

미만성 경화 아형 유두 갑상선암: 단일 기관에서의 17년 경험

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Diffuse Sclerosing Variant of Papillary Thyroid Carcinoma: A 17-year Experience at a Single Institution

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Purpose: The diffuse sclerosing variant of papillary thyroid carcinoma (DSPTC) is a rare histological subtype characterized by unique morphological features and aggressive behavior. The aim of this study was to evaluate the clinicopathologic features and outcome of DSPTC over 17 years.

Methods: Twenty-six cases of DSPTC (0.5%) were identified among 5,527 patients with papillary thyroid carcinoma treated between July 1990 and June 2007 at the Department of Surgery, Yonsei University College of Medicine. The clinicopathological features and outcome of these patients with DSPTC were evaluated. The median follow-up period was 46 months (range, 1-202 months).

Results: Twenty patients were females and 6 were males, the age ranged from 5 to 70 years (median 30.5 years). Histologically, most of the patients demonstrated diffuse involvement of one or both lobes of the thyroid, variable degree of lymphocytic infiltration, squamous metaplasia, psammoma bodies, extensive sclerosis and extracapsular extension, along with a high incidence of lateral neck node metastases (17/26, 65.4%). Treatment was by complete surgical resection by means of a total thyroidectomy (24/26, 92.3%), modified radical neck dissection (17/26, 65.4%) and

postoperative radioactive iodine therapy (22/26, 84.6%). Recurrences were noted in 6 cases (5 locoregional recurrence and 1 distant metastasis in bone). One patient died of an unrelated disease. The 10-year overall survival and disease specific survival rates were 83.8% and 100%, retrospectively, but disease free survival was 48.0%.

Conclusion: DSPTC showed an unfavorable clinical course with a low overall disease free survival. An aggressive therapeutic approach and close follow-up are recommended. (*J Korean Surg Soc* 2008;74:98-104)

Key Words: Papillary thyroid carcinoma, Diffuse sclerosing variant, Psammoma body, Squamous metaplasia

(papillary carcinoma)
(follicular carcinoma) (well-differentiated thyroid carcinoma, WDTC),
(medullary thyroid carcinoma, MCT), (undifferentiated carcinoma)
(anaplastic carcinoma)
(1,2)
1953 Crile Fisher(3)
(diffuse sclerosing variant)
0.7 5.3%
(4,7)

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중심 단어: 유두 갑상선암, 미만성 경화 아형, 사종체, 편평상피화생

22 16 (72.7%)
 1,998 IU/ml (, 11.7 17,947 IU/ml)

1990 1 2007 6 24 (92.3%)
 5,527 2 (7.7%)

26 (0.5%)
 (Table 1).

, 3 6
 , 10

1 (Table 2).
 2.1 cm (, 0.3 5.0 cm)

2007 6
 49 (, 4 205)
 10 6
 , 8

SPSS v12.0 for windows (Copyright 2003
 SPSS Inc. Chicago, Illinois, USA)
 Kaplan-Meier

6 (23.1%)
 4
 13 (50%)
 17 (65.4%) 18 (69.2%)
 (Table 2). (squa-
 mous metaplasia),
 (Fig 2).

11 9 , 42 13
 6 , 20
 305 (, 5 70) , 13
 (50%) 30
 5 (19.2%),
 1 (3.9%) , 20
 (76.9%) . 52 (, 1
 24) (Table 2). 100 mCi) , 17
 (150 200 mCi) (Table 2).

1 ,
 1 ,
 2
 (pseudo-inclusion) , (groove),
 25 (96.2%), 1
 (3.8%) . 1 (disease unrelated
 death)

(heterogenous hy-
 poechoogenicity), (ill-defined margin),
 (scattered microcalcification)
 (Table 3 Fig 1). 5 (19.3%)
 3 , 1 (Table 2).
 (thyroglobulin antibody) 41 (2 91)
 5

Table 1. Summary of patients with DSPTC (n=26)

