



RIGA FEDE DISEASE- AN UNUSUAL PRESENTATION

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ABSTRACT

A 7-year-old female child came to hospital with history of swelling over left side of tongue from 10 days. Swelling was identified accidentally by her mother. There were no complaints per se from the patient. On examination there was 2x1cm smooth swelling over dorsal surface of the left side of tongue in anterior one-third. It was a firm swelling with well-defined margins. Magnetic Resonant Imaging of neck with contrast was done which showed a T1 Hypointense with T2 hyperintense peripherally enhancing lesion of the left genioglossus which was suggestive of Fibroma/Dermoid cyst/mucous retention cyst. Cyst was excised in toto under General Anaesthesia and sent for Histopathological examination which showed features of Riga Fede disease (Eosinophilic ulcer of the tongue). Patient was followed up for 3 months and no signs of recurrence were seen.

KEYWORDS

Riga Fede Disease, Traumatic Ulcerative Granuloma with Stromal Eosinophilia (TUGSE), Tongue swelling.

INTRODUCTION:

Riga Fede Disease (RFD) presents as traumatic sublingual ulceration that occurs in neonates and infants involving the ventral surface of tongue. It is most commonly associated with natal and neonatal teeth in infants. (1) It can also occur in older infants due to repetitive trauma by erupted primary lower incisors. Lesion begins as an ulcer over the ventral surface of tongue which can progress to fibrous mass.(1) It was first described by Italian physician Antonio Riga in 1881 subsequently histological findings were published by F. Fede in 1890.(2) Elzay noted similar presentation in older children and adults, termed this condition as Traumatic Ulcerative Granuloma with Stromal Eosinophilia (TUGSE) where there is an increased eosinophilic infiltration found in the connective tissue below these lesions.(3)

It can be further classified into Precocious RFD and late RFD. Precocious RFD associated with natal or neonatal tooth in the first 6 months of life while Late RFD is seen in cases with dental eruption after 6-8 months of life. (4) Here we are discussing about a rare presentation of late RFD.

CASE REPORT:

A 7-year-old female came to hospital with history of swelling over left side of tongue from 10 days. Swelling was identified accidentally by her mother. It was not associated with pain or difficulty in swallowing or chewing or speech production. She had no previous history of recurrent trauma to tongue or problems with dentition. Examination revealed single non-tender firm swelling of 2cm x 1cm with well-defined margins over the dorsal surface of anterior one-third of tongue; extending from midline up to 0.5cm from the lateral border of the tongue. The overlying and surrounding mucosa was normal. There were no sharp tooth and the child had good oro-dental hygiene. A provisional diagnosis of mucous retention cyst, lingual dermoid cyst was made.

Magnetic Resonant Imaging (MRI) of neck with contrast was done which showed T1 Hypointense with T2 hyperintense peripherally enhancing lesion measuring 1.8 x 1.1 x 1.1 cm involving the left genioglossus which was suggestive of Fibroma/Dermoid cyst/mucous retention cyst. (Fig.1)

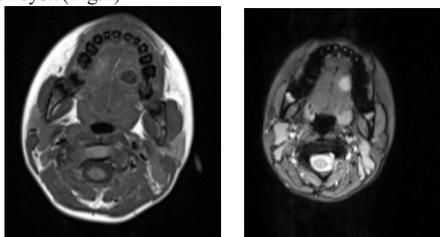


Fig.1MRI Neck with contrast showing T1 Hypointense with T2

hyperintense peripherally enhancing lesion measuring 1.8 x 1.1 x 1.1 cm involving the left genioglossus.

Under general anaesthesia transoral examination revealed a 2cm x 1cm well defined cyst which was excised submucosally. There was no infiltration of adjacent tongue musculature. The cyst was excised in toto (Fig.2) and sent for Histopathological analysis. Wound closed with sutures. Histopathological examination (HPE) revealed an inflammatory infiltrate predominantly composed of eosinophils, lymphocytes and macrophages with ill-defined granuloma suggestive of RFD(Fig.3)



Fig.2 Intraoperative picture showing the dissected specimen

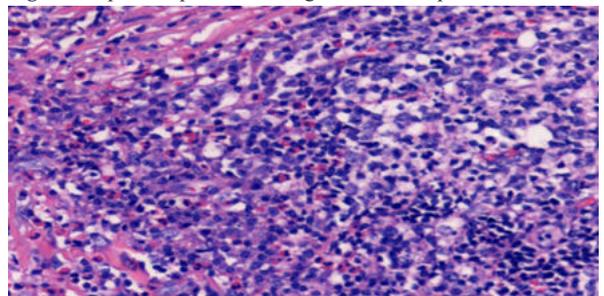


Fig.3 HPE showing inflammatory infiltrate predominantly composed of eosinophils, lymphocytes and macrophages [magnification 20x]

Immediate post-operative period was uneventful. At three-month post-operative follow up, a well healed wound was observed and the child

was comfortable with regular oral diet.

DISCUSSION:

RFD is a benign and uncommon mucosal disorder caused by chronic and recurrent trauma to tongue against the mandibular incisors (5). It is also termed as TUGSE by Elzay and is classified into Precocious RFD and late RFD (3, 4, 5). There is no gender predilection (5). Most common presentation is an ulcer over the oral cavity mucosa however it can also present as an indurated submucosal granuloma. It is known to be confined within the oral cavity, with tongue being the most common affected site (6). Lesions can be painful or asymptomatic. Microscopically numerous eosinophils and large histiocytes with pale nuclei are characteristic (7).

RFD is known to occur in infants and children younger than 2 years of age in association with the eruption of first dentition, mainly the mandibular incisors (8). However, it has also been reported in adults due to chronic trauma to the oral cavity as a result of ill fitted dentures, broken tooth or due to involuntary tongue movements associated with psychological conditions. Ours is a rare case with a late presentation at the age of 7 years without any history of chronic localised trauma to the tongue or problems with dental eruption.

Most cases in literature have reported infants presenting with painful ulcer over the ventral surface of tongue due to natal and neonatal tooth (2,5,9). Wollina et al reported a 70-year-old female with painful ulcer over the tongue with raised borders for four years due to repetitive trauma from an ill-fitting denture. Within 2 weeks of not using ill fitted dentures complete resolution was observed (10). Ours is a unique case as there was no ulcer in oral cavity and the presentation was a swelling over the dorsal surface of the tongue which was quite uncommon in RFD. Intraoperatively we noticed it to be a well-defined submucosal cystic swelling based on which lingual dermoid was suspected. However, HPE which showed inflammatory infiltrate predominantly composed of eosinophils, lymphocytes and macrophages with ill-defined granuloma indicative of RFD.

A variety of management options were discussed in literature such as modification of feeding habits, weaning, nasogastric tube feeds, filing of teeth, dental extraction (4, 6, 10). In our case no specific causative factor could be identified. Post-operative follow-up was uneventful with good wound healing and no signs of recurrence.

CONCLUSION:

Our aim is to highlight the rare presentation of RFD with no underlying causative factor in a child presenting with tongue swelling. In absence of unremarkable features on radiological evaluation, a diagnosis of RFD should be kept in mind. Excision of the lesion is a safe treatment option and long term follow up is recommended to note the possible recurrence.

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