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Case Report Open Access

Isolated Neurofibroma of Lid Margin in a Case of Paediatric Sjogren's Syndrome - A Rare Case Report

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Abstract

Sjögren syndrome (SS) is a chronic, inflammatory and autoimmune disease characterized by progressive lymphocytic and plasma cell infiltration of the salivary and lacrimal glands. It is observed in 90% of women in the age group 35-45 yr. The primary SS is very rare in childhood. It may be a primary or secondary disorder to other autoimmune disorders, such as systemic lupus erythematosus (SLE), rheumatoid arthritis, scleroderma and biliary cirrhosis; but association with neurofibromatosis is not reported till date. Isolated neurofibroma of lid is very rare, though it can be associated with Von Recklinghausen's disease. We report here a case of Primary Pediatric Sjogren's Syndrome presented with isolated neurofibroma of lid margin.

Keywords: Paediatric sjogrens syndrome; Neurofibroma; Schirmer's test

Introduction

Sjögren syndrome, a slowly progressive inflammatory disorder that involves the exocrine glands is a rare entity in paediatric patients [1]. In the late 1800s, Mikulicz and others recognized the findings of kerato-conjunctivitis and xerostomia as an entity [2]. In 1933, Sjögren recognized the association of this symptom complex with polyarthritis.

Primary disease is rare in childhood, as only 145 cases of primary disease have been described in the international pediatric literature [3]. Only 5% of adult patients report onset of symptoms before twelve years of age.

The clinical manifestations and classification schema differ in pediatric patients; therefore, this disorder might be under reported. The female-to-male ratio for Sjögren syndrome was 7:1 in one of the multicenter international pediatric cohort and 6:1 in a systematic review of the literature by Cimaz et al. [3,4]. In adults, the female-to-male ratio is approximately 9:1 [5]. The constellation of symptoms seen in children (i.e., lower frequency of Sicca syndrome, higher rates of parotid enlargement, higher prevalence of immunologic markers) may be similar to those found in young adult patients (i.e, <thirty five years) [6].

Neurofibroma is a neurogenic tumour arising from Schwann cells or peripheral tissues of nerve sheaths. It is usually associated with Von Recklinghausen's disease rather than presenting as a solitary tumour. It is commonly found in the head and neck and the flexoral surfaces of the upper and lower extremities [7]. Till date no relationship has been documented between a chronic inflammatory disease and neurofibromatosis .We report a case of Pediatric Sjogrens Syndrome (PSS) with a unique presentation in the form of bilateral swelling of parotid glands and isolated neurofibroma of lid margin.

Case Report

A thirteen years old girl presented in Ophthalmology clinic with a mass at the lid margin near lateral canthus of her left eye (Figure 1). On examination, a nodular swelling about four mm in size, firm in consistency, was present at lower lid margin near lateral canthus. It was initially small about size of a two mm, non-tender to touch and gradually increased to current size within two year period. Her visual acuity was 20/20 OU associated with signs of conjunctival xerosis. Her mother pointed out that since birth she had a tearless cry. Her anterior and posterior segment examination was normal. On external examination she had a typical facial appearance.

Examination of lower half of face revealed enlargement of both parotid and submandibular gland along with chapped lips (Figure 2). Oral examination done by an oral surgeon demonstrated red and congested mucosa, gingivitis, and decalcification of the teeth at the cervical margin.

The dorsal surface of the tongue showed generalized atrophy of the filiform papillae and glossitis (Figure 3). She also complained of difficulties in chewing and swallowing, burning sensation while eating spicy and acidic foods.

Systemic evaluation done by pediatrician revealed no abnormality in her nervous and musculoskeletal system except that she was small

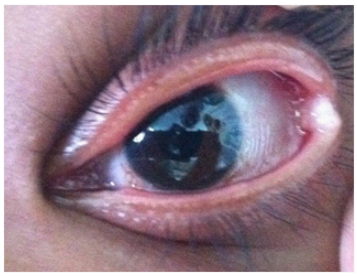

Figure 1: Clinical photograph of girl with a mass at the lid margin near lateral canthus of her left eye.

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for her age. Her Schirmer's test was significant in both eyes less than five mm in left eye and less than ten mm in right eye.