Sex/ Age	Symptom duration, month	Surgery*	Tumor size [†] (cm)	Extra thyroidal invasion	Neck node metastasis			RAI (mCi)	Follow up, months	Recur
					Central neck node	Lateral neck node	Total			
M/21	Neck mass (8)	TT&MRND (ip)	Nodule (bi) 2.5	Y	5/5	7/33 (ip)	12/38	60	71	Yes
F/24	Neck mass (12)	TT&MRND (bi)	Nodule (uni) 0.6	N	16/16	15/24 (ip) 8/24 (contra)	39/64	100	57	Yes
F/27	Neck mass (21)	LT	Nodule (uni) 4.0	N				No	196	No
F/70	Neck mass (1)	LT&CCND	Nodule (uni) 0.3	N	/			No	131	No
F/42	Neck mass (1)	TT	Nodule (uni) 5.0	Y				60	66	Yes
F/5	Neck mass (3)	TT&MRND (bi) SMD	Nodule (uni) 4.0	Y	/	9/24 (ip) 9/24 (contra)	22/52 (SMD 4/4)	30	142	Yes
F/32	Neck mass (8)	TT&MRND (bi)	Diffuse goiter (bi)	N	4/4	11/25 (ip) 12/2 (contra)	27/52	60	71	No
M/13	Neck mass (3)	TT&MRND (ip)	Nodule (bi) 3.0	Y	8/9	6/29 (ip)	14/38	60	138	No
F/26	Neck mass (2)	TT&MRND (bi)	Nodule (bi) 1.1	Y	16/20	9/13 (ip) 4/12 (contra)	29/45	200	17	No
F/26	Neck mass (5)	TT&MRND (ip)	Nodule (uni) 1.0	Y	6/8	10/35(ip)	16/43	200	16	No
F/50	Incidentaloma (breast cancer)	TT&CCND	Nodule (bi) 1.0	N	/			No	17	No
F/31	Neck mass (1)	TT&MRND (bi)	Diffuse goiter (bi)	Y	2/2	8/41 (ip) 7/26 (contra)	17/69	200	13	No
F/15	Neck mass (2)	TT&MRND (ip)	Diffuse goiter (uni)	N	7/7	12/36 (ip)	19/43	60	118	No
F/20	Neck mass (7)	TT&CCND	Diffuse goiter (uni)	Y	0/4		0/4	60	96 (death)	Yes
M/30	Incidentaloma	TT&MRND (bi)	Nodule (bi) 2.5	Y	20/20	11/47 (ip) 9/12 (contra)	40/79	150	5	No
M/39	Neck mass (1)	TT&MRND (bi)	Diffuse goiter (bi)	N	14/17	8/26 (ip) 5/16 (contra)	27/59	200	1	No
F/43	Incidentaloma	TT&MRND (ip)	Nodule (bi) 1.1	Y	10/11	14/32 (ip) 3/3 (contra)	27/46	200	1	No
F/50	Neck mass (6)	TT&CCND	Nodule (bi) 0.3	N	0/5		0/5	No	202	No
M/32	Neck mass (1)	TT&CCND	Nodule (uni) 3.0	Y	10/19		10/19	100	5	No
F/35	Incidentaloma	TT&MRND (ip)	Nodule (bi) 1.3	Y	8/9	10/28	18/37	150	5	No
F/39	Neck mass (1)	TT&CCND	Nodule (bi) 5.0	Y	13/18		13/18	100	2	No
F/26	Hoarseness (24)	TT&MRND (bi)	Nodule (bi) 0.5	Y	11/11	11/33 (ip) 8/25 (contra)	30/69	‡	2	No
F/37	Neck mass (1)	TT&CCND	Nodule (bi) 2.5	Y	3/9			30	1	No
F/20	Neck mass (1)	TT&MRND (ip)	Nodule(bi) 1.5	Y	7/8	5/31	12/39	‡	1	No

Table 1. Continued

Sex/ Age	Symptom duration, month	Surgery*	Tumor size [†] (cm)	Extra thyroidal invasion	Neck node metastasis			RAI (mCi)	Follow up, months	Recur
					Central neck node	Lateral neck node	Total			
F/32	Incidentaloma	TT&MRND (bi)	Nodule (bi) 1.1	Y	/	7/28 (ip) 7/25 (contra)	14/53	150	13	No
M/9	Neck mass (1)	TT&MRND (bi)	Diffuse goiter (bi)	Y	15/16	8/30 (ip) 5/24 (contra)	28/70	100	12	No

