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Research Article

Fibrous Hamartoma of Infancy: An Unusual Site

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Abstract

Fibrous hamartoma of infancy is a rare entity typically occurring in the first two years of childhood. So far head and neck contributes to only 6.5 percent of total cases. Diagnosis is by characteristic features in radiology and histopathology. Meticulous surgical excision is the treatment of choice with an excellent prognosis. We present a case report of Fibrous hamartoma of Infancy (FHI) at a tertiary pediatric care centre in a 2 year old male presenting in the parotid space. Post MRI imaging the patient underwent a surgery following which the diagnosis was confirmed with immunohistochemistry of the histopathology specimen. Postoperative period was relatively uneventful. Hence accurate radiodiagnosis in the form of MRI and meticulous surgical excision teamed with tissue diagnosis is the mainstay in achieving the optimum results.

Keywords: Hamartoma; Infancy; Parotid, Surgery; Immunohistochemistry; Facial Nerve

Introduction

Most of the head neck tumours of infancy are benign. Hamartoma is multilineage tumor with mature cells and tissues in a disarrayed fashion. It is a benign tumor-like malformation that consists of a collection of architecturally disorganized cells located in an area of the body where the cells are normally found. It is often due to abnormal development. Fibrous hamartoma of infancy (FHI) is a rare, benign tumor of the subcutis and lower dermis, which usually occurs within the first 2 years of life.

We hereby give case report of an unusual site of fibrous hamartoma in a 2 year old child at a tertiary centre.

Materials and Methods

A two year old male child presented in the outpatient department with swelling in the right parotid region which was noticed since 2 months. The swelling progressively increased to the present size. The swelling was not associated with fever, pain or any other systemic complaints. On examination, the swelling was approximately 5 cm x 5 cm on examination, firm in consistency, non-tender, mobile and skin over the swelling was free. There were no pulsations, no restriction of mouth opening, facial nerve function was unaffected and no bulge was seen on intraoral examination

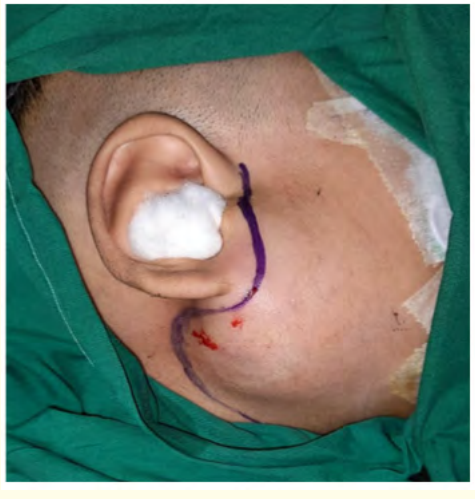

Figure 1: Right pre and infra auricular swelling.

Magnetic Resonance Imaging of the swelling revealed a well-defined lobulated lesion of about 5.1 x 5.4 x 4.3 cm arising from the parotid space including the superficial and deep lobe of parotid mildly displacing the carotid space and in between the medial pterygoid and masseter with displacement of the mandibular condyle. There was no involvement of pharyngeal mucosal space, vascular invasion or external auditory canal involvement. T1 and T2 hyperintense components of fat and isointense to hypointense solid components on contrast and ossified components were seen. Differential diagnosis of teratoma or a fibrous hamartoma of infancy was made.

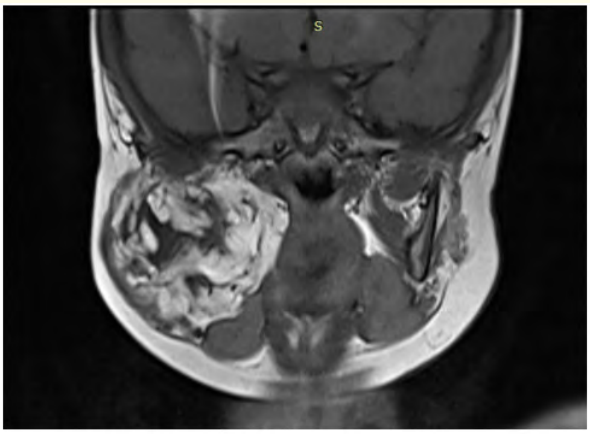

Figure 2: Magnetic Resonance Imaging showing T1 Coronal view.

Fine needle aspiration cytology revealed scattered spindle cells and on suggestion of the pathologist, the patient underwent a trucut biopsy. Biopsy report revealed benign cartilaginous components. As teratoma was one of the differential diagnosis considered, Beta HCG, serum Lactate Dehydrogenase and serum Alpha Fetoprotein levels were tested preoperatively and were normal.

The decision of surgical excision was made. The child underwent a total parotidectomy under general anaesthesia. Nerve monitoring was used to preserve Facial nerve function. The superficial parotid was thinned out and dissected. The facial nerve was stretched over the enlarged deep lobe of parotid gland which was could not be appreciated separately from the mass. The dissection posed a challenge and the nerve had to be mobilized superiorly along with branches while dissecting the tumor from the base. The tumor was firmly attached to the ramus of the mandible and the deeper tissues. Post dissection the facial nerve was preserved along with the branches as shown in the figure 3.

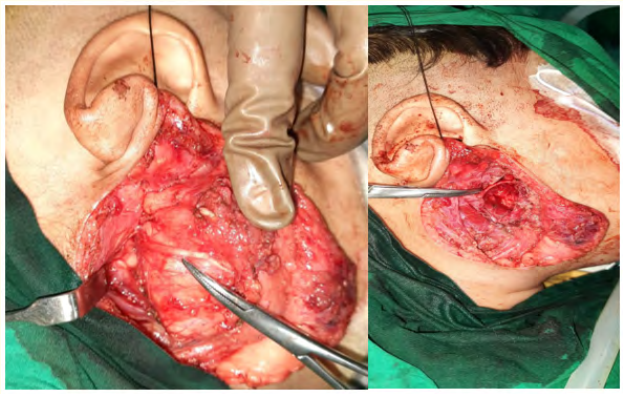

Figure 3: Preservation of the facial nerve after total excision.

