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Juvenile recurrent parotitis: clinical, sialographic and ultrasonographic features

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Objective. Juvenile recurrent parotitis (JRP) is a rare salivary gland disease of obscure aetiology that affects children. The aim of this study was to investigate the patterns of clinical presentation, and the sialographic and ultrasonographic features of JRP in Sri Lankan children.

Methods. The authors analysed the hospital records of 26 subjects who had been diagnosed with JRP between January 2003 and April 2006.

Results. The subjects consisted of 15 males and 11 females (male:female ratio = 1.4:1). The age range of the sample was 2.5–16 years (mean = 8.4 years). The age of onset was biphasic, with two major peaks at 6 years (n = 6) and 10 years (n = 5) (mean = 6.73 years). Unilateral involvement was seen in 69.2% of patients. The commonest clinical features were swelling (100%), pain (80.8%) and fever (50.0%). The average frequency of recurrences of JRP in 18 patients was 7.1 times per year. The average duration of an individual episode, also in 18 patients, was 5.44 days. Sialography in 17 patients had revealed punctate sialectasis, whereas ultrasonography in 16 patients had demonstrated multiple hypoechoic areas and heterogeneous echoes

Conclusions. This study documents the clinical features of JRP in Sri Lankan children. It has established the usefulness of sialography and ultrasonography in the diagnosis of JRP.

Introduction

Juvenile recurrent parotitis (JRP) is a well-recognized salivary gland disease in children, being second only to mumps in prevalence[1,2]. The disease is characterized by recurrent unilateral or bilateral parotid swellings accompanied by pain, fever and redness of the skin overlying the gland[3]. Juvenile recurrent parotitis is diagnosed mainly on the basis of a history of two or more episodes of inflammation of one or both parotid glands with or without the clinical features described above. Differentiating JRP from mumps may be difficult in small children[1].

Juvenile recurrent parotitis appears to have a world-wide distribution, with reports of cases coming from many different countries[1,3–10]. Very little is known of the incidence of JRP since most studies are of case series, implying that the condition is comparatively rare[3]. Although the aetiology and pathogenesis of this condition are largely unknown, several causative factors such as congenital malformation of the ductal system[11], hereditary and genetic factors[4,8], bacterial infection[9], autoimmune disease[10,12], and immunodeficient states such as hypogammaglobulinaemia[13], IgA deficiency[5,14] and IgG3 deficiency[15] have been implicated. There have also been reports indicating that JRP may resolve when the affected patient enters adulthood[16,17].

The aim of this paper is to analyse the patterns of clinical presentation, and the sialographic and ultrasonographic features of JRP in Sri Lankan children.

Subjects and methods

This study was carried out at the Oral Medicine Clinic of the University Dental Hospital, Peradeniya, Sri Lanka. The Dental Hospital is a tertiary care institution for dental, oral and maxillofacial diseases that deals with approximately 30 000

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patients per year. It receives referrals from medical and dental practitioners in district and peripheral hospitals, as well as private practice. Many patients are also referred at a primary level from the Admission Clinic of the Dental Hospital itself. The study was undertaken as a retrospective review of the case records of all patients under the age of 16 years who were diagnosed with JRP during the period from 1 January 2003 to 30 April 2006.

The diagnosis of JRP had been made on the basis of a history of two or more episodes of inflammation of one or both parotid glands in children under the age of 16 years presenting with unilateral or bilateral parotid swellings with or without pain, fever and redness of the overlying skin. The records were evaluated for demographic data, and information on age of onset, number of recurrences, duration of individual episode and family history of JRP, for example. In addition, data on clinical features such as unilateral or bilateral involvement, presence of swelling, fever, pain, redness of overlying skin, dry mouth, and purulent discharge were retrieved. The results of haematological, sialographic and ultrasonographic studies performed on these patients were also evaluated. The clinical records were evaluated by the first, second, fourth and fifth authors. The results of the sialographic and ultrasonographic examinations were evaluated by the first, second and third authors. Wherever available, the results of colour Doppler sonography (CDS) were also assessed. Clearance for the research was obtained from the Faculty Research and Ethical Committee.

Results

Twenty-six patients were identified who had been diagnosed with JRP based on the clinical criteria stated above. Table 1 shows the age and gender distribution among these patients. There was a slight preponderance of male subjects, with a male:female ratio of 1.36:1. The overall age range was 2.5–16 years, with a mean age of 8.44 years. Twenty-two patients (84.6%) had presented by the age of 10 years. The age of onset varied greatly (see Fig. 1), with a mean age of onset of 6.73 years. The minimum age of onset among the boys was 2 years and the maximum was 13. Among the girls, the minimum age of onset was 3 years and the maximum was 14. The time lapse between the onset of JRP and presentation to the hospital ranged from 3 months to 4 years.