*TT = total thyroidectomy; LT = less than total thyroidectomy; CCND = central compartment node dissection; MRND = modified radical neck dissection; [†] Nodule (uni) = nodule on unilateral lobe; Nodule (bi) = nodule on bilateral lobes; Diffuse goiter (uni) = papillary cancer involve unilateral lobe without nodular lesion; Diffuse goiter (bi) = cancer involve bilateral lobes without nodular lesion; Ip, ipsilateral; Bi = bilateral; Contra = contra lateral; SMD = superior mediastinum dissection; RAI = radio active iodine treatment; = lymph node dissection was not done; / = lymph node dissection was done but no report; [‡] RAI at other hospital.

Table 2. Clinicopathologic features (n=26)

Gender	
Male	6 (23.1%)
Female	20 (76.9%)
Age at first diagnosis	
Mean (years)	30.5±13.7
Range (years)	5-70
Symptoms and Signs	
Anterior neck mass	20 (76.9%)
Incidentaloma	5 (19.3%)
Hoarseness	1 (3.8%)
Operation type	
Total thyroidectomy only	7 (26.9%)
Total thyroidectomy with MRND (Ip)	7 (26.9%)
Total thyroidectomy with MRND (Bi)	10 (38.5%)
Less-than total thyroidectomy	2 (7.7%)
Size of tumor (nodule forming case)	
Mean (cm)	2.1±1.5
Range (cm)	0.3-5.0
Capsule invasion	18 (69.2%)
Multifocality	13 (50%)
Bilaterality	17 (65.4%)
Postoperative RAI treatment	22
Low dose (30-100 mCi)	13
High dose (150-200 mCi)	7
Unknown dose	2
Recurrence	5 (19.3%)
Central and lateral lymph node	1
Lateral lymph node	3
Mediastinum lymph node	1
Duration of recurrence	
Mean (month)	41
Range (month)	2-91
Distant metastasis (bone, metachronous)	1 (3.8%)

MRND = modified radical neck dissection; Ip = ipsilateral; Bi = bilateral; RAI = radio-active iodine treatment.

Table 3. Ultrasonographic features (n=26)

Echogenicity	
Hyperechoic	1 (3.8%)
Isoechoic	1 (3.8%)
Hypoechoic	24 (92.4%)
Microcalcification	
Diffuse scattered	25 (96.2%)
Absent	1 (3.8%)
Mass forming	
Present	20 (76.9%)
Absent	6 (23.1%)
Margin (n=20)	
Blurred	18 (90%)
Defined	2 (10%)

(regional lymph node dissection)

, 1 2 7

. 5

(150-200 mCi)

26 8 6

1 (3.8%)

100%

Kaplan-Meier

(10 year overall survival rate, OS) 83.3%, 10

(10 year disease-free survival rate, DFS) 48.0%

(Fig. 3).

Crile Fisher(3)

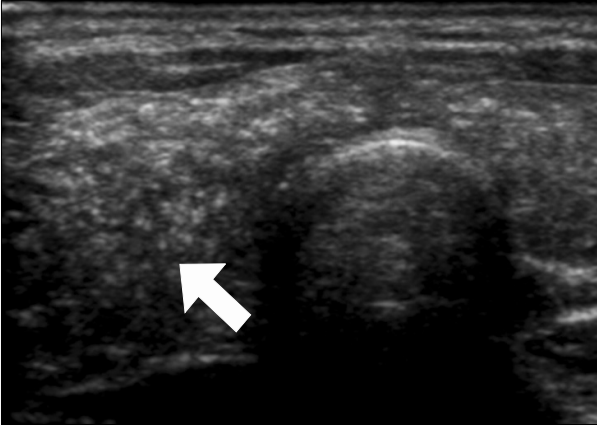


Fig. 1. Ultrasonographic findings in transverse section showing diffuse scattered microcalcifications (white arrow) occupying nearly the entire right thyroid.

