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Case report

Two patients with TAFRO syndrome exhibiting strikingly similar anterior mediastinal lesions with predominantly fat attenuation on chest computed tomography

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ABSTRACT

We herein report on two middle-aged men with TAFRO (thrombocytopenia, anasarca, fever, reticulin fibrosis or renal failure, and organomegaly) syndrome, a unique clinicopathological variant of multicentric Castleman's disease recently proposed in Japan. Strikingly similar anterior mediastinal fat swellings with soft tissue density were observed in the patients on chest computed tomography. In TAFRO syndrome, bilateral pleural effusion and slight lymph node swelling are common in the thoracic region; however, anterior mediastinal lesions have not been previously observed. Although the mechanisms of anterior mediastinal lesions have not been defined, these lesions seem to have a close relationship with TAFRO syndrome.

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1. Introduction

TAFRO (thrombocytopenia, anasarca, fever, reticulin fibrosis or renal failure, and organomegaly) syndrome is considered as a clinicopathological variant of multicentric Castleman's disease (MCD) that has been identified in Japan [1]. Recently, newly proposed diagnostic criteria of TAFRO syndrome were published [2]. In the thoracic part, previous reports have shown that affected patients usually had bilateral pleural effusion and slight lymph node swelling. However, mediastinal lesions in patients with TAFRO syndrome have not yet been described [1–11].

We herein report two cases of TAFRO syndrome patients who showed characteristic anterior mediastinal lesions on chest computed tomography (CT).

2. Case presentation

2.1. *Case 1*

A 46-year-old Japanese man who smoked came to our hospital with general fatigue, dyspnea, and chest discomfort. He had no remarkable medical history. Physical examination at the initial visit showed a height of 176.0 cm, body weight of 79.8 kg, arterial blood pressure of 106/58 mmHg, pulse rate of 96/min, respiratory rate of 24/min, and body temperature of 38.0 °C. Breath sounds were low in the bilateral lower lungs. Pitting edema was present in the lower extremities. No superficial lymphadenopathy was observed. His laboratory examination findings are shown in Table 1. He had a slightly increased white blood cell count, thrombocytopenia, elevation of C-reactive protein (CRP) levels, slight renal dysfunction, liver dysfunction, hypoalbuminemia, and elevated d-dimer levels. Although his antinuclear antibodies were 80x, the results of the tests for other autoantibodies were negative. The results of the tests for the antibodies of human immunodeficiency virus (HIV), human T-lymphotrophic virus type 1 (HTLV-1), human herpes virus type 8 (HHV-8), and Histoplasma were all negative. The serum immunoglobulin levels were normal. Soluble interleukin-2 receptor (sIL-2R), IL-6, and vascular endothelial growth factor (VEGF) values were high. A chest X-ray on admission showed bilateral pleural effusion and enlargement of the mediastinal shadow (Fig. 1A). Chest and abdominal CT revealed bilateral pleural effusion, hepatosplenomegaly, slight mediastinal lymph node swelling, and anterior mediastinal fat swelling with soft tissue density distributed in a patchy fashion and vascular proliferation (Fig. 1B and C). T1-weighted fat-suppressed magnetic resonance (MR) imaging of the thorax showed anterior mediastinal lesion mainly with soft tissue intensity (Fig. 1D). Although we started the patient on a regimen of antibiotics, he became increasingly ill. Biopsy of the anterior mediastinal lesions was performed using a thoracoscope on day 8 of hospitalization. The biopsy

Table 1 – Laboratory data of the two patients at the initial hospital visit.

Laboratory data								
	Case 1	Case 2			Case1	Case 2		
WBC	9000	39730	/μL	CRP	23.21	22.75	mg/dL	
Net	78.6	95	%	IgG	1577	1104	mg/dL	
Mon	9.9	5	%	IgA	354	196	mg/dL	
Eos	0.2	0	%	IgM	47	243	mg/dL	
Baso	0.2	0	%	Anti-nuclear antibody	80	40		
Lym	11.1	0	%	Ferritin (<280)	467	735	ng/mL	
RBC	438×10^4	444×10^4	/μL	EBV VCA IgM	<10	<10		EBV: Epstein-Barr virus
Hb	14.5	12.4	g/dL	EBV VCA IgG	80	40		VCA: viral capsid antigen antibody
Ht	42.9	35.5	%	EBV EBNA	20	40		EBNA: EBV nuclear antigen
Plt	8.4×10^4	4.3×10^4	/μL	HIV antibody	(–)	(–)		HIV: human immunodeficiency virus
TP	6.1	5.7	g/dL	HTLV1 antibody	(–)	(–)		HTLV-1: human T-cell leukemia virus-1
ALB	2.5	2.7	g/dL	HHV-8 antibody	(–)	(–)		HHV-8: human herpesvirus-8
UN	12.9	26	mg/dL	Histoplasma antibody	(–)	Not done		
Cre	1.21	1.43	mg/dL					
Na	137	139	mmol/L	Soluble IL-2R (<421)	1735	1896	U/mL	IL-2R: interleukin-2 receptor
K	4.5	3.8	mmol/L	IL-6 (<2.41)	32.6	49.7	pg/mL	IL-6: interleukin-6
Cl	108	104	mmol/L	VEGF(plasma) (<38.3)	86.1	224	pg/mL	VEGF: vascular endothelial growth factor
AST	18	37	IU/L					
ALT	18	13	IU/L	**Urinalysis**				
γ-GTP	133	67	IU/L	PH	5.5	5.0		
T-bil	1.18	4.07	mg/dL	SG	1.019	1.015		
ALP	497	616	IU/L	Protein	+	+		
LDH (<230)	185	369	IU/L	Glucose	–	–		
				Occult blood	–	+		
PT	14.1	15.9	sec					
APTT	30.2	64.2	sec					
FIBG	695	482.5	mg/dL					
FDP-DD	5.9	42.8	μg/mL					

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