Table 1. Age and gender distribution of patients with juvenile recurrent parotitis.

Variable	Males	Females
Minimum age (years)	2.5	4
Maximum age (years)	16	15
Age group (years):		
2–4	3	1
5–7	3	6
8–10	4	1
11–13	3	2
14–16	2	1
Mean age (years)	8.7	8.1
Total number of patients (%)	15 (57.7)	11 (42.3)
Side of the parotid affected [*n* (%)]:		
right	3 (11.5)	4 (15.4)
left	6 (23.1)	5 (19.2)
bilateral	6 (23.1)	2 (07.7)

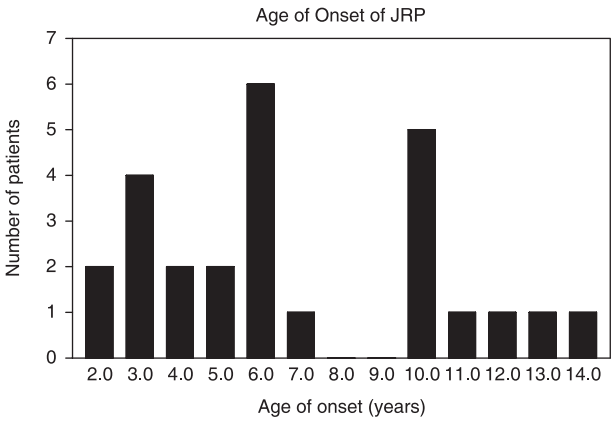

Fig. 1. Age of onset of juvenile recurrent parotitis.

The frequency of recurrences of JRP was recorded in only 18 patients (69.2%), and ranged from two to 15 times per year, with a mean of 7.1 times per year. Information on the duration of an individual episode was only available for an equal number of patients and varied from 4 days to 7 days, with a mean of 5.44 days.

Three patients (11.5%) described a positive family history of JRP. One of these subjects had a younger sibling as well as a mother with a positive history, while the other two had a mother and grandmother, respectively, who suffered from JRP.

Table 2. Clinical symptoms and signs of juvenile recurrent parotitis.

Clinical finding	Number of patients	Percentage
Swelling	26	100.0
Pain	21	80.8
Fever	13	50.0
Cloudy/viscous salivary secretion	11	42.3
Redness of overlying skin	7	26.9
Dry mouth	2	7.7
Purulent discharge from parotid papilla(e)	1	3.9

The right and left parotid glands were involved in seven (26.9%) and 11 (42.3%) subjects, respectively. Bilateral involvement was recorded in eight (30.8%) individuals. Table 1 shows the affected side according to gender. Simultaneous involvement of both the right and left parotid glands was encountered only in two (7.7%) patients.

Table 2 summarizes the clinical findings among the 26 subjects. Although a complaint of dry mouth had been recorded in two patients (7.7%), no measurement of salivary flow rate had been performed in any individual. Haematological investigations had been carried out in only 14 subjects. Six of these patients (23.1%) were mildly anaemic, with haemoglobin levels slightly less than the normal range (12–18 g dL^{-1}). Leucocytosis had been found in five of these 14 subjects. None of the patients had any significant history of systemic diseases.

The records showed that radiological investigations in the form of sialography (SG), ultrasonography (US) or both had been carried out in 22 patients (84.6%). Of these individuals, 16 (61.5%) had US performed on a total of 22 parotids, and 17 patients (65.4%) had been subjected to sialography on a total of 20 glands, with an overlap of 11 patients who had undergone both investigations (see Table 3). The typical SG finding in JRP is punctate sialectasis, and this feature was found in all 20 parotid glands in the 17 patients who underwent SG. No dilation or kinks were found by SG in the main parotid ducts of any of these subjects. Marked hypoechoic areas and heterogeneous echoes were the characteristic ultrasonographic findings in 21 of 22 parotids investigated with US. Of the 16 patients who had US, information on CDS was available only for seven individuals. Table 3 also shows the results of CDS in these seven patients. Follow-up CDS of three glands that had previously displayed hypervascularization showed normal blood flow after the resolution of symptoms.

Figures 2 and 3 show typical SG and US images, respectively.

All 26 patients had been managed conservatively with antibiotics, analgesics and oral hygiene aids with satisfactory results. The antibiotics prescribed varied from phenoxymethyl penicillin (penicillin V) for early episodes to amoxycillin for later ones. Erythromycin had also been used, alternating with amoxycillin, for recurrences or in those patients with a history of allergy to penicillin.

Discussion

As far as it can be ascertained, this is the first report of a case series of subjects with JRP

Table 3. Radiological investigations in children with juvenile recurrent parotitis*.

