Juvenile onset of primary Sjögren's syndrome: changes in imaging findings during a 7-year progression

Sirs,

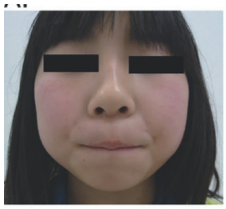
We present the case of a 10-year-old girl with primary juvenile Sjögren's syndrome (JSS); the onset and progression of salivary gland lesions were demonstrated by periodic magnetic resonance imaging (MRI) and ultrasonography (US) before the development of sicca symptoms.

The girl presented with erythema on her cheeks (Fig. 1A). She had no symptoms of oral or ocular dryness; however, we suspected JSS based on the cheek erythema and laboratory findings (Fig. 1B). Based on lymphocyte infiltration around the ducts (focus score: 2.28) in the labial gland biopsy, primary JSS was diagnosed by a paediatric rheumatologist. Therefore, treatment was initiated with an immunosuppressive agent and corticosteroid (Fig. 1B), and the erythema soon disappeared. Corticosteroid dosage was maintained within a range that avoided growth suppression or osteoporosis, while the immunosuppressive drugs were adjusted as needed (Fig. 1B). Nonetheless, at age 13, her serum amylase level was high; one year later, it markedly increased and remained high. Her IgG levels had been high since age 10 and further markedly increased at age 17 (Fig. 1B).

Although sicca symptoms were absent during the follow-up, at age 16, she developed acute left parotitis, which resolved after 3 days of antibiotic treatment. Two weeks later, Schirmer's and Saxon's tests were performed; only Schirmer's test was positive (Fig. 1B).

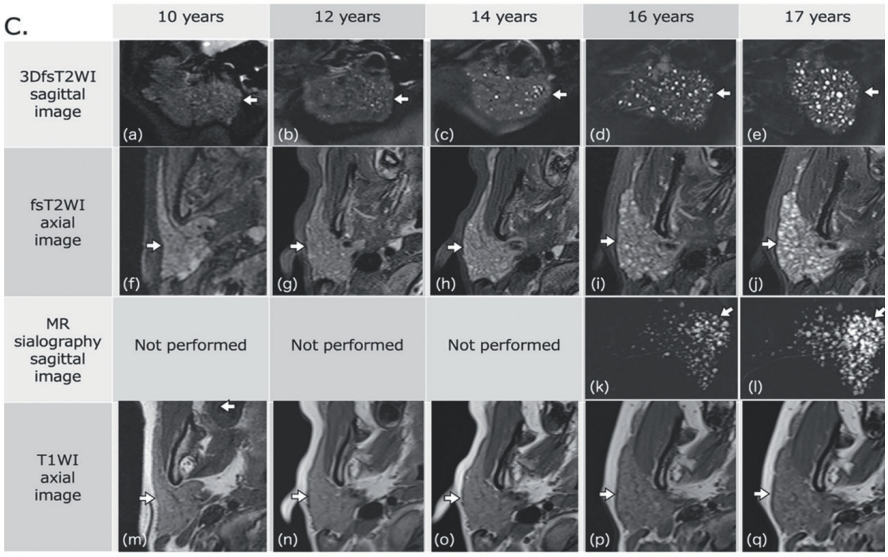
Periodic MRI and US from age 10 to 17 are shown in Figures 1C and D. The salivary gland MRI and US findings in patients with SS are characterised by fatty degeneration and punctate sialectasis (1, 2). In our case, hyperintense spots suggestive of punctate sialectasis appeared in both parotid glands from age 10 on fat-suppressed T2-weighted MR images (fsT2WI) using isotropic 3D turbo spin-echo imaging (3DfsT2WI) [Fig. 1C (a)], and from age 14 on conventional fsT2WI [Fig. 1C (h)]; the spots progressively increased in number and size during the follow-up [Fig. 1C (b-e), (i, j)]. MR sialography preformed only at ages 16 and 17 showed that the number and size of hyperintense spots increased between ages 16 and 17 [Fig. 1C (k, l)]. In contrast, the lace-like hyperintense areas on T1-weighted images suggesting slight fatty degeneration of the glandular parenchyma, were observed from age 16 [Fig. 1C (p, q)].

Although no abnormal findings were observed in the submandibular glands on MRI throughout the follow-up, the US at ages 16 and particularly 17 showed multiple



B.

