fibers are also noted. No atypia/mitosis/necrosis is however seen. Fibrous hamartoma of infancy is often misdiagnosed as lipoma, neurofibroma, hemangioma, or dermatofibroma [6]. The histological picture of the incisional biopsy in our case was similar to that described. The HPE diagnosis of the postoperative specimen too showed similar results. Diagnosis of FHI was further confirmed by IHC.

The treatment of choice is local excision with a very low recurrence rate [7]. Surgical excision of the tumor in our case was challenging due to its size and proximity to important neurovascular structures in the neck. We avoided tracheostomy in the first instant due to fear of difficult decannulation in pediatric age group. Fiberoptic guided nasal intubation was performed. We were prepared for mandibulotomy for complete exposure and dissection of tumor but the tumor mass had already made enough “space" around it and we could dissect the whole tumor from the neck and parapharyngeal area without any mandibulotomy. Facial nerve could also be salvaged though it was very stretched over the surface of the tumor. Excessive bleeding was avoided as we had already done and external carotid artery ligation beforehand.

In these large sized tumors in pediatric age group, excessive intraoperative bleeding is a major concern which we anticipated beforehand. External carotid ligation was done beforehand and adequate blood products were arranged keeping in mind the low blood reserve in this age group patients.

Injury to important neurovascular structures lies the carotids and the lower cranial nerves in the neck are always a possible complication which we did not encounter in our case. Facial nerve could also be saved and our patient had no facial nerve paresis in the postoperative period.

Incomplete removal of tumor is always a possibility in such large tumors. We could remove the tumor completely and no residual lesion was left behind.

The differential diagnosis for subcutaneous swelling in an infant includes both benign and malignant soft tissue tumors such as epidermoid cyst, recurring digital fibrous tumor, juvenile aponeurotic fibroma, juvenile hyaline fibromatosis, histiocytoma, dermatofibroma, leiomyosarcoma, and fibrosarcoma [8].

Fibrous hamartoma of infancy is not a very common pathology that too in the head and neck region making our case a unique one to report. The management was challenging but successfully achieved without any adverse outcomes.

CONCLUSION

The clinical course of FHI is benign and slowly progressive, and the prognosis is excellent. Its diagnosis is confirmed on the basis of histopathological criteria which needs to be confirmed on immunohistochemistry. Owing to its rarity, a misdiagnosis of malignancy is common. With adequate surgical management, recurrence is negligible. Hence, a wide clinical awareness is necessary to avoid diagnostic delay, and to aid in effective and efficient management of this rare condition.

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Author Contributions

Ragini Raina – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Kranti Bhavana – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Swati Suneha – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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