Biopsy obtained through normal-appearing mucosa of minor salivary gland revealed focal lymphocytic sialo-adenitis. Serology tests for SS-A and SS-B were positive, but negative for antinuclear antibody (ANA) and rheumatoid factor (RF). Patient 's ocular signs, oral signs, and results of serological test were fulfilling the criteria [8] of not only Paediatric Sjogren Syndrome but also American-European Consensus Group (AECG) criteria [9] confirming her as case of Paediatric Sjogren syndrome (PSS). In her family, her younger sister also had similar complaint of tearless cry but she didn't report to our clinic.

The plan of management was excision of lid mass which was done successfully and was sent for histopathological examination. Surprisingly, her histopathology report showed unremarkable 'conjunctival lining epithelium with deeper tissue showing extensive neural bundles forming fascicles' (Figure 4).

Cells in the tissue were elongated with the serpentine nucleus which was suggestive of neurofibroma. We re-evaluated the tissue and it confirmed the diagnosis of neurofibroma. Again her systemic evaluation was done to look for the other signs of neurofibromatosis [10] as proposed by National Institutes of Health or any other similar growth present anywhere in body along with CT scan brain, but signs related to neurofibromatosis were absent. Until this time no such case has been reported in literature where isolated neurofibroma of eyelid was seen in a patient of PSS.

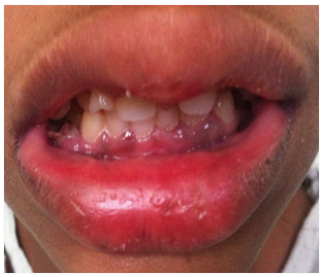

Figure 2: Oral examination shows red and congested mucosa, gingivitis, and decalcification of teeth at the cervical margin.

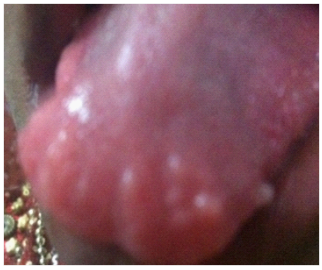

Figure 3: The dorsal surface of the tongue demonstrates generalized atrophy of the filiform papillae and glossitis.

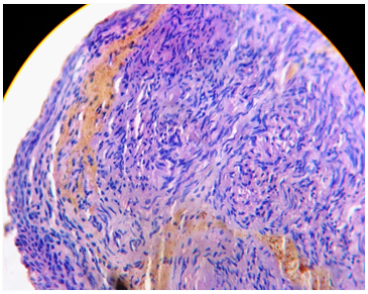

Figure 4: Conjunctival lining epithelium (thin arrow) with deeper tissue showing extensive neural bundles (thick arrow) forming fascicles.

Discussion

Sjögren syndrome (SS) is a rare and possibly under diagnosed condition in pediatric age group. Nikolas et al. reported a case of primary SS (PSS) in a four-year-old Venezuelan girl .The patient presented with bilateral recurrent parotid enlargement, predominantly on the right side. She did not complain of dry mouth or eyes [11]. Similarly our patient didn't complain of dry mouth or dry eyes, but she had a swelling of salivary glands.

The mean age of pediatric patients with SS is 9.4 (2.2) years, ranging from six to fourteen years [12]. Various studies conducted till date have demonstrated that Sjögren syndrome may be a primary disorder or may be secondary to other autoimmune disorders, such as systemic lupus erythematosus (SLE), rheumatoid arthritis, scleroderma, and biliary cirrhosis. It may overlap with other pediatric autoimmune disorders and, less commonly, may present as a primary condition, but never presented with neurofibromatosis, as per literature review, which was seen in our patient.

Kobayashi et al. reported four cases of childhood SS complicated by chronic thyroiditis, interstitial nephritis or sweat gland inflammation. Additionally, in one of these cases, the central nervous system was involved, but in our patient, no systemic abnormality was present [13]. Renal tubular acidosis should be excluded in children and adolescents with SS as this can be a cause of weakness, fatigue or growth failure. Early recognition would reduce long-term complications such as growth failure [14]. Though our patient had growth failure but her kidney function tests were within normal limits. Recurrent parotid gland enlargement is the most common clinical finding at disease onset in children, but chief complaint of our patient was tumour at lid margin though she was having symptoms of dry mouth, difficulty in swallowing, and irritation in mouth and swelling of cheeks.