Investigation	Number of patients	Number of parotid glands	Hypoechoic areas in US†	Punctate sialectasis in SG†	Hyper-vascularization in CDS†	Normal Findings†
US only	5	8	8	–	–	0
CDS	7‡	10‡	–	–	7	3
SG only	6	6	–	6	–	0
Both US and SG:						
unilateral	8	8	7	8	–	1§
bilateral	3	6	6	6	–	0
Total	22	28	21	20	7	–

*Key: (US) ultrasound; (CDS) colour Doppler sonography; and (SG) sialography. †Number of parotid glands. ‡Included in US. §US.

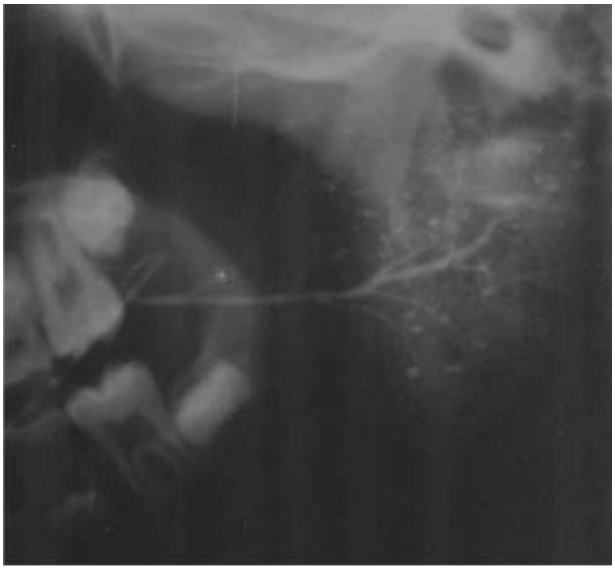

Fig. 2. Typical sialograph of juvenile recurrent parotitis showing punctate sialectasis.

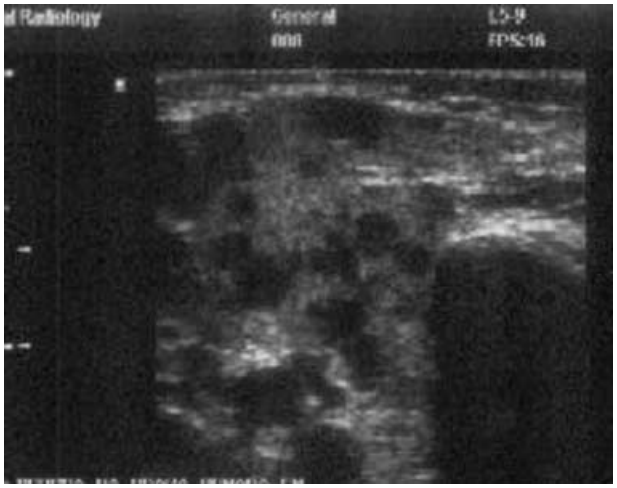

Fig. 3. Typical ultrasonogram of juvenile recurrent parotitis showing hypoechoic areas and heterogeneous echoes.

from Sri Lanka. The only patients included in the series are those who were referred to the University Dental Hospital during the period of the study; however, many cases of JRP might also have been referred to the paediatric departments of the two large general hospitals in the same area as the dental hospital.

The main condition from which JRP needs to be differentiated by the clinician is mumps. The latter condition occurs in epidemics, being spread by droplet infection, and is not associated with a history of recurrence. Mumps is also accompanied by a greater number of marked constitutional symptoms, such as a high fever, malaise, headache and chills[1], whereas JRP is associated with much milder systemic symptoms, and its diagnosis requires the fulfilment of the criterion of two or more episodes of parotid swelling. Moreover, ultrasonograms of parotids affected by mumps would display a homogenous echogenicity in contrast to the multiple hypoechogenic areas and heterogeneous echoes of JRP.

The age of onset in this study parallels that reported by Leerdam *et al.*[3], with a biphasic distribution. The first major peak in the former report was at 6 years of age, however, whereas that in the latter study was at 3 years, although there was also a minor peak in the present study group at 3 years. The second major peak in both studies occurred at 10 years of age. The cases in this study had a relatively late onset, with only 84.6% ($n = 22$) of the patients having undergone onset of the condition by 10 years of age in comparison with the findings of Leerdam *et al.*[3] (94%). The present study demonstrated unilateral involvement of JRP in 69.2% of patients and bilateral involvement in the remainder. This closely resembles the unilateral involvement of 66% in the report referred to above.

As in other reports, there was a slight preponderance of males among this group of subjects[3,18,24], but the gender difference was not as marked as in these other studies. Although Table 1 shows that a higher proportion of male patients were affected bilaterally, this observation is not statistically significant (Fisher's exact test, $P = 0.536$). Kolho *et al.*[4] and Reid *et al.*[8] reported a familial tendency among patients with JRP, with the former authors reporting a familial involvement as high as 22%. In the present series of patients, only three subjects (11.5%) had definite familial involvement. The mean duration of an individual episode in this series of patients (5.44 days) varied from the findings of Leerdam *et al.*[3] (3 days), probably because of the easy access to and early commencement of medical treatment for the individuals of the latter study.