The nerve monitor did not detect any impulse in the buccofacial branch post operatively. It did detect an impulse in the zygomaticotemporal branch. The excised specimen, preparotid and level II lymph nodes were sent for histopathological examination.

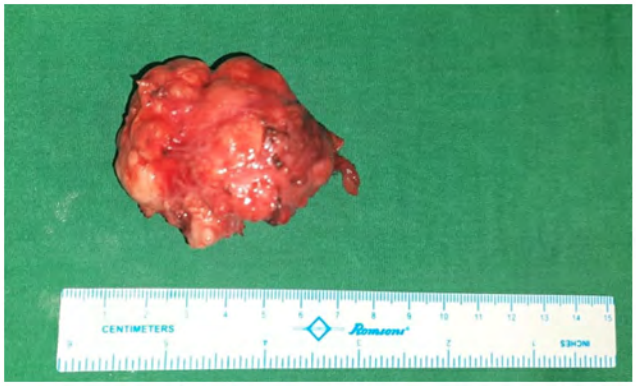

Figure 4: The excised tumor.

Results

In postoperative period the patient had a grade 4 paresis of the right lower facial region. The child received short term systemic corticosteroids and facial physiotherapy after suture removal and the facial nerve function recovered completely in one month.

The histopathology report revealed normal superficial parotid lobe. The deep lobe had benign cell infantile fibromatosis which was a circumscribed lesion of bands of heavily collagenised tissue of spindle and stellate cells in vague fascicles. There was myxoid stromal change and interspersed tissue of mature adipocytes. Metaplasic bone was also seen. There were no lipoblasts or hemangiopericytomatous vasculature or immature mesenchyme. Immunohistochemistry showed the spindle cells which expressed CD34 and Desmin. It was negative for SMA, myogenin, betacatenin and S100 Protein suggestive of Fibrous Hamartoma of Infancy.

Discussion

Fibrous hamartoma of infancy is a rare benign lesion most frequently occurring within the first two years of life. It was first described by Reye in 1956 and was named as FHI by Enzinger in 1965 [11,12]. The vast majority of the cases (91%) occur within the first year of life and about 25% of them are congenital [12].The lesions most frequently involve the superficial soft tissues and may occur anywhere in the body, for example, axilla, upper arm, trunk, external genital area, and so on. In our case the subject is a 2 year old male with parotid swelling.

On MR imaging, there were reports that the tumours of FHI had signal intensity similar to fibromatosis and some had intensity similar to lipoma. All findings displayed triphasic morphology, but varied widely in the relative percentages of fat, fibroblastic fascicles, and primitive mesenchyme. Hyalinized zones with cracking artifact, mimicking giant cell fibroblastoma were also present. A component was considered 'predominant' if it occupied equal or more than 45% of the entire lesion [7,8]. Parallel or whirling appearance, and dilated vessels [5]. The presence of fat containing subcutaneous mass with intersecting bands of fibrous signal tissue in the appropriate age group is highly suggestive of the diagnosis [8].

In a retrospective review study by S. Lee., *et al*. of the sonographic features of confirmed 13 cases of fibrous hamartoma of infancy in pediatric patients all masses demonstrate heterogeneous hyperechogenicity with a "serpentine pattern" of intervening hypoechoic portions in the hyperechoic mass. The margins were ill-defined or lobulated [17].

Our report showed a variable intensity with T1 T2 hyperintense components of fat and isointense to hypointense on contrast.

The histological appearance of the tumor is characterized by three different tissues: fibrous trabeculae, mature adipose cells, and immature mesenchymal tissues which is specific for diagnosis of FHI. There have been two large clinicopathological reviews. A review of 60 patients with FHI by Saab., *et al*. reported an unusual pseudoangiomatous pattern in about half of the cases [1]. In a study of 145 cases by El Ibrahimi., *et al*. at Mayo clinic confirms the classic clinicopathologic features, including the triphasic morphologic appearance of most cases. All displayed triphasic morphology, but varied widely in the relative percentages of fat, fibroblastic fascicles, and primitive mesenchyme.

C Sotelo-Avila., *et al*. had a study of 40 cases of subdermal fibrous hamartoma of infancy all had the characteristic mixture of fibrous and adipose tissue [3].

Our report showed myxoid stromal change and interspersed tissue of mature adipocytes.

Immunohistochemistry is used to confirm the diagnosis. IHC findings show Vimentin positivity was noted in both areas, whereas desmin and actin positivity was found mainly in the fascicular-fibroblastic regions [16]. Saab., *et al*. demonstrated in immunohistochemistry (IHC) that there was reactivity for smooth muscle actin (SMA) and CD34 in the mature fibrous tissue and variable CD34 expression in the primitive mesenchyme [1].

Our report showed that spindle cells expressed CD34 and Desmin. It was negative for SMA, myogenin, betacatenin and S100 Protein.

Conclusion

Fibrous hamartoma of infancy sometimes presents at unusual sites like in our case. The differential diagnosis can be a teratoma or a fibromyoma. Magnetic resonance imaging cytology and immunohistochemistry are the mainstay for diagnosis. Treatment is surgical excision and gives a permanent cure with minimal complications. Prognosis is excellent. Wait and watch policy will only allow the tumor to keep growing and further compromise function.

Conflict of Interest

This is no financial interest or any conflict of interest exists.

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