	10 years	11 years	12 years	13 years	14 years	15 years	16 years	17 years
SS-A (U/mL)	125.9	126.6	134.7	-	>1200	-	>1200	-
ANA	1:160	-	-	-	1:1280	-	1:2560	1:2560
RF (IU/mL)	25.7	-	-	-	-	-	-	-
IgG (mg/dL)	2156	1717	1937	1950	1890	2218	2389	2803
Amylase (U/mL)	98	87	93	141	241	247	235	232
Saxon's test (g/2min)	-	-	-	-	-	-	5.18	-
Schirmer's test (mm/5min) *right, **left	-	-	-	-	-	-	*10, **5	-
Oral dryness	none	none	none	none	none	none	none	none
Ocular dryness	none	none	none	none	none	none	none	none
Other symptoms	Erythema on cheeks	none	none	none	none	none	Left parotitis	none
Imaging examination	MRI	-	MRI	-	MRI	-	MRI US	MRI US
Body weight (kg)	26	30	33	36	40	42	45	46
Immunosuppressive agent	Azathioprine 25mg/day				Mizoribine 50→100mg/day			Mycophenolate Mofetil 500mg/day
Corticosteroids	10→5→4mg/day			3mg/day			2mg/day	



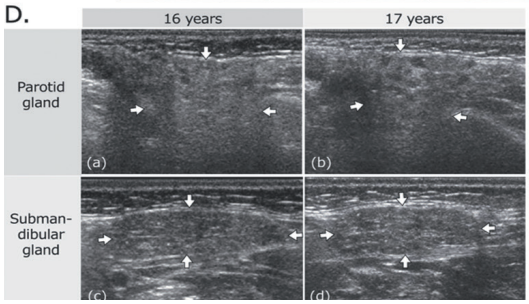


Fig. 1. A. On the initial visit at age 10, erythema was observed on bilateral cheeks.
B. Laboratory, clinical, and imaging results: ages 10-17 years. SS-A: anti- Ro/SSA antibody; ANA: antinuclear antibody; RF: rheumatoid factor; IgG: immunoglobulin G; MRI: magnetic resonance imaging; US: ultrasonography.
C. Periodic magnetic resonance imaging (MRI) of the right parotid gland: ages 10-17 years. Since the MRI findings were similar bilaterally, only the right side is presented. i) On 3DfsT2WI at 10 years of age, several hyperintense spots are observed (a); these spots progressively increased in number and size (b-e); ii) on conventional fsT2WI, at ages 10 and 12 years were unremarkable (f, g), at age 14 several hyperintense spots appeared (h); these spots progressively increased in number and size (i, j); iii) on MR sialography, the number and size of hyperintense spots increased between ages 16 and 17 (k, l) ; and i) T1WI at ages 10, 12, and 14 were unremarkable (m, n, o). T1WI at 16 years of age revealed abnormal lace-like hyperintense areas suggesting slight fatty degeneration (p) with a progressive increase during the follow-up (q). The arrows indicate the right parotid gland. 3DfsT2WI: fat-suppressed T2-weighted images with isotropic 3D turbo spin-echo imaging; fsT2WI: fat-suppressed T2-weighted images; MR sialography, magnetic resonance sialography; T1WI: T1-weighted images.
D. Ultrasonography images obtained at ages 16 and 17 reveal multiple hypoechoic areas in the right parotid and submandibular glands, more so at age 17 than at age 16.

hypoechoic areas within the parotid and submandibular glands (Fig. 1D), suggestive of punctate sialectasis of the peripheral duct and/or lymphocyte infiltration in the glandular parenchyma (1, 3, 4). Thus, the onset and progression of salivary gland lesions in primary JSS were clearly demonstrated by MRI and US before the development of sicca symptoms.

This is the first report of imaging changes concurrent with SS progression in a single patient. It has been reported that patients with JSS have fewer sicca symptoms than those with adult SS and that recurrent parotitis is the most common early symptom of JSS (5, 6); our patient developed acute parotitis at age 16 without sicca symptoms during the follow-up. Periodic MRI showed progressive punctate sialectasis suggestive of SS before the development of parotitis, suggesting that MRI can detect early slight abnormalities in JSS. Furthermore, this case supports the fact that inflammatory ductal changes caused by SS precede fatty degeneration in the glandular parenchyma (1); therefore, for early detection of JSS, it is important to check for peripheral ductal changes (1). Diagnosis of primary JSS is challenging. A labial gland biopsy is a useful yet invasive diagnostic tool. Hence, MRI and US should be considered useful non-invasive diagnostic tools for early diagnosis and evaluation of JSS progression.

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