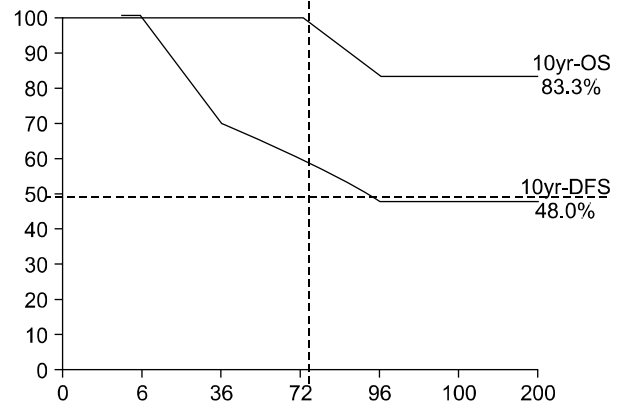


Fig. 3. 10-year overall survival (OS) and 10-year disease-free survival (DFS).

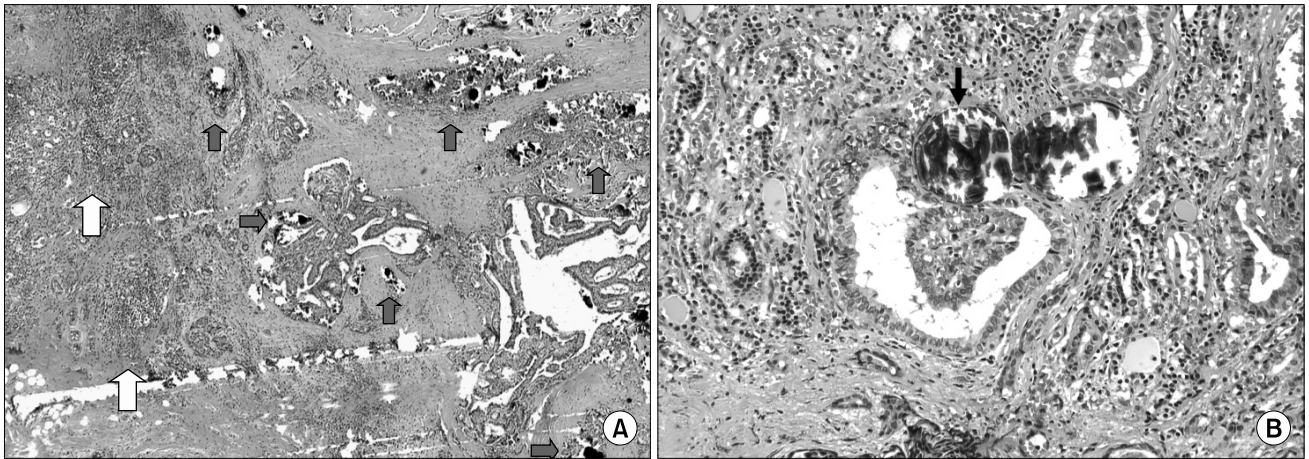


Fig. 2. Microscopic pathologic findings of DSPTC. (A) Lower power view showing interstitial fibrosis, a lymphocytic inflammatory infiltration (white arrow), and psammoma bodies (gray arrow) (H & E stain $\times 40$). (B) Higher power view presenting the usual cytological features of papillary thyroid carcinoma, and psammoma bodies (black arrow) (H & E stain $\times 400$).

2/3

1/3

(5,7,8)

(overlapping)

(9)

(11,12)

(thyroglobulin)
(carcinoem-

cytokeratin, calcitonin
bryonic antigen, CEA)

(10)

(12)

(papillae)

, 1
 . Lam Lo(15)
 , 1
 .(57,10,11,13 Falvo (14)
 X
 .(25)
 5 , 1
 , 10 48.0%
 Lam Lo(15)
 , 83%
 30
 ,(11,14,15) Moreno (21)
 .(12,14,17)
 Carcangju Bianchi(11) 20
 10%
 20 35%
 30.5 , 30
 50%, 6 20 , 19.3%
 (150 200 mCi)
 ,(11,15) (20 , 76.9%)
 ,(7,19,22)
 ,(12,14,23)
 40%
 .(7,11)
 21 cm (, 0.3 5
 cm), 13 (50%),
 17 (65.4%), 18 (69.2%)
 .(7,11,12,14,19,22,23)
 ,(5,11,14,15)
 ,(11,15,24) 17 (65.4%)

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