Peripheral nerve-sheath tumours are divided into neurofibroma, schwannoma and neurogenic sarcoma. Neurofibroma and schwannoma are classified as benign, and neurogenic sarcoma as malignant.

Solitary neuroma is rare, especially in the eyelids, conjunctiva or orbit. Naoko et al. reported two cases of solitary neurofibroma of tarsal plate in woman without signs of cafe-au-lait spots, iris nodules or other neuromas [15].

PSS can present as an acute onset of generalized lymphadenopathy mimicking lymphoma as reported by Buonuomo et al. [16] but PSS associated with neuofibroma is an atypical form of presentation seen in our case.

Sjögren syndrome, a slowly progressive inflammatory disorder that involves the exocrine glands, itself is rare in pediatric patients, its relationship with neurofibromatosis, which is characterized by neuroectodermal tumors arising within multiple organs and autosomal-dominant inheritance has not been documented. Though close follow up is kept with patient, as with age other signs of neurofibromatosis can develop.

References

1. Fox RI (2005) Sjögren's syndrome. Lancet 366: 321-331.

2. Jonsson R, Haga HJ, Gordon TP (2001) Sjogren's syndrome. In: Koopman WJ (Ed) Arthritis and Allied Conditions: A Textbook of Rheumatology. (14th edn) Lippincott, Williams, & Wilkins, Philadelphia, PA: 1736-1759.
3. Civilibal M, Canpolat N, Yurt A, Kurugoglu S, Erdamar S, et al. (2007) A child with primary Sjögren syndrome and a review of the literature. Clin Pediatr (Phila) 46: 738-742.
4. Cimaz R, Casadei A, Rose C, Bartunkova J, Sediva A, et al. (2003) Primary Sjögren syndrome in the paediatric age: a multicentre survey. Eur J Pediatr 162: 661-665.
5. Anaya JM, Ogawa N, Talal N (1995) Sjögren's syndrome in childhood. J Rheumatol 22: 1152-1158.
6. Ramos-Casals M, Solans R, Rosas J, Camps MT, Gil A, et al. (2008) Primary Sjögren syndrome in Spain: clinical and immunologic expression in 1010 patients. Medicine (Baltimore) 87: 210-219.
7. Kim YD, Bai CH, Suh JS, Song KW (1997) Transnasal endoscopic excision of an isolated neurofibroma of the nasal septum. Rhinology 35: 89-91.
8. Houghton K, Malleson P, Cabral D, Petty R, Tucker L (2005) Primary Sjögren's syndrome in children and adolescents: are proposed diagnostic criteria applicable? J Rheumatol 32: 2225-2232.
9. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, et al. (2002) Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 61: 554-558.
10. Gutmann DH, Aylsworth A, Carey JC, Korf B, Marks J, et al. (1997) The diagnostic evaluation and multidisciplinary management of neurofibromatosis 1 and neurofibromatosis 2. JAMA 278: 51-57.
11. Nikitakis NG, Rivera H, Lariccia C, Papadimitriou JC, Sauk JJ (2003) Primary Sjögren syndrome in childhood: report of a case and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 96: 42-47.
12. Drosos AA, Tsiakou EK, Tsifetaki N, Politi EN, Siamopoulou-Mavridou A (1997) Subgroups of primary Sjögren's syndrome. Sjögren's syndrome in male and paediatric Greek patients. Ann Rheum Dis 56: 333-335.
13. Kobayashi I, Furuta H, Tame A, Kawamura N, Kojima K, et al. (1996) Complications of childhood Sjögren syndrome. Eur J Pediatr 155: 890-894.
14. Pessler F, Emery H, Dai L, Wu YM, Monash B, et al. (2006) The spectrum of renal tubular acidosis in paediatric Sjögren syndrome. Rheumatology (Oxford) 45: 85-91.
15. Shibata N, Kitagawa K, Noda M, Sasaki H (2012) Solitary neurofibroma without neurofibromatosis in the superior tarsal plate simulating a chalazion. Graefes Arch Clin Exp Ophthalmol 250: 309-310.
16. Buonuomo PS, Maurizi P, Bracaglia C, Margani G, Riccardi R (2009) Primary Sjögren's syndrome mimicking lymphoma in a pediatric patient. A rare and possibly misdiagnosed condition. Minerva Pediatr 61: 119-122.