Several authors[10,12,19–22] have advanced the possibility that JRP is a precursor of adult Sjogren syndrome (SS), with xerostomia as a presenting symptom and the existence of serological

evidence of the autoimmune disease in these patients. This concept appears to contradict the reports by Ericson *et al.*[16], and Galili and Marmary[17], who suggested that JRP abates when children enter adulthood. A plausible explanation for this is that only a subset of patients with JRP may be affected by SS. In this study, only two patients appeared to have dry mouth, although no eye symptoms were reported. It is tempting to speculate that xerostomia could also have been present in many of these patients, since this symptom was not investigated during history-taking and because young children are unlikely to complain of a dry mouth as a result of lack of experience of normal salivary flow status. Future prospective studies could pay attention to this aspect of investigation, together with tests for the detection of autoantibodies, and to rule out immunodeficiencies.

The incidence of common clinical findings in this study was generally similar to the findings of Leerdam *et al.*[3], particularly that of swellings of the parotids (100% in both studies). The percentage of patients with pain was, however, slightly lower in this study, and the proportion of patients who experienced fever was slightly greater than in the latter study. Many authors[3,23] have asserted that purulent discharge from the parotid duct is unusual in JRP. In this study, only one child had a purulent yellowish discharge.

Six of 14 children (42.9%) who underwent haematological investigations were found to be mildly anaemic in this study, and leucocytosis was found in five of these 14 patients (35.7%). Although these figures appear somewhat significant, it is not possible to make any comparisons since this aspect was not addressed in the literature surveyed.

Both SG and US are very successful methods of confirming sialectasis. As shown in Table 3, among the 22 patients who had these investigations performed, 14 parotids had been subjected to both investigations in 11 patients. Sialography had detected sialectasis in all 14 parotids, whereas US had found typical features of multiple hypoechoic areas and heterogeneous echogenicity in 13 parotids. Thus, there appeared to be a concordance of 92.86% ($n = 13$ of 14) when both investigations had been performed in same subjects. Sialography is an invasive procedure that requires considerable patient cooperation; however, it has also been reported to have a beneficial effect in terms of improving symptoms[1] and reducing the frequency of recurrence[25]. This has been the experience of the present authors too, although it is anecdotal. Nahlieli *et al.*[1] reported dilation and kinks in the main ducts in SG of their patients. Such features were not found in the present series.

Ultrasonography is a very useful technique for examining small children who are less cooperative. Shimizu *et al.*[26] recommended the use of US because they found that it has superior sensitivity to SG but the present study did not totally confirm this finding. Steiner *et al.*[27] and Gritzmann *et al.*[28] showed that CDS can be useful in the subset of subjects with JRP who have SS because it reveals hypervascularization of affected glands, although these results correspond to the extent of glandular destruction. This feature is not specific to SS since it is seen also in malignant lesions of the parotid[29]. The records of only seven patients in this series recorded the results of CDS investigations (Table 3). In these seven subjects, 10 parotid glands had been subjected to CDS. Seven parotids showed hypervascularization while three failed to show this feature. Follow-up CDS, after the resolution of the symptoms of three parotid glands that had previously displayed hypervascularization, showed normal blood flow. This may indicate that hypervascularization may also be a feature of an acute inflammatory response. No definitive conclusions can be drawn regarding this aspect because the information available in the records is insufficient.

All 26 patients were managed conservatively with antibiotics and supportive therapy. This conforms with the report by Leerdam *et al.*[3], the majority of whose patients had been treated with antibiotics. Isaacs[23], however, has stated that antibiotics are of little benefit.

A main caveat in this study is that the total number of subjects was relatively small. Moreover, the patients were not investigated for evidence of either SS or immunodeficiencies. This aspect must be taken into consideration in future prospective studies.

What this paper adds

- This study puts the Sri Lankan experience of JRP in the world-wide context.
- It describes the similarities and variations in the clinical presentation of JRP in a hitherto-unreported group of patients.
- It adds to the increasing awareness of the usefulness of SG and US, and to a limited extent, that of CDS, in the overall management of patients with JRP.

Why this paper is important to paediatric dentists

- This study enables them to recognize the clinical features of JRP and makes them aware of the condition as a distinct clinical entity.
- It underlines the need to refer individuals with JRP to appropriate specialists in order to properly investigate such patients.

Conclusions

This study found several similarities with other reports of JRP, including a preponderance of male subjects. It puts the Sri Lankan experience of JRP in the world-wide context, and documents the demographic and clinical features of this disease in a previously unreported group of patients. This study also demonstrates the usefulness of SG and US in diagnosis, and underlines the need to refer children with parotid swellings to